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CLINICAL CASES

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Epidural hematoma of the vertex. Report of three cases

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Abstract

Introduction: *Epidural hematomas of the vertex occur after trauma that produces diastasis or fracture of the sagittal suture, accompanied by tearing of the superior sagittal sinus, diploe bleeding, or dural tear.* **Methods:** *The objective of the study was to show the different treatment options for patients with epidural hematomas of the vertex.* **Discussion and Conclusions:** *The series presented here contrasts the management offered to each patient, where the first case had a scheduled surgical resolution after medical surveillance, while the other two cases were not surgically managed and were only kept under surveillance.*

Keywords: *Vertex epidural hematoma. Skull fracture. Head injury. Epidural hematoma.*

Introduction

Epidural hematomas (EH) are extra-axial bleeding caused by hemorrhage from the middle meningeal artery in 85% of cases and, to a lesser extent, related to venous sinus tears (e.g., transverse or sigmoid sinuses in the posterior fossa)[1-6]. HE of the vertex (HEV) differs by bleeding of venous origin from the superior longitudinal sinus[1-6]. These frequently occur due to trauma that produces diastasis or fracture of the sagittal suture, accompanied by a tear of the superior sagittal sinus, bleeding from the diploe, a dural tear of the internal table of the skull, or associated diseases (e.g., vascular diseases in the skull)[1,7,8].

The incidence of HE in traumatic brain injury (TBI) is 2.5-4%, and only 8% of these correspond to the vertex; HE pre-dominates in the male population (87%), between the second and third decade of life, the main causes of which are road accidents (48%), armed robbery (21%), and falls (17%)[1-3,8-12].

The diagnosis suspicion of HEV exists due to a history of a direct impact on the vertex, the presence of diastasis, and/or fractures that cross the coronal or sagittal suture, as observed by imaging[5,11]. The prognosis is favorable even though the growth of the hematoma is rapid and its stabilization is also rapid. They have an average of 6.45 days of hospital stay and a complication rate of 2.97%.1,4 An associated mortality is estimated at 18 to 50%.

Herein, three similar cases are discussed with the aforementioned history and with complementary imaging studies with epidural hematomas of the vertex.

Case reports

Case 1

A 28-year-old man, with no significant history, denies ingestion of alcohol or recreational drugs, suffers, the previous day, a fall from his own height with direct contusion

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in the occipital region, a loss of awake state for 30 min (approximately), and does not have abnormal movements or seizures reported. Two hours after the accident, he began to experience intense headaches, dizziness, nausea, and vomiting without deterioration in alertness. The bone series of the skull and cervical spine show an image suggestive of a fracture in the cranial vault. 48 h after the event, the patient is conscious, oriented, and active, with a Glasgow Coma Scale (ECG) score of 15 points, with no alterations in the cranial nerves, locomotor system, or sensory organs. He presents pain on palpation in the right occipital and parietal regions. A simple skull tomography was performed, which shows epidural hematoma at the parietal level toward the convexity and hemorrhagic contusion in the left temporal lobe, with evidence of a linear fracture of the right frontal bone and partial diastasis of the sagittal suture (Fig. 1).

A craniotomy was performed at the vertex, leaving a subgaleal drainage. The patient evolved favorably, with resolution of headache and nausea, without neurological sequelae, and was discharged 7 days after admission.

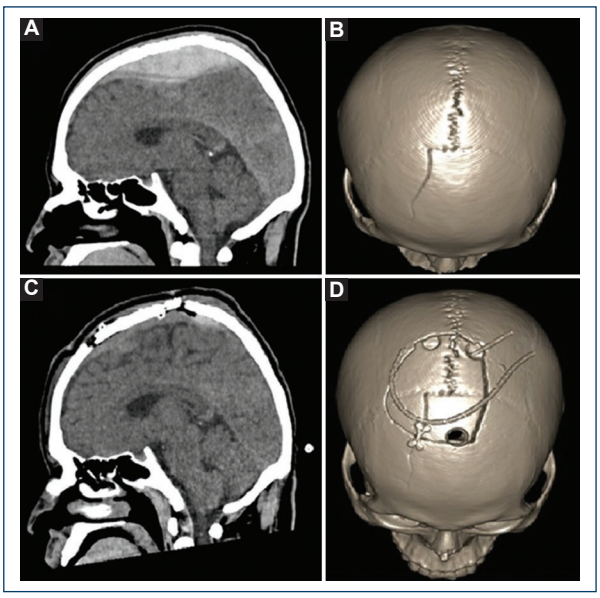

Figure 1. Simple skull tomography. **A:** sagittal sections are seen at the midline level where a hyperdense image of biconvex lenticular characteristics is observed that displaces the cerebral cortical surface from ventral to caudal. **B:** three-dimensional reconstruction, sagittal suture diastasis is seen with an irregular line drawn from posterior to anterior in the frontal bone just in front of the metopic suture. **C:** sagittal control section, post-surgical changes are observed at the level of the vertex with a residual image of smaller volume that does not displace the cerebral cortex. Adequate differentiation of sulci and gyri is seen without apparent evidence of associated pneumocephalus. **D:** three-dimensional reconstruction at the level of the vertex, craniotomy limits can be seen with the presence of a single initiating trephine, as well as the presence of osteosynthesis material at the level of the right frontal.

Case 2

A 29-year-old man, with no significant history, suffered aggression by a blunt object with multiple impacts to the head and had an apparent loss of consciousness of undefined evolution, associated with intense headaches and vomiting on three occasions, without convulsive seizures. On physical examination, the patient was conscious, with an ECG at 15 points, bipalpebral ecchymosis, 3 mm pupils, photomotor and consensual reflexes present, cranial nerves without alterations, and motor and sensory systems without alterations. Head without evidence of depression on palpation, without breaks in the scalp; extremities with bruises on the upper limbs.

The tomography showed a skull fracture in the right frontoparietal region associated with right frontal contusion and subdural hemorrhage (Fig. 2A and B).

Due to his stable neurological status, expectant management was decided, with no neurological changes or targeting data. The control computed tomography scan 4 days after his injury showed no changes compared to the previous one (Fig. 2C). After 9 days of monitoring, there was a remission of symptoms without complications.

Case 3

A 36-year-old man with road accident caused by a motor vehicle ejected three meters away. He has no loss of wakefulness, seizures, nausea, or vomiting. On ECG examination of 14 (O3V4M6) non-cooperative, 4 mm normoreflex pupils, facial symmetry, adequate hearing, shoulder symmetry, overall strength 5/5 in MRCs, and preserved sensation.

The simple skull tomography (Fig. 3) performed 1 day after the accident showed a complex bilateral parietal fracture with sagittal suture diathesis and subsidence, HEV < 15 mm, right laminar frontoparietal acute subdural hemorrhage, left frontal and temporal punctate hemorrhagic contusion, and pre-dominantly post-traumatic subarachnoid hemorrhage. in the posterior fossa at the level of the left optic-carotid cisterns, ambiens, crural, left lateral prepontine, and lateral pre-bulbar, as well as in the quadrigeminal plate Greene 2. The patient was treated with mannitol, diphenylhydantoin, buprenorphine, haloperidol

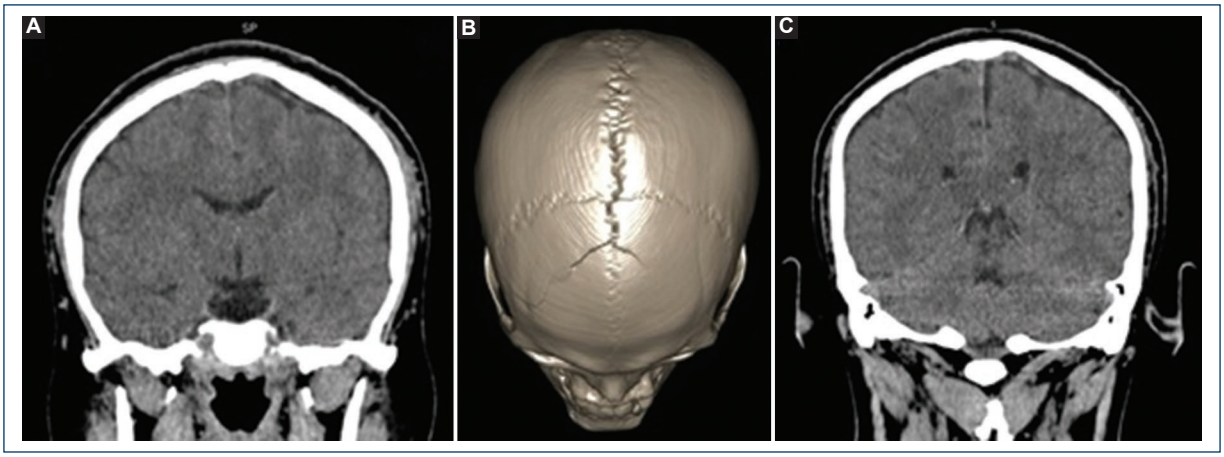

Figure 2. Simple skull tomography and three-dimensional reconstruction. **A:** coronal sections at the level of the frontal horns where a laminar image of minimal volume can be seen at the level of the vertex, which does not affect the mass effect. No dilation and/or morphological changes are seen at the level of the ventricular system. **B:** three-dimensional reconstruction, a linear trace is seen in the direction of the sagittal suture from posterior to anterior that covers the midline and is directed toward both sides in relation to pre-dominance on the right side. **C:** coronal cuts after 9 days of hospital stay. A well-known laminar image is observed without changes in relation to the previous tomography.

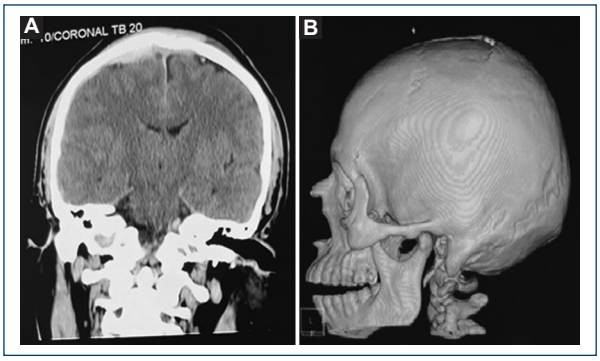

Figure 3. Simple skull tomography. **A:** coronal section where a laminar hyperdense image is observed at the parietal level that crosses the midline with bilateral distribution. In addition, there is probable soft tissue edema and/or evidence of subgaleal hematoma at the vertex level on the right side of the head. **B:** three-dimensional reconstruction lateral view. Fracture-sinking is observed at the level of the cranial vertex.

hypertonic solutions, and ondansetron. He presents a favorable evolution and is discharged after a week of surveillance.

Discussion

The case series presented here is consistent with the clinical and epidemiological characteristics known through current medical literature reports. Although HEVs are already a known entity, the objective of this document was to recapitulate the topic due to its diagnostic difficulty that still represents a diagnostic and therapeutic challenge, due to the technical limitations associated with the lack of efficient tomographs within some second-level hospital units.

The clinical evolution with an initial loss of awake state is well known, followed by a period of lucidity to again suffer neurological deterioration in almost half of patients with HE; however, the clinical course of HEV is variable.

The clinical manifestations vary from rapid neurological deterioration to a slow and progressive onset of symptoms, where most of the symptoms will depend on the location, the most common being an intense headache associated with nausea or vomiting that may be accompanied by paraparesis that is related to compression of the motor cortex. In addition, there may be secondary manifestations due to blockage of arachnoid granulations such as hydrocephalus and intracranial hypertension associated with visual alterations, papilledema, or anisocoria related to a mortality rate of 29.7%. On the other hand, the cranial nerves usually do not present alterations. However, paralysis of the first, third, fourth, and sixth cranial nerves has been reported[2,3,5,6,8,10,13-17].

The difficulty of imaging diagnosis is described due to the confusion of the isodensity of the hematoma with the adjacent bone and the alignment of the hematoma

along the tomographic section. Furthermore, the different tomographic projections of thick sections (commonly 10 mm) prevent or limit visualization of the hematoma. Today, there are more improved diagnostic tools whose software makes 3D reconstructions of bone structures possible. Likewise, there are alternatives such as angiography, which provides information on the location of the hematoma, which is why its use has been suggested in head trauma when there are signs of intracranial hypertension without tomographic alterations. Another less invasive diagnostic tool is magnetic resonance angiography, which, unfortunately, is not accessible in all medical units[5,6,8,15].

Treatment should be individualized according to the patient's neurological conditions, lesion volume, location, and clinical evolution. Thus, small hematomas with no symptoms may not require surgical intervention. The current indications for surgery, according to the Brain Trauma Foundation (BTF), are HE > 30 cm^3 without considering the ECG, and surveillance can be maintained in HE < 30 cm^3 and < 15 mm thick with < 5 mm deviation of the midline (MLD) in patients with ECG > 8 without focal deficits. Follow-up with simple tomography should be considered within the first 6 or 8 h for those in whom expectant treatment is chosen[4-6,18].

Surgical management is recommended by means of a wide decompressive craniectomy through the MLD, which includes the margins of the hematoma, its evacuation, and control of bleeding. The craniectomy approach is performed at the vertex with a single flap or bilaterally parasagittally. There are less-used alternatives, such as percutaneous aspiration, unilateral craniectomy, or trephine drainage. The most common and important complication is associated with blood loss that occurs when exposing the superior sagittal sinus on which the hematoma sits and air embolism[5-7,11].

High mortality is also influenced by scores < 8 in the ECG assessment at hospital admission. It has been observed that the results are favorable in 54.3% of cases if there is drainage of the hematoma within the first 70 min after the appearance of mydriasis[1,2].

The series of cases presented here contrasts the management offered to each of the patients, where the first case had a scheduled surgical resolution after medical surveillance. This was due to the persistence of symptoms suggestive of intracranial hypertension. On the other hand, the other two cases did not require surgical management; they were only kept under surveillance with symptomatic management due to the increase in symptoms and signs that could lead to neurological deterioration. The surgical approach in the first case was performed through a craniotomy that contained the margins of the HEV, with a satisfactory evolution.

Conclusions

In this series of cases with HEV, the different most common etiologies were presented (accidental falls, skull contusions, and traffic accidents); all cases presented headache as the pre-dominant symptom. Likewise, in all of them, a skull fracture is demonstrated, which includes the coronal or sagittal sutures and the presence of subsidence in the third case. Only the third case had moderate TBI at the time of the initial evaluation; however, it did not require surgical management, which demonstrates the correct use of the BTF criteria.

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Conflicts of interest

The authors declare no conflicts of interest.

Ethical disclosures

Protection of human and animal subjects. The authors declare that no experiments were performed on humans or animals for this study.

Confidentiality of data. The authors declare that they have followed the protocols of their work center on the publication of patient data.

Right to privacy and informed consent. The authors have obtained the written informed consent of the patients or subjects mentioned in the article. The corresponding author is in possession of this document.

Use of artificial intelligence for generating text. The authors declare that they have not used any type of generative artificial intelligence for the writing of this manuscript or for the creation of images, graphics, tables, or their corresponding captions.

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Left pulmonary artery ectopia with origin over ascending aorta: case report

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Abstract

Introduction: *The anomalous origin of a pulmonary artery over the ascending aorta is an extremely rare congenital malformation that comprises only 0.3% of congenital cardiac malformations.* **Case presentation:** *The presentation of this condition can be severe and early after birth, or asymptomatic, as in those cases where the ectopia is isolated. Diagnosis is complicated and requires a high level of suspicion due to the non-specific clinical picture.* **Conclusion:** *Direct reimplantation of the ectopic pulmonary artery on the pulmonary trunk remains the treatment of choice for these patients. We present the case of a 21-year-old female patient with this pathology.*

Keywords: *Pulmonary artery. Ascending aorta. Congenital malformation. Ectopia.*

Introduction

The ectopic origin of a pulmonary artery on the ascending aorta is an extremely rare congenital malformation that occurs in only 3% of congenital cardiac malformations, with the left pulmonary artery being affected 4-8 times less than the right[1,2]. The hemodynamic and pulmonary repercussions of this condition require early diagnosis and surgical correction to avoid irreversible pulmonary, cardiac, and hemodynamic complications[2-4].

The aim of this article is to present an atypical case of this congenital malformation that usually manifests in the first months of life and is associated with severe cardiac malformations.

Case presentation

The patient is a 21-year-old female who denies smoking or alcohol consumption. She has no known chronic degenerative diseases or other relevant pathological personal history. The only relevant background is her parents reporting growth retardation during infancy, with weight and size lower than appropriate for her age during her childhood.

Her condition began 2 years earlier with episodes of high-intensity migraine, after evaluation by a neurologist, she was referred to cardiology when a cardiac murmur was found. An audible *holosystolic* murmur was identified in all auscultation points, with greater intensity at the aortic point. Subsequently, more symptoms began to appear, such as palpitations, dyspnea, intolerance to moderate physical activity, stabbing precordial pain with no apparent association, tingling sensation in thoracic limbs, hemoptysis on multiple occasions, and epistaxis. A transesophageal echocardiogram was requested, which reported left ventricular dilatation with an ejection fraction of 41%, double aortic lesion with severe

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stenosis, and moderate valvular insufficiency; for these reasons, it was decided to perform an aortic valve replacement. During the surgical procedure, an intact aortic valve was found, with no stenosis or insufficiency. An exploration of the large vessels found a probable double aortic arch; it was decided to conclude the surgical procedure without performing valvular replacement. The patient recovered satisfactorily from the procedure; however, symptoms persisted. She underwent a CT angiography for a detailed revaluation (Fig. 1), where an anomalous origin of the left pulmonary artery over the ascending aorta, right aortic arch without retroesophageal component, and left lung with thickening of the interstitium and diffuse ground glass appearance (suggesting of congestive etiology) were described (Figs. 2 and 3). The patient was admitted for cardiac catheterization and diagnostic ventriculography to confirm the findings and better characterize them. After the procedure, the emergence of the left pulmonary artery from the ascending aorta, and the single drainage of the right ventricle over the right pulmonary artery was demonstrated, and no other abnormalities were found. Surgery was decided to reconnect the left pulmonary artery to the pulmonary trunk (Figs. 4 and 5).

The procedure was carried out in which aortic plasty with subsequent anastomosis of the left pulmonary artery to the pulmonary trunk was performed, no complications or eventualities occurred during surgery. The patient recovered satisfactorily after 5 days of hospital stay. In the long-term follow-up, the patient denies any of the previous symptoms such as dyspnea, fatigue, or chest pain.

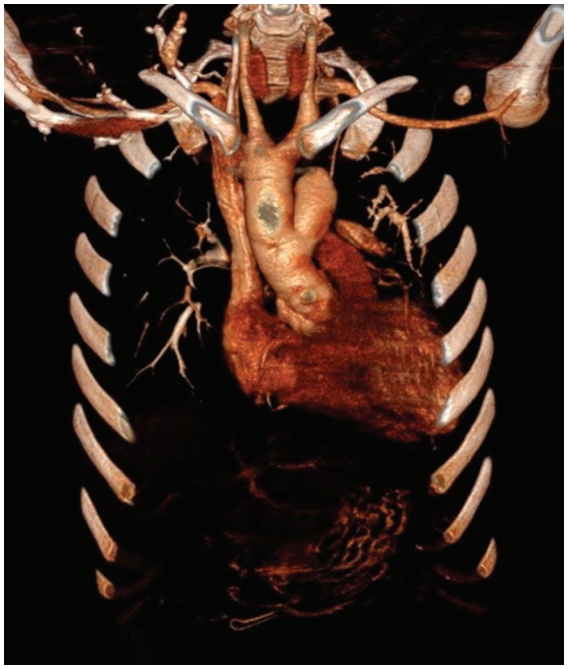

Figure 1. Coronal Volume rendering from CT angiography shows the abnormal left pulmonary artery arises from the left posterior vascular wall of the ascending aorta above the aortic valve annulus.

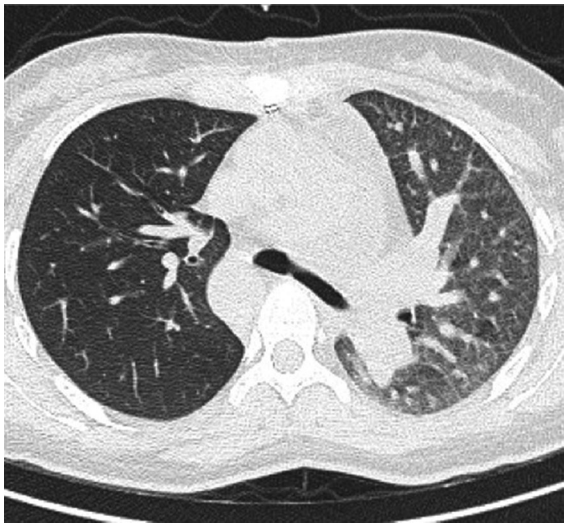

Figure 2. Axial CT lung window shows interstitial thickening and ground-glass appearance of left lung parenchyma.

Discussion

The origin of one pulmonary artery over the ascending aorta, in conjunction with the normal location of the other pulmonary artery on the pulmonary trunk, is an uncommon abnormality, accounting for only 0.05-0.3% of congenital heart diseases[1,5]. The right pulmonary artery is more frequently affected, with the left pulmonary branch being 4-8 times more uncommon[1-7]. The association with other cardiac malformations is frequent, being observed in up to 40% of cases[1-3,5,8]. These cardiac malformations include tetralogy of Fallot, persistent ductus arteriosus, right aortic arch (as in our case), ventricular and/or atrial septal defects, and anomalous origin of the right subclavian artery; most being more frequently seen when the left pulmonary artery is involved[2,5,7,8]. Among these malformations, the persistent ductus arteriosus is the most frequent, present in up to 75% of cases[3,5,7].

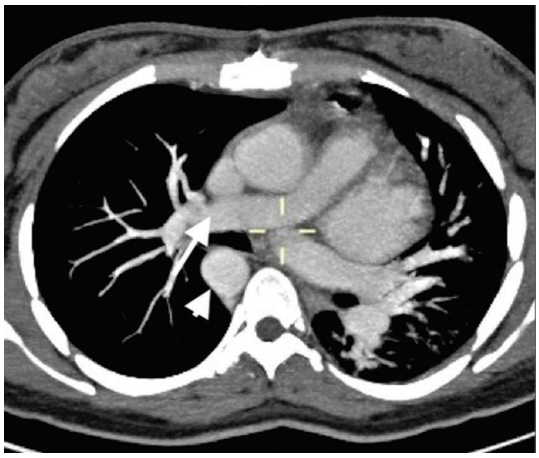

Figure 3. Axial CT Angiography shows normal right pulmonary artery (white arrow) with no left pulmonary arising from the pulmonary trunk and right-sided aorta (Arrowhead).

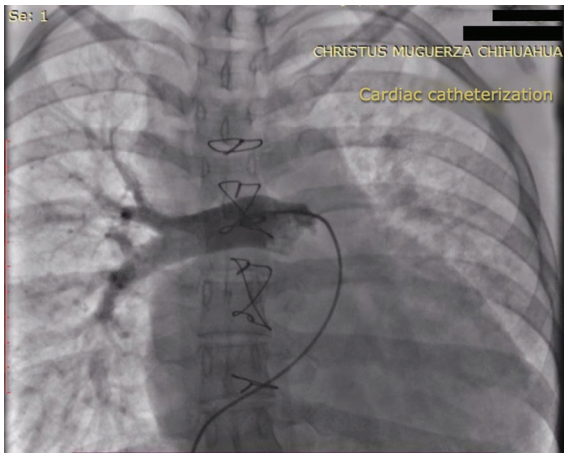

Figure 5. Image taken with flouroscopy during diagnostic cardiac catheterization utilizing contrast, pulmonary trunk catheterization shows the right pulmonary artery as its only branch.

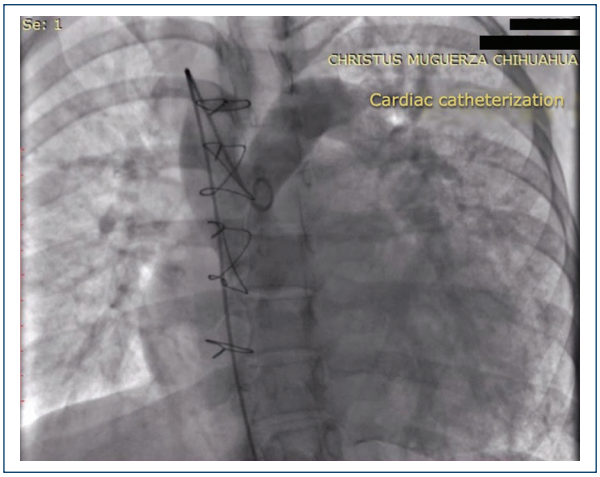

Figure 4. Image taken with flouroscopy during diagnostic cardiac catheterization utilizing contrast dye, shows the left pulmonary artery arising from the ascending aorta.

The embryological basis for the development of this abnormality is not clearly understood or defined; however, it is thought that the truncus arteriosus, the aortopulmonary septum, and the sixth aortic arches are the main structures involved in its development[2,8,9].

During normal fetal development, the aortopulmonary septum divides the truncus arteriosus into two structures, the pulmonary trunk (posteriorly) and the ascending aorta (anteriorly)[2,5,8]. According to the theory proposed by Cucci, malrotation of this septum is what gives rise to ectopia of the pulmonary arteries, where a left malposition gives rise to ectopia of the right pulmonary artery, while a right malposition affects the left branch[2,5,9]. After malposition of the septum, the sixth dorsal arch of the affected side will be integrated within the anterior division of the truncus arteriosus (which will continue its development until it becomes the ascending aorta) giving rise to ectopia of the pulmonary artery over the ascending aorta[7,9].

The clinical picture derives from the perfusion of one lung with systemic output and pressure, whereas the other lung receives all the arterial flow from the right ventricle[2,5,7]. The presentation of the condition can vary from severe and early after birth, to asymptomatic and late, as in those cases where ectopy is present in isolation[1,2,4,8]. Congestive heart failure is the predominant clinical picture, accompanied by other complications such as cyanosis, pulmonary hypertension, progressive respiratory failure, or repeated respiratory infections since birth[1,2,4,5].

Diagnosis is complicated as it warrants a high level of suspicion due to the non-specific clinical picture, usually requiring different imaging studies to confirm and characterize the pathology. The findings in chest radiography are indirect and of poor specificity, being an increase in pulmonary vascularity, cardiomegaly, and right aortic arch those mainly observed[1,5]. Ultrasound is a fast, effective, and non-invasive method that can be confirmatory in some cases; however, due to the complexity for proper visualization of the pulmonary arteries and being operator-dependent, most cases require confirmation by catheterization[1,2]. The best windows are the long parasternal, short parasternal, and subcostal views; special

emphasis is placed on looking for the emergence of the pulmonary artery over the aorta, most commonly originating on the posterior wall[4-6]. Other signs that can be looked for are the absence of bifurcation of the pulmonary trunk as well as the absence of confluence of the pulmonary arteries[4,6]. Catheterization continues to be the method of choice for diagnosis, which also allows characterization of the vascular anatomy and hemodynamic measurement of pulmonary circulation blood pressure[1,4,6]. Another modality that can be used is angio-tomography, which allows complete characterization and pre-surgical assessment of the lesion[1,2].

Early surgical repair is recommended for all patients, as the long-term prognosis is poor if not performed[1-6]. The need to seek early surgical intervention is based on avoiding the development of pulmonary hypertension and progression to obstructive pulmonary vascular disease, both secondary to the presence of systemic arterial pressure on the affected lung[2-4,6,9]. Surgery is recommended as soon as the diagnosis of the disease is made, including the neonatal period, since changes in the pulmonary vasculature can be observed as early as 3 months of life[3-5].

Different surgical techniques for repair have been described, but direct reimplantation of the ectopic pulmonary artery on the pulmonary trunk continues to be the treatment of choice in technically feasible cases[1,3,5,7]. The use of pericardial grafts or patches is also possible as extension techniques for anomalous arteries with insufficient length (commonly when the right pulmonary artery is affected)[1-3,5]. The operative mortality reported in the literature has decreased over time, with an incidence of 0-21% nowadays[2].

The main complication after surgery is stenosis of the implantation site, seen in up to 60-72% of patients and being a significant cause of morbidity[3-5]. Subsequent monitoring is recommended in all cases with either ultrasound, catheterization, or other imaging methods, as some studies reveal its development up to 14 months after surgery[3,4]. The use of interventional catheterization for dilatation of stenosis sites has had encouraging results[2,3]. It is estimated that the rate of reintervention by interventional catheterization for the management of stenosis at the anastomosis site is 2.5-36%[2].

Conclusion

The anomalous origin of a pulmonary artery on the ascending aorta is an extremely rare congenital malformation that comprises only 0.3% of congenital cardiac malformations. The embryological basis for its development is not clearly defined; however, it is thought that the truncus arteriosus, the aortopulmonary septum, and the sixth aortic arches are the structures mainly involved in its development. Congestive heart failure is the predominant clinical picture, accompanied by other complications such as cyanosis, pulmonary hypertension, progressive respiratory failure, or repeated respiratory infections after birth. Diagnosis is complicated, as it merits a high level of suspicion due to the non-specific clinical picture, usually requiring different imaging modalities to confirm and characterize the pathology. Early surgical repair is recommended for all patients. Direct reimplantation of the ectopic pulmonary artery on the pulmonary trunk remains the treatment of choice in technically feasible cases.

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CLINICAL CASES

Tuberculous meningitis and septic shock due to miliary tuberculosis in Down syndrome: a case report

José C. Mendoza-Torres[1], Georgina Aguilar-Gallardo[2], and Rogelio Morán-Ortega[3]*

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Abstract

Introduction: *Tuberculosis is an infectious disease caused by Mycobacterium tuberculosis. Only 1% of tuberculosis cases present as septic shock.* **Case presentation:** *A 41-year-old male with a history of trisomy 21. The condition began with holocranial headache, paresis of the left upper extremity, and paresis of the ipsilateral pelvic limb. As he did not present perfusing blood pressure, management with norepinephrine was initiated. Sampling showed positivity for M. tuberculosis. Antituberculous therapy planned for 12 months was started and dexamethasone was added.* **Conclusion:** *In patients with Down syndrome, it is necessary to intentionally look for extrapulmonary forms.*

Keywords: *Tuberculosis. Tuberculous meningitis. Tuberculosis septic shock. Miliary tuberculosis. Down syndrome.*

Introduction

Tuberculosis is an infectious disease caused by *Mycobacterium tuberculosis*, listed by the World Health Organization in 2020 as the second cause of death from a single infectious agent, after coronavirus disease-19. That same year, a global incidence rate of 127 cases/100,000 inhabitants was estimated[1].

In Mexico, 22,285 new cases of tuberculosis were reported in 2019; the majority are pulmonary (80%) and then meningeal (2.1%)[2].

Extrapulmonary tuberculosis represents 20% of tuberculosis cases, distributed in order of extrapulmonary involvement in lymph nodes, pleura, bones/joints, genitourinary tract, meninges, and peritoneum[3].

The main risk factors for the development of extrapulmonary disease are infection with the human immunodeficiency virus, living with people with tuberculosis, female sex, chronic alcoholism, smoking, diabetes mellitus, drug use, and desnutrition[3,4].

Only 1% of tuberculosis cases present as septic shock due to tuberculosis, which is a rare entity and may or may not be associated with hematogenous dissemination called Landouzy septicemia[5].

Case report

A 41-year-old male with trisomy 21, of low socioeconomic level. The patient began the condition with upper respiratory tract infection, chills, and non-productive cough predominantly at night, weight loss of 10 kg in 3 months, and use of multiple antibiotics without improvement. After this, he presented intense holocranial headache, paresis of the left upper extremity, and paresis of the ipsilateral pelvic limb that made ambulation difficult. He had a saturation of 89% to room air, tachycardia, tachypnea, and hypotension of 80/50 mmHg with a temperature of 38.4°, so it was decided to admit him to the hospital.

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On physical examination, he presented a body mass index of 17.7 kg/m^2, a tendency to drowsiness, pale integuments, dehydrated oral mucosa, bilateral cervical and axillary lymphadenopathy, bilateral infrascapular crackles, abdomen without visceromegaly, hypotrophic extremities with the presence of transverse palmar groove, bilateral sandal sign and 4-s capillary refill.

In the neurological examination, the patient showed a tendency to inattention, neck rigidity, positive Kerning sign, positive Brudzinski sign, positive Binda sign, hypotonia of the left hemibody, decreased muscle strength in the left upper and lower extremity, and response extensor plantar in the left pelvic limb.

Fluid therapy, supplemental oxygen, and antibiotic therapy were initiated; however, as there was no perfusion arterial pressure, norepinephrine was administered, achieving temporary hemodynamic improvement. Given the refractory septic shock, vasopressin and hydrocortisone were added with suspension of the second vasopressor 2 days after improvement.

The chest X-ray showed a diffuse bilateral micronodular pattern (Fig. 1), and the chest tomography detected multiple bilateral micronodules (Fig. 2), as well as images of a budding tree (Fig. 3) and multiple mediastinal lymphadenopathy.

Fiberoptic bronchoscopy sampling for acid-fast bacilli, mycobacterial culture, and GenExpert *M. tuberculosis* (MTB)/rifampicin (RIF) ULTRA demonstrated positivity for *M. tuberculosis*. The HIV enzyme-linked immunoassay test was negative and no other site of infection or co-infection was identified by blood or urine cultures.

The skull tomography showed a hypodense lesion with a digitiform appearance in the right frontoparietal lobe related to vasogenic edema and associated leptomeningeal enhancement (Fig. 4). Magnetic resonance imaging of the skull showed an area of right frontal encephalomalacia and leptomeningeal enhancement (Fig. 5).

Lumbar puncture showed pleocytosis with a predominance of neutrophils, hypoglycorrhachia, and hyperproteinorrhachia. Cerebrospinal fluid culture for mycobacteria and GenExpert MTB/RIF ULTRA was positive for *M. tuberculosis*, with elevated adenosine deaminase.

Tomography of the abdomen and pelvis showed regional lymph node dissemination with evidence of multiple retrocavoaortic, paracaval, and bilateral inguinal lymphadenopathy. Antituberculosis therapy was started with RIF, isoniazid, pyrazinamide, and ethambutol for 12 months and dexamethasone was added. Bone marrow aspirate and urine mycobacterial culture were negative.

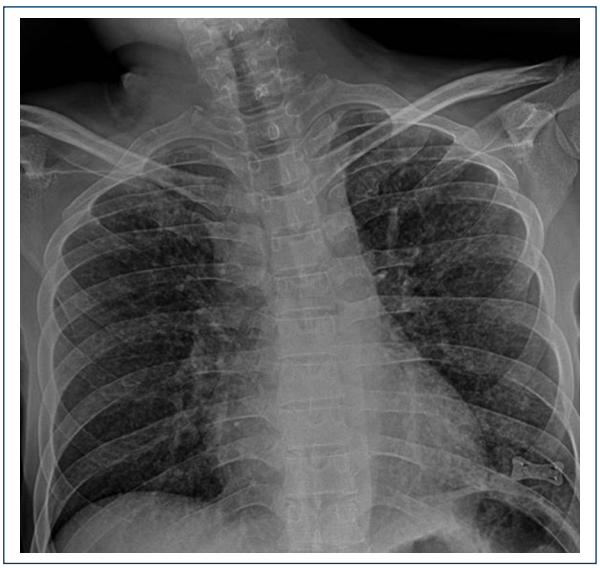

Figure 1. Chest X-ray with diffuse micronodular infiltrate in both hemithorax.

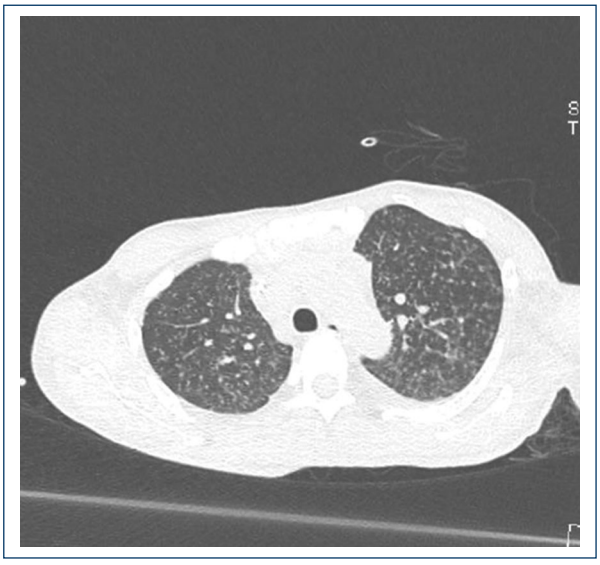

Figure 2. Chest tomography with bilateral micronodular pattern.

The diagnoses of septic shock due to *M. tuberculosis*, miliary tuberculosis, and tuberculous meningitis were integrated. There was remission of septic shock 5 days after starting vasopressor support and fluid therapy, adding neurological improvement 7 days after starting glucocorticoid therapy, and completing a 15-day hospital stay. As part of the follow-up, chest X-rays were

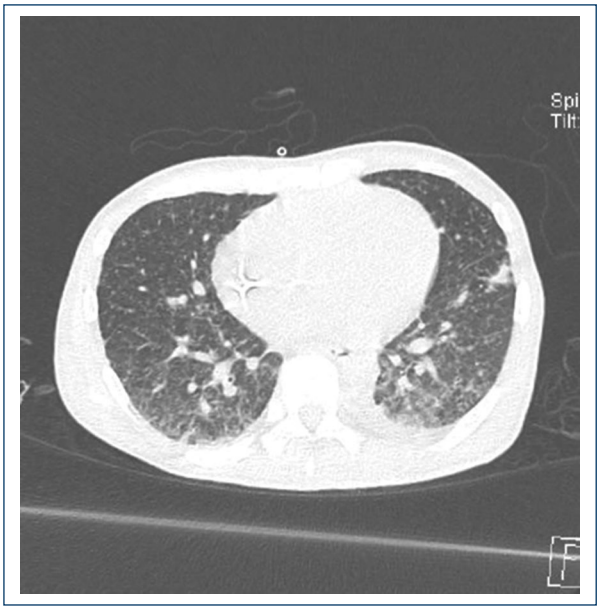

Figure 3. Chest tomography with budding tree images.

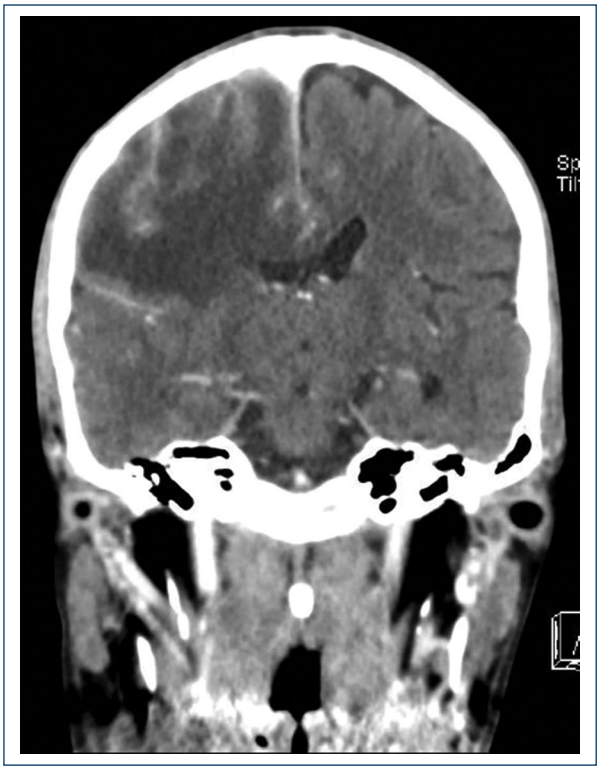

Figure 4. Contrast-enhanced tomography of the skull with hypodense digitiform lesion and right frontoparietal meningeal enhancement.

taken at 4 and 8 months, which showed a bilateral decrease in the micronodular pattern. At present, the patient shows no evidence of neurological sequelae.

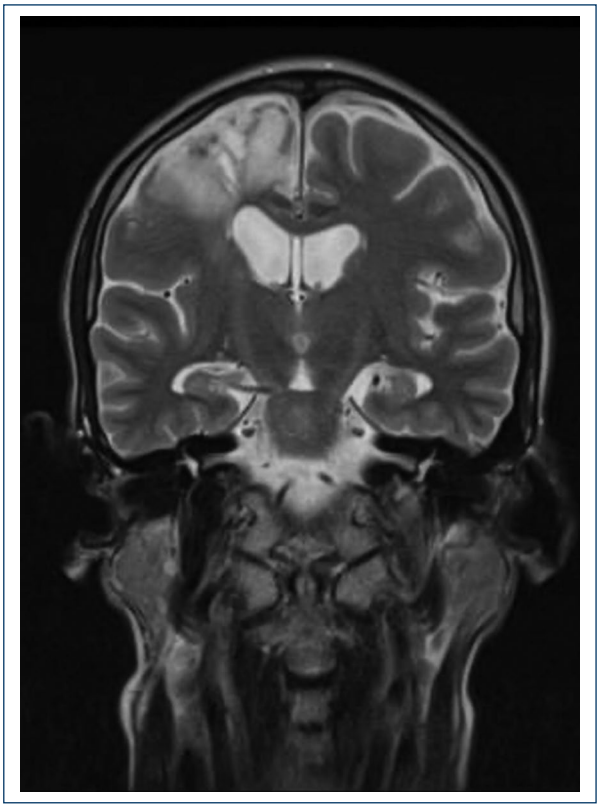

Figure 5. Magnetic resonance imaging shows right frontal leptomeningeal enhancement associated with encephalomalacia.

Discussion

Septic shock occurs in 1% of tuberculosis cases, of which 90% are associated with concomitant lung disease as in our patient, and with mortality of 79%, which improves with the timely administration of anti-tuberculosis treatment[6].

Admission to the intensive care unit for septic shock due to tuberculosis occurs in 12.3% of cases with critical illness due to respiratory failure, hemodynamic instability, or tuberculous meningitis. Vasopressor support and associated lung disease are the main indicators of mortality in the intensive care unit[7]. In turn, the hospital stay in patients with tuberculous meningitis is an average of 15 days, the same length of hospitalization observed[8].

Our patient had a low body mass index (17.7 kg/m^2) that doubles the risk of developing tuberculosis compared to the general population and is associated with higher mortality in patients with septic shock due to *M. tuberculosis*[6,9].

Meningeal infection begins with hematogenous dissemination by *M. tuberculosis* in the subarachnoid

space, forming a caseous lesion called Rich's focus. Later it suffers a rupture, which can cause meningitis and other types of tuberculous neuroinfection[10].

Tuberculous meningitis presents as headache, weight loss, fever, altered level of consciousness, neck stiffness, and only 17% present neurological focal data, consistent with the clinical findings of our patient who presented left hemiparesis[8].

The findings in the cerebrospinal fluid in tuberculous meningitis are hypoglycorrhachia, hyperproteinorrachia, and lymphocytic pleocytosis; however, it is possible to identify a predominance of neutrophils in the early stages of the disease, a situation observed in our patient[11]. Likewise, the determination of adenosine deaminase has a sensitivity and specificity of 79% and 91% for the diagnosis of TM[12]. In tuberculous meningitis, dexamethasone is indicated together with anti-tuberculosis treatment since it reduces mortality by 22%[13].

Down syndrome causes an immunological dysfunction that affects innate immunity and adaptive immunity with decreased cytokine production, low expression of T cell receptors, decreased chemotaxis, alterations in opsonization and phagocytosis, as well as T and natural killer cell dysfunction[14]. Furthermore, the anatomical alterations inherent to the disease and the presence of gastroesophageal reflux, together with immunological dysfunction, cause a higher incidence of respiratory infections regardless of the infectious agent, being predominant in childhood, including tuberculosis[15]. In our patient, multiple risk factors such as malnutrition, low socioeconomic level, and his underlying genetic pathology favored the presence of disseminated tuberculosis.

In the population with Down syndrome, there are only reported cases of neurological involvement due to tuberculosis in pediatric patients. A first case presented due to tuberculous radiculomyelitis manifested with urinary retention and lung involvement[16], and another case presented multiple tuberculomas without lung involvement[17]. There is only one reported case of septic shock due to tuberculosis in a pediatric patient with the presence of active lung disease without central nervous system involvement[18]. Therefore, this is the first reported case of tuberculous meningitis and septic shock due to miliary tuberculosis in an adult population with Down syndrome.

Conclusion

Tuberculosis is an entity that affects global public health. In patients with Down syndrome who have risk factors for tuberculosis, it is necessary to intentionally search for extrapulmonary forms. It is essential to highlight the need to maintain a high suspicion of extrapulmonary disease to expedite the initiation of the diagnostic protocol and positively impact the long-term prognosis.

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CLINICAL CASES

Hepatic inflammatory pseudotumor: a rare entity difficult to diagnose. A case report

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Abstract

Inflammatory pseudotumor (IPT) of the liver, also known as inflammatory myofibroblastic tumor, is a rare benign lesion that mimics malignancy. Its etiology and pathogenesis are unclear, but a relationship has been found with several diseases. Patients usually present with non-specific symptoms. The imaging diagnostic studies of choice are triphasic computed tomography and magnetic resonance imaging. Definitive diagnosis may require histological confirmation. The treatment of choice is still surgical resection in those patients with severe symptoms, an indeterminate diagnosis, or a lack of improvement with medical treatment. We hereby present a case of a patient undergoing laparoscopic resection of an IPT of the liver.

Keywords: *Hepatic inflammatory pseudotumor. Liver. Plasma cell tumor. Hepatic neoplasms. Fever of unknown origin.*

Introduction

Hepatic inflammatory pseudotumor (IPT), also known as inflammatory myofibroblastic tumor, is a rare benign lesion that simulates malignancy clinically, imaging, and sometimes histopathologically[1]. These were first described in the lungs in 1939 and have subsequently been reported in numerous areas including the liver, spleen, kidneys, omentum, ovaries, skin, soft tissues, lymph nodes, spinal cord, salivary glands, breasts, and orbits eyepieces[2-5]. Liver involvement was described for the 1st time in 1953 by Pack and Baker, mentioning that it can cause biliary obstruction, portal hypertension, cirrhosis, and, finally, liver failure[6].

The etiology and pathogenesis of IPT are not clear; a relationship has been found with many pathological entities, such as Crohn's disease, diabetes mellitus, Sjögren's syndrome, gout, chronic ascending cholangitis, primary sclerosing cholangitis, Kostmann's disease, acute myeloblastic leukemia, HIV and autoimmune pancreatitis among others[1,7-9].

IPT is a benign tumor, usually single, located in the right liver, which can be confused with other benign and malignant liver lesions (liver abscesses, liver metastases, hepatocarcinoma or cholangiocarcinoma)[10]. The clinical presentation, laboratory data, and radiological findings are variable, making the differential diagnosis difficult. The definitive diagnosis is made by histopathological study, sometimes requiring immunohistochemistry. The treatment of choice continues to be surgical resection, especially in patients with severe symptoms and an indeterminate diagnosis[5].

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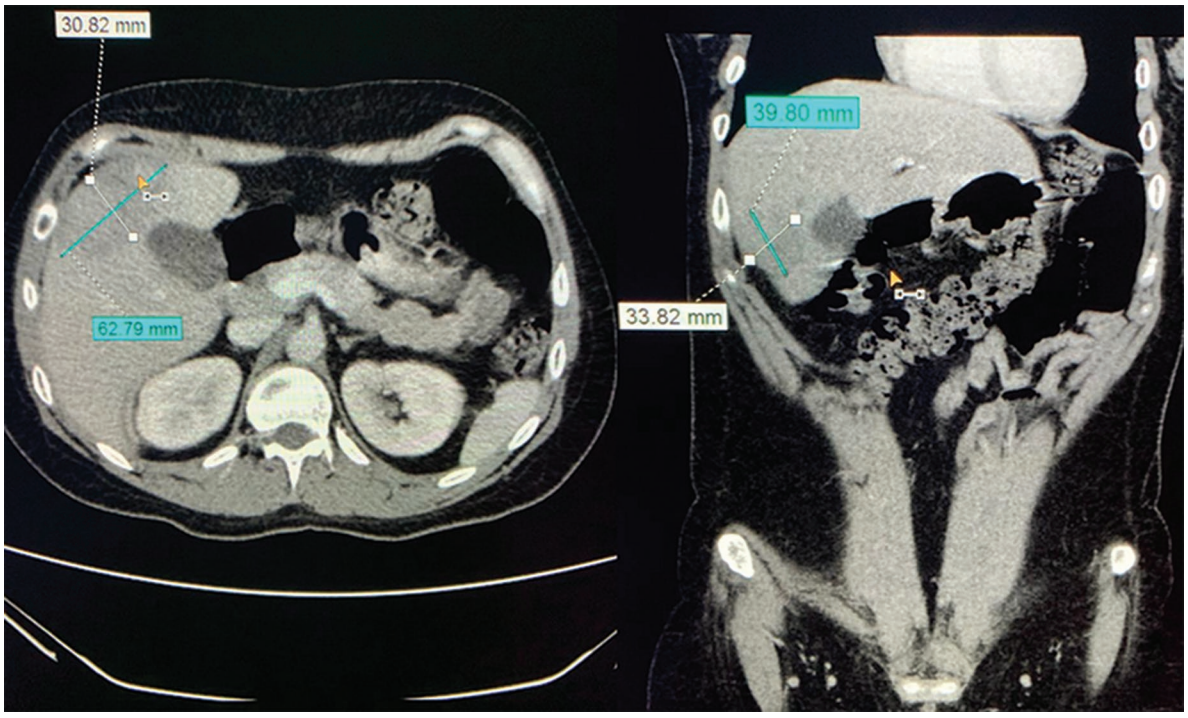

Figure 1. Abdominal computed tomography prior to surgery. There is an oval image, with defined borders, heterogeneous with a hypodense peripheral zone with reinforcement of the contrast medium in late phase and a hyperdense central zone that presents reinforcement after the application of contrast of all phases with measures of 61 x 31 x 54 mm approximately.

Case presentation

A 31-year-old female patient with no chronic, surgical, or any other history of importance for the current condition.

He went to the emergency department due to a 3-week history of unquantified fever managed with paracetamol. Since the patient did not show improvement, she went to her health center where they indicated symptomatic management for the fever as well as unspecified antibiotics. The ultrasound reports an ovoid, heterogeneous image, with an anechoic area and discrete posterior acoustic reinforcement, which is why she was sent for evaluation by a specialist doctor.

Upon questioning, the patient reported intermittent abdominal pain of approximately 1 week's duration in the right hypochondrium (visual analogue scale 7/10) without irradiation, without improvement in the intake of analgesics, without weight loss or any other accompanying symptoms. On physical examination, hemodynamically stable, without jaundice with adequate hydration status, cardiopulmonary without compromise, slightly ballooning abdomen associated with adipose panniculus, peristalsis present, percussion without alterations, palpation with mild pain in the right hypochondrium, negative Murphy's sign, unable to identify masses or megalias, without signs of peritoneal irritation, extremities without alterations. The studies report: leukocytes 12.9 × 10^3 mm^3, neutrophils 8.8%, hemoglobin 10.4 g/dL, hematocrit 31%, platelets 729 10^3 mm^3, glucose 78 mg/dL, urea 12.6 mg/dL, creatinine 0.4 mg/dL, uric acid 3.7 mg/dL, sodium 140 mmol/L, chloride 107 mmol/L, potassium 3.2 mmol/L, cholesterol 138 mg/dL, proteins 7.5 g/dL, albumin 3.4 g/dL, total bilirubin 0.3 mg/dL, direct bilirubin 0.2 mg/dL, indirect bilirubin 0.1 mg/dL, aspartate aminotransferase 15 IU/L, alanine aminotransferase 25 IU/L, alkaline phosphatase 162 IU/L, lactic dehydrogenase 122 IU/L and viral panel for hepatitis negative. The contrast-enhanced abdominal tomography shows a liver of preserved size, in segment V with an oval image, with defined edges, heterogeneous with a hypodense peripheral area (30 HU) with contrast medium enhancement in the late phase and a hyperdense central area that presents enhancement. After contrast application in all phases with measurements of 61 × 31 × 54 mm and volume of 44 cc, in relation to liver abscess (Fig. 1). Tumor markers show: CA 19-9 11.8 U/mL (0.80-25 U/mL), Alpha feto protein 3.0 ng/mL (0-9 ng/mL), CA125 24.20 U/mL (0-35 U/mL). It was decided to request

image-guided percutaneous drainage, but the study could not be performed.

Due to the persistence of abdominal pain and the tomography data, a diagnostic laparoscopy was performed, finding a liver tumor in segments V and VI measuring approximately 4 cm in diameter. It was decided to perform resection with the help of an ultrasonic scalpel, leaving a margin. Free of 5 mm throughout the periphery of the lesion, in the end a piece measuring 4 cm in diameter × 1.5 cm wide was obtained, in the rest of the laparoscopy no more apparent alterations were found (Figs. 2 and 3).

In the post-surgical period, the patient presents remission in pain and fever, drainage output and antibiotic management are monitored, and a pathology report is collected which mentions IPT. Two days after the intervention, the patient no longer mentions pain or fever, tolerates the oral route adequately, and shows no evidence of an inflammatory response, so she is discharged to continue follow-up in the outpatient clinic.

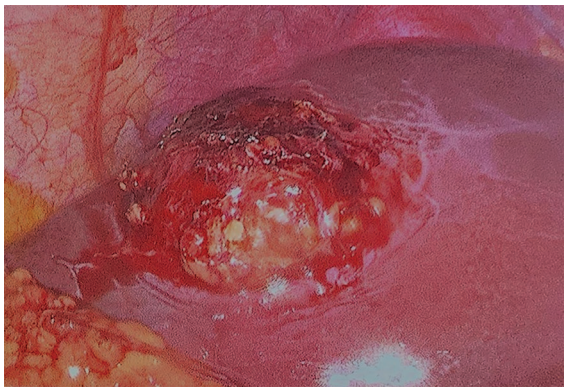

Figure 2. Image of the tumor during laparoscopic resection, it presented adhesions to the wall and omentum, which are already released in the image.

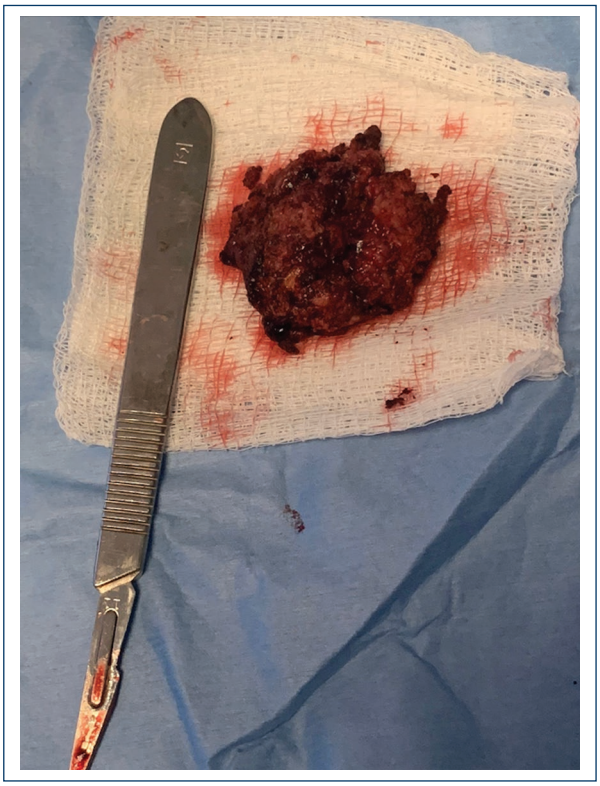

Figure 3. Tumor already resected and removed from the patient for pathological examination.

Discussion

Hepatic IPT is considered a condition of benign reactive inflammation with unclear pathogenesis[11]. Since pack and baker first described it in 1953[12], hepatic IPT has become increasingly recognized, probably due to advances in imaging methods. Histologically, the lesion is composed of proliferation of fibrous tissue with the presence of numerous inflammatory cells (plasma cells, lymphocytes, neutrophils, macrophages, multinucleated giant cells, and eosinophils)[11].

We present the case of a patient in whom the diagnosis was made after a laparoscopic resection. It is mentioned that the incidence of IPT is low, and that the clinical presentation is very variable, even being an incidental diagnosis in some cases as it is asymptomatic[13]. In this case, the patient comes for evaluation due to fever of 3 weeks' duration as well as abdominal pain of 5 days and an external ultrasound with the characteristics already mentioned. Since we are in an endemic area, a study protocol for liver abscess is initiated, initially of origin to be determined. However, since clinical or laboratory studies do not correspond to the diagnosis of liver abscess and there is no improvement after antibiotic treatment, it was decided to request a percutaneous puncture, which cannot be performed.

Laboratory studies in IPT can be within normal parameters or non-specific alterations of the liver profile and sometimes present elevation of tumor markers such as CAE and CA 19-9[1,2,14]. The radiological results show a late enhancement in the computed tomography with contrast, specifically in the periphery of the lesions, it is considered to be characteristic of IPT, but it can also be compatible with cholangiocarcinoma or metastatic lesions[14] so the imaging diagnosis ends up being nonspecific.

In our case, despite antibiotic treatment, no remission of symptoms was achieved, in addition to the fact that the control tomographic study showed slight growth of the lesion. For this reason, it was decided to perform diagnostic laparoscopy without identifying abscesses, performing complete exeresis for histopathological analysis. In the literature they are reported macroscopically as non-encapsulated and nodular whitish-yellowish lesions, agreeing with the report of our case[10].

Surgical resection, which was initially considered the management of choice, should be reserved for some situations such as persistence of unresolved systemic symptoms despite medical treatment, evidence of growth, involvement of the hepatic hilum, biliary obstruction, and/or hypertension. Portal or inability to rule out malignancy[14]. In patients who were diagnosed by biopsy alone, liver resection may also be necessary as definitive treatment when steroid therapy is futile[15].

Conclusion

IPT is a rare entity whose etiopathogenesis is still being studied. Given such a non-specific clinical presentation, the diagnosis is generally made by exclusion and may be incidental in asymptomatic patients.

Undoubtedly, current imaging studies help us direct the diagnosis, but since our environment does not have high-cost studies, it is necessary to consider laparoscopy as a diagnostic/therapeutic tool in cases in which there is suspicion of neoplasia or that fulfills with any of the criteria already mentioned to perform some type of resection, thus avoiding highly invasive treatment.

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CLINICAL CASES

Microsurgical reconstruction of the SCALP with fasciocutaneous perforator free flaps: presentation of four cases

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Abstract

Introduction: *Scalp is essential for the protection of intracranial contents and contributes to external aesthetic appearance; it is a unique hair-bearing surface. Total or partial lack of scalp directly affects patient's quality of life.* **Case presentation:** *The objective of this paper is to present four cases of complex scalp wounds solved with fasciocutaneous free flaps. Microvascular free tissue transfer is associated with high success rates in scalp reconstruction (90% success rate). Microsurgery was the last step on the reconstructive ladder, nowadays with reconstructive elevator concept; microsurgery can be used as the first step in reconstruction.* **Conclusion:** *Microvascular scalp reconstruction is an invaluable part of a reconstructive surgeon since the advantages outweigh the disadvantages.*

Keywords: *Scalp. Microsurgery. Fasciocutaneous free flap. Perforator free flap. Anterelateral thigh free flap. Medial sural arterial perforator flap.*

Introduction

The scalp is the soft tissue covering the bony calvarium. It is essential for the protection of intracranial contents and contributes to the external aesthetic appearance, in addition, it is a unique hair-bearing surface of the body[1,2]. It consists of the skin, sub-cutaneous tissue, galea aponeurosis, loose connective tissue, and pericranium layers (mnemotechnic SCALP)[1]. The scalp includes an inelastic galea aponeurosis layer and has limited adjacent tissue for the above[2]. Damage to this structure such as from trauma, burns, radiation, or tumor excision can result in poor quality of life. The complex anatomy of the scalp makes reconstruction difficult[1,3,4].

Since the description of the free omental flap for the reconstruction of large scalp defects by McLean in 1972[5] advances in knowledge of vascular anatomy and improvement in microsurgery techniques have allowed the development of better reconstructive options for this type of defect with minimal donor site morbidity.

The ideal donor tissue should be of appropriate thickness and surface area to cover convexity of the calvarium with a robust blood supply[4]. Due to the characteristics, the latissimus dorsi (LD) free flap was the primary choice for microsurgical scalp reconstruction for many years. Nowadays, fasciocutaneos flaps such as anterolateral thigh (ALT) flap and medial sural artery perforator (MSAP) flap add up as good reconstructive options[6].

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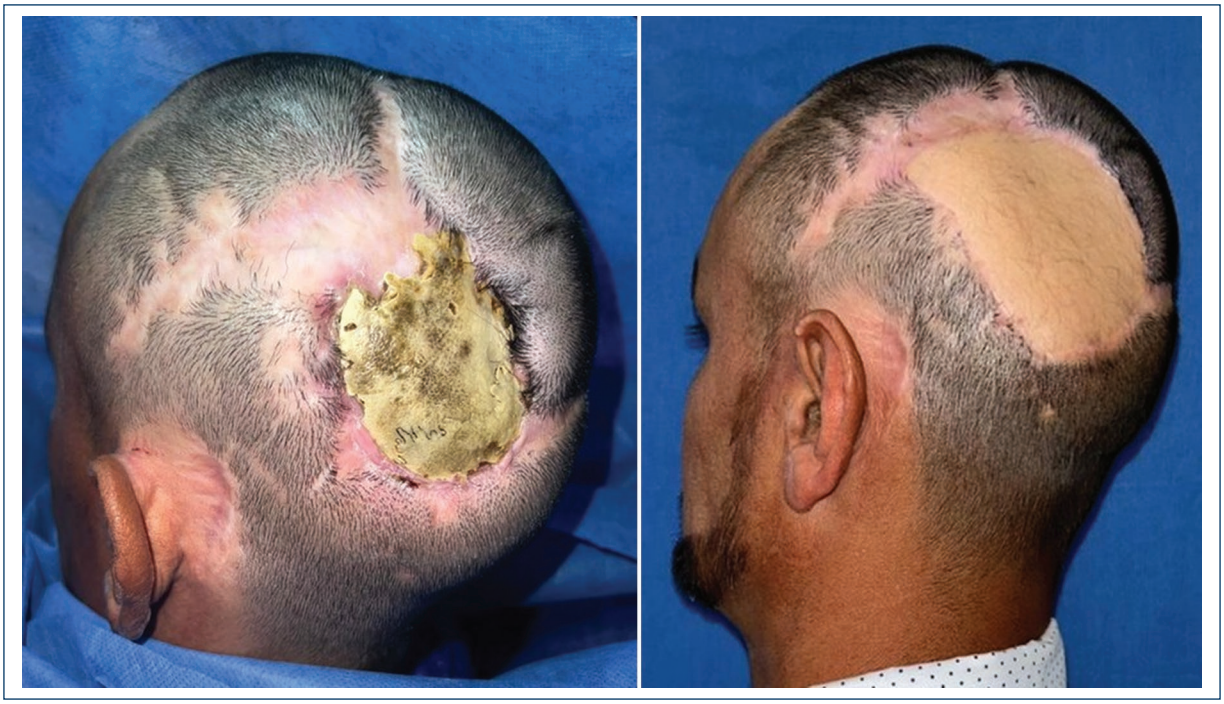

Figure 1. Case 1: pre-surgical photography and 8 moths after anterolateral thigh free flap surgery.

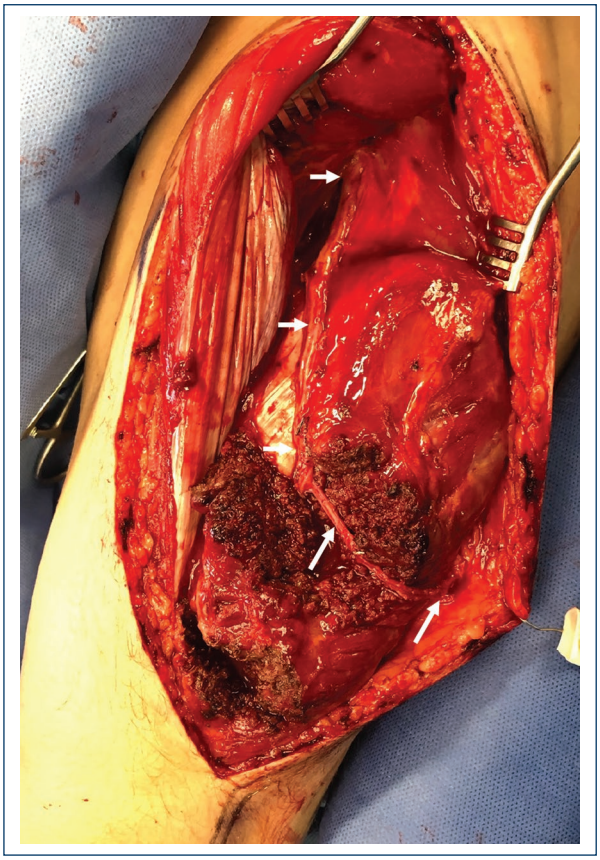

Figure 2. Skin perforator vessel increased in caliber until emerged from the lateral femoral circumflex artery at intermuscular septum (arrows).

An advanced understanding of the anatomy of the scalp, as well as an awareness of the full complement of reconstructive options, is essential to providing the patient the best result.

The objective of this paper is to expose four cases of reconstruction of scalp defects using fasciocutaneous perforator-free flaps.

Cases presentation

Case 1 (Fig. 1)

A 44-year-old man who developed skull bone exposure after an electrical burn. He had two previous failed reconstruction attempts (retroauricular total thickness skin graft and O-S regional flap). After these procedures, he evolved with bone exposure of 6 × 5 cm in the left parieto-occipital surface and abundant scar tissue around. In conjunction with neurosurgery, we removed devitalized bone and scar tissue and we performed skin coverage with ALT-free flap.

Technique of flap harvest

The patient is placed in the supine position and the thigh is circumferentially prepared. A line is drawn extending from the anterior superior iliac spine to the lateral superior aspect of the patella. At the midpoint of this line

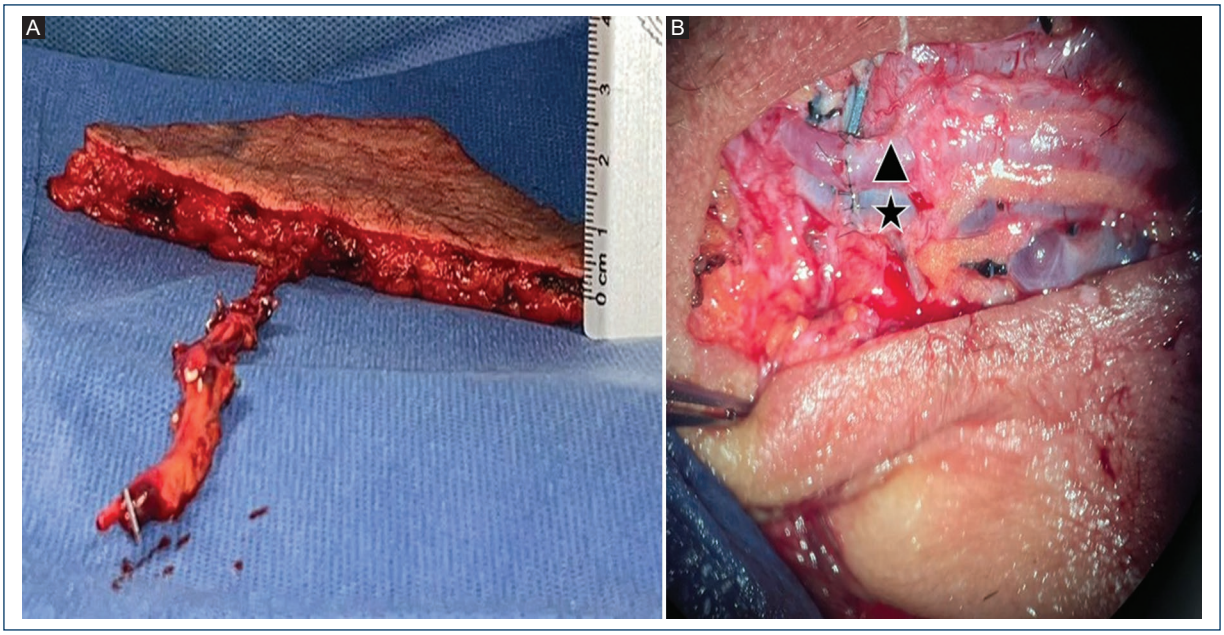

Figure 3. A: superthin ALT flap (8 mm). **B:** microvascular anastomosis under magnification, artery (triangle) and vein (star).

a circle with a radius of 3 cm is drawn, delineating the predominant location of the skin perforations vessels using handheld Doppler, our preference is to include more than one perforator, but it is not always possible. Skin island of the flap (7.5 × 8.5 cm) was designed on the left thigh. The medial margin of the flap incision is first made through the skin and subcutaneous tissue, dissection proceeds above the fascia in a lateral way until the previously Doppler skin perforator vessels are reached. A fascial incision is made in the direction of the skin perforator vessel and the vessel is traced in a retrograde fashion until adequate vessel length and caliber are achieved until the perforator vessel emerges from the lateral femoral circumflex artery at intermuscular septum between the rectus femoris and vastus lateralis muscles (Fig. 2). Lateral incision of the flap is made a dissection is made until medial dissection obtaining a superthin flap (Fig. 3A) One intramuscular perforator vessel was found during the dissection. We performed end-to-end anastomosis to the left superficial temporal vessels (one vein and one artery) under magnification (Fig. 3B) the patient evolved favorably, without complications. He was discharged 5 days after surgery.

Case 2 (Fig. 4)

A 46-year-old man who had biparietal contusion. He evolved with skin necrosis and subsequently bone exposure (7.5 × 9 cm) and osteomyelitis. First neurosurgery performed daily debridement of devitalized tissue. Once the wound was completely clean, we performed a MSAP-free flap.

Technique of flap harvest

The patient is placed in the supine position and the leg is circumferentially prepared. A line is marked from the midpoint of the popliteal crease to the central point of the medial malleolus. A handheld Doppler is used to find the perforators, which are located mostly over this line, usually at 8 cm from the popliteal crease. The island of the flap (7.5 × 9 cm) was designed on the right leg, lateral incision is performed to the until de fascia, dissection continues medially until the perforator vessel is located. A fascial incision is made in the direction of the skin perforator vessel and the vessel is traced in a retrograde fashion intramuscularly until the perforator length and diameter are adequate to suit the defect (Fig. 5). We performed end-to-end anastomosis to left superficial temporal vessels (one vein and one artery) under magnification. The patient evolved favorably. He was discharged 5 days after surgery without complications.

Case 3 (Fig. 6)

A 59-year-old man in 2011 was diagnosed with meningioma, neurosurgery performed craniectomy, tumor

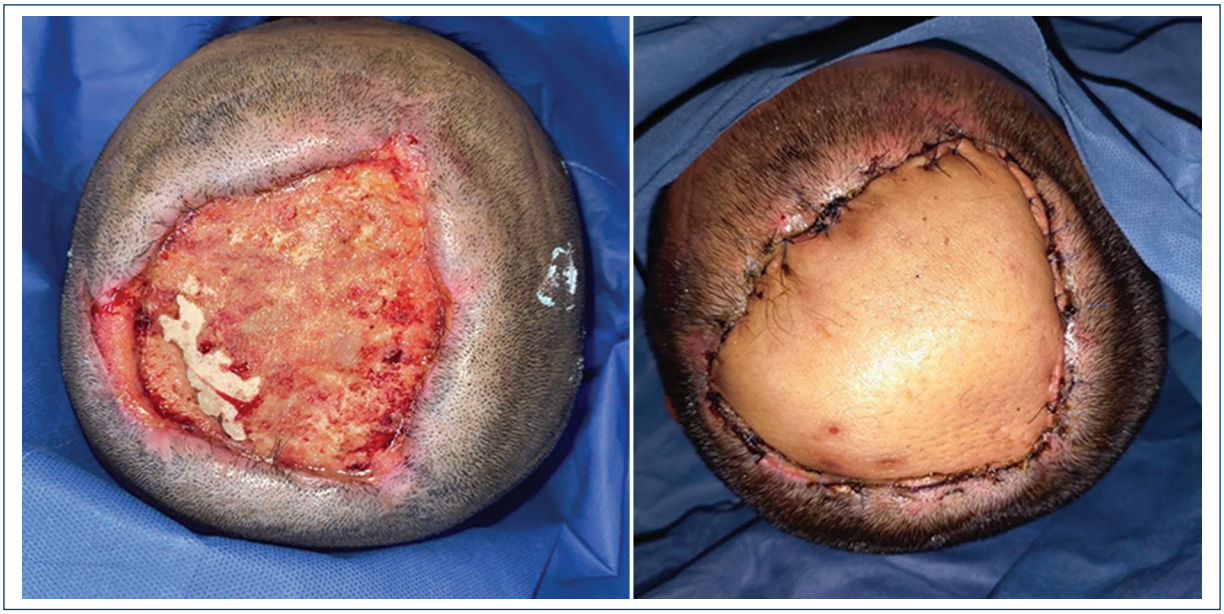

Figure 4. Case 2: coronal view before and after medial sural artery perforator free flap reconstruction.

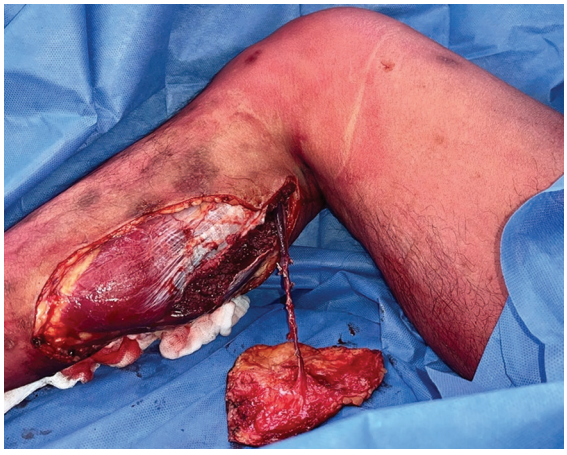

Figure 5. Medial sural artery perforator flap dissection before separating the flap from its native feeding vessel.

resection, and cranioplasty with titanium mesh placement. Ten years later, he developed titanium mesh and bone exposure through a chronic wound that progressively increased in size on the left frontal and parietotemporal surface. In our first evaluation, the lack of skin coverage was 15 × 7 cm. During surgery, titanium mesh was removed and bone exposed with osteomyelitis was resected by neurosurgery team, meanwhile we performed a left ALT free flap. Surgical technique was performed as described above. The skin island (20 × 10 cm) was designed on left thigh, dissection was performed suprafascially to obtain a superthin flap. We performed end-to-end anastomosis to left superficial temporal vessels (two veins and one artery) under magnification. The patient evolved favorably. He was discharged 5 days after surgery without complications.

Case 4 (Fig. 7)

A 30-year-old man with a history of severe head trauma with right temporal bone fracture treated with decompressive craniectomy. After a year, he underwent cranioplasty with methyl methacrylate plate under the right temporal bone surface. Four years later, he developed plate exposure through a chronic wound that progressively increased in size (3 × 3 cm). During surgery, methyl methacrylate plate was removed by neurosurgery team, meanwhile we performed a right MSAP free flap, surgical technique was performed as described above. The island of the flap (4 × 3 cm) was designed on the right leg. We performed end-to-end anastomosis to the right facial vessels (one veins and artery) under magnification. The patient evolved favorably. He was discharged 5 days after surgery without complications.

Discussion

Free tissue transfers in scalp reconstruction are reserved for medium (10-50 cm^2), or large (> 50 cm^2) defects[3,7], previously radiated patients, exposed cranial

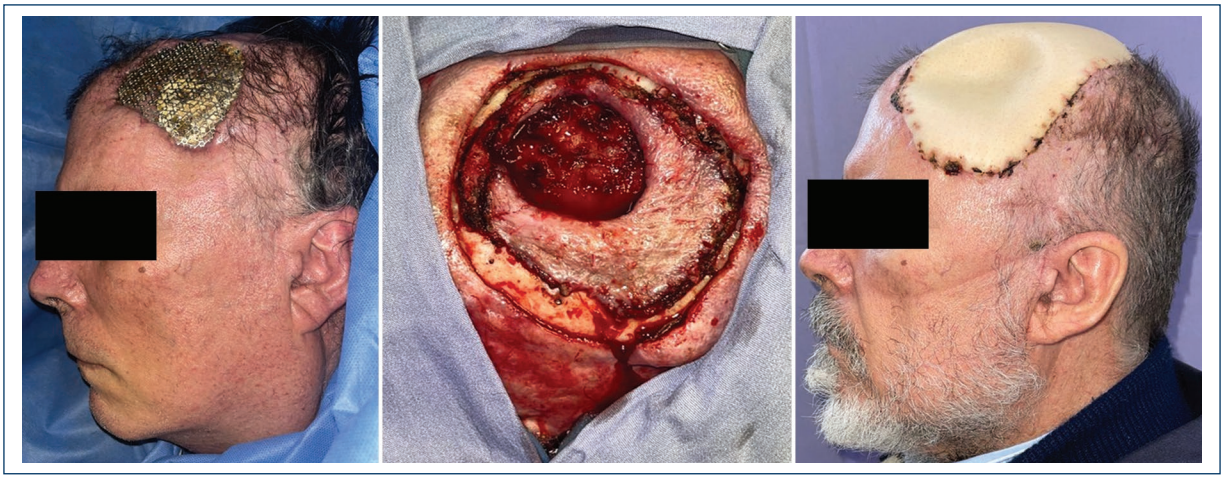

Figure 6. Case 3: before, trans-surgical, and 2 months after microsurgical reconstruction with left ALT free flap.

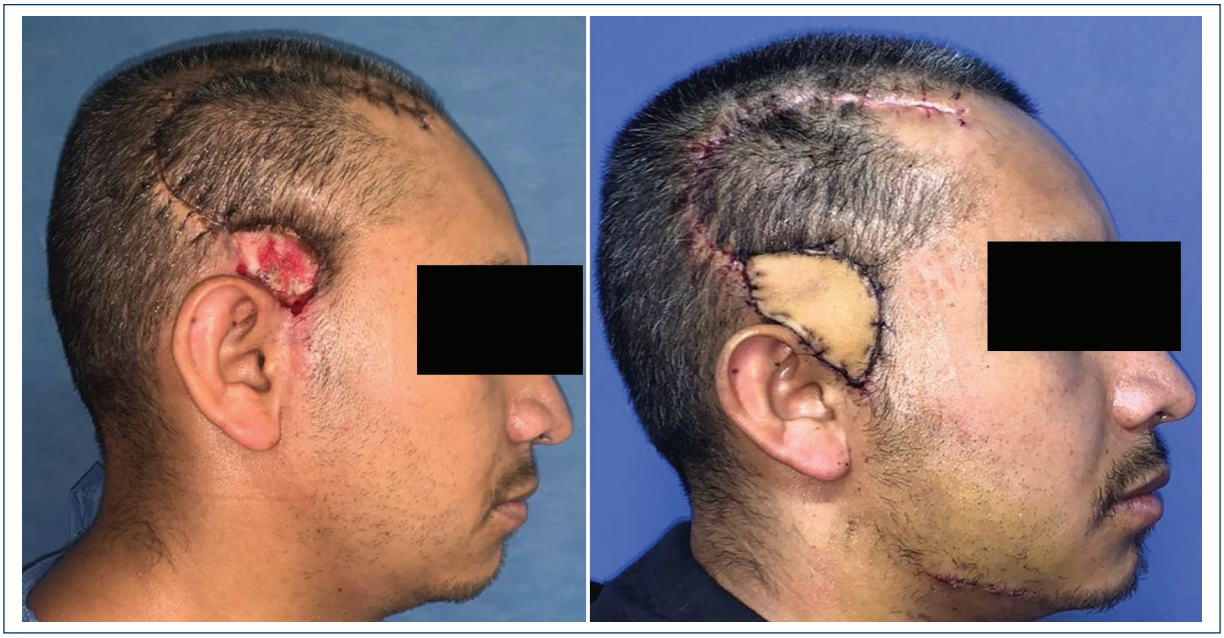

Figure 7. Case 4: before and 1 month after microsurgical reconstruction with right medial sural artery perforator-free flap.

contents, or chronic infection[7]. Advantages of free tissue transfer include bulk of vascularized tissue. Disadvantages are alopecia and color and contour mismatch[1,7].

Microvascular free tissue transfer is associated with high success rates in scalp reconstruction over 90% success rate[1,4].

In our initial approach to scalp reconstruction, using the reconstructive elevator concept described by Gottlieb and Krieger in 1994[8], our preference is to use microsurgery-free flaps in large and secondary defects. We agree with Song and Pu[4] a favor to fasciocutaneous free flap (ALT or MSAP) because the obvious benefits compared with the LD free flap (avoidance of muscle sacrifice, minimal donor site morbidity, and thin flaps). Both ALT and MSAP flaps can be comfortably done in the supine position without the need to position the patient in a lateral decubitus position during the flap dissection or total repositioning of the patient after flap dissection. Fasciocutaneous flaps can be lifted in different planes depending on the desired thickness as

described by Kwon et al.[6] muscle flaps lack this advantage. However, in scalp defects > 90% of the surface, we prefer a LD-free flap rather than two fasciocutaneous-free flaps.

Jang and Choi[3] published a total of 98 cases of scalp reconstruction, 3% of scalp defects were reconstructed with free flaps mostly for large defects and cranial bone or dura mater exposure. As a major complication, they presented two cases of infection without specifying if they occurred in the group of free flaps (two LD muscle flaps and one ALT flap), in the cases presented we had no infections. Of the others, major complications reported by Jang and Choi (flap congestion, hematoma, skin necrosis, and dehiscence)[3] we did not present any of the above.

Park et al.[2] published a total of 180 cases of scalp reconstruction. From those 36 cases were free flaps. As a complication in those patients who underwent free flap reconstruction, they reported four cases of partial flap loss and three cases of seroma or hematoma. We did not present any cases of partial flap loss, seroma, or hematoma.

In muscle-based flaps are often initially bulky, however, flap atrophy over time yields an acceptable cosmetic result with tissue thickness comparable to adjacent scalp tissue; however, fasciocutaneous-free flaps could be rise in suprafascial plane even if the flap is too thick some secondary procedures could be performed to become very thin flap[6].

Conclusion

As a conclusion, scalp reconstruction is challenging for the plastic surgeon and requires an excellent understanding of the surgical anatomy for optimal reconstructive outcomes. Microvascular reconstruction is an invaluable part of a reconstructive surgeon since the advantages outweigh the disadvantages. With reconstructive elevator concept, microsurgery can be used as the first step in reconstruction when the expected result is achieving functional and aesthetic outcome with it. In our cases, fasciocutaneous-free flaps were the first and only option for scalp reconstruction.

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Conflicts of interest

The authors declare no conflicts of interest.

Ethical disclosures

Protection of human and animal subjects. The authors declare that no experiments were performed on humans or animals for this study.

Confidentiality of data. The authors declare that they have followed the protocols of their work center on the publication of patient data.

Right to privacy and informed consent. The authors have obtained the written informed consent of the patients or subjects mentioned in the article. The corresponding author is in possession of this document.

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CLINICAL CASES

Incidental finding of choledochal cyst in a pregnant woman: case report

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Abstract

Introduction: *Choledochal cysts are a rare congenital anomaly of non-defined etiology, commonly diagnosed in childhood, whose main manifestations are abdominal pain, jaundice, and a palpable mass. Although magnetic resonance cholangiography is the gold standard for diagnosis, incidental findings have been increasing due to the extensive use of imaging studies.* **Case presentation:** *A 25-year-old woman with an incidental finding of a choledochal cyst (Todani Ia) by prenatal control ultrasound, being managed with resection of the cyst and hepatojejunal Roux-Y anastomosis, leaving a remnant that warrants surveillance by the risk of malignancy.* **Conclusion:** *The relevance of this type of cases is knowing about their existence, having a high index of suspicion and being able to carry out a complete diagnostic approach for adequate surgical planning.*

Keywords: *Choledochal cyst. Pregnancy. Todani. Case report.*

Introduction

Choledochal cysts are a rare congenital anomaly in the Western population; they are characterized by one or multiple dilations of the intra and extrahepatic bile duct. Diagnosis commonly occurs during childhood with the presence of jaundice, palpable mass, and abdominal pain. The most accepted classification was proposed by Todani in 1977, divided into five types depending on their anatomical location. Type 1 cysts are the most common, representing up to 80% of cases, and are characterized by fusiform dilation[1]. The importance of the diagnosis lies in the high rate of complications, the cholangiocarcinoma being the most serious, with an incidence between 1000 and 2000 times higher compared to the healthy population[2].

Case report

The patient is a 25-year-old woman with no significant history who began her condition in 2016 during her first pregnancy with colicky epigastric pain of moderate intensity radiating to the right subscapular region, which is exacerbated by the ingestion of cholecystokinetics accompanied by nausea and vomiting of gastro-alimentary content without choluria, acholia, or jaundice. Conservative treatment was initiated based on analgesics and antispasmodics as well as dietary modifications due to clinical suspicion of watery cholecystitis. She carried her pregnancy to term without complications, with resolution by elective cesarean section. It continues the following years asymptomatically. In July 2020, she came to our unit for the 1st time during her second pregnancy

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due to a recurrence of painful symptoms with characteristics identical to those that occurred in the first pregnancy. Laboratory studies (blood biometry, blood chemistry, liver function tests, amylase, and lipase) show normal parameters. The prenatal control ultrasound reveals a pregnancy of 16 weeks of gestation by fetometry, a live singleton with normal ultrasound characteristics, a heart rate of 156 beats/min, a Grade I anterior body insertion placenta, and a probable delivery date of December 23, 2020. Ultrasound of the liver and bile duct detects an inflamed gallbladder with signs of cholesterolosis and gallbladder polyps, a common bile duct cyst that extends to the head of the pancreas with stones inside. Due to these findings and limited studies free of ionizing radiation, cholangioresonance was performed, which shows a dilation of the saccular appearance dependent on the common bile duct of 70 × 37 × 42 mm with stones inside that cause anterior displacement of the pancreatic head. It is reported as a Todani 1 choledochal cyst (Fig. 1).

Given these findings, the patient was scheduled for complete resection of the cyst, cholecystectomy, and bilioenteric reconstruction in Roux-en-Y after resolution of the pregnancy, being managed with hygienic-dietary measures as well as analgesics for necessary reasons. Normal evolutionary pregnancy continues until resolution by cesarean section in December 2020, obtaining a single live product without complications. I was re-admitted a month later for surgery.

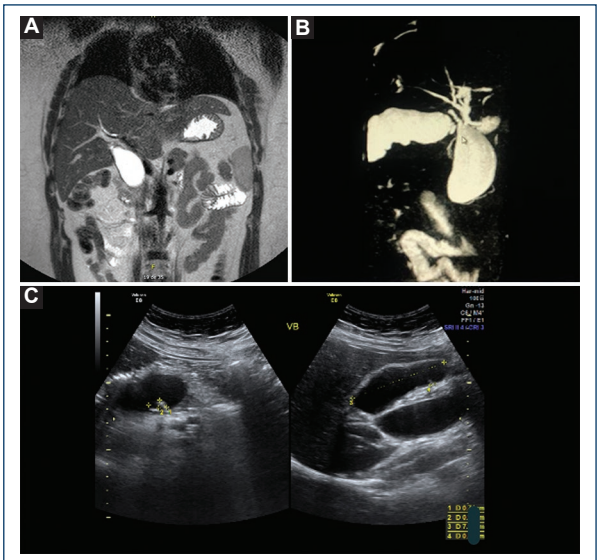

Figure 1. A: cholangioresonance where the choledochal cyst is visualized. **B:** cholangioresonance where the gallbladder and Todani 1a cyst are visualized. **C:** ultrasound shows cyst with stones inside.

Surgical findings

An 8-cm, Todani 1a-type choledochal cyst was detected with firm adhesions to the first portion of the duodenum and extension toward the head of the pancreas. For this reason, resection of the cyst, cholecystectomy, and Roux-en-Y hepatojejunal anastomosis were performed. Due to the involvement of the head of the pancreas, a complete resection of the cyst was not possible, completing the surgery without incident. The patient progressed favorably and was discharged on the eighth post-operative day, continuing monthly monitoring by outpatient general surgery consultation. The only complication was the formation of a seroma (Fig. 2).

The pathology report reported the presence of chronic cholecystitis, a preserved area with partial epithelial lining without atypia, and the presence of subepithelial histiocytes (cholesterolosis), as well as a cystic lesion made up of the simple epithelial lining of cuboidal cells without atypia (Fig. 3).

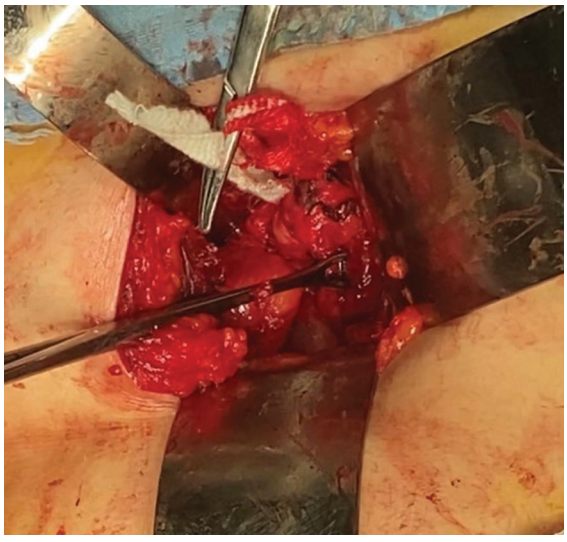

Figure 2. Cyst between Babcock tweezers.

Discussion

Choledochal cysts have an incidence of 1:100,000-150,000 with a pre-dominance in women (4:1). They are usually diagnosed during childhood, even prenatally; however, up to 25% of cases are detected in adults[1]. Although the etiopathogenesis is still unclear, the most accepted theory is that of Babbitt et al. This theory establishes an anomalous union of the pancreatobiliary

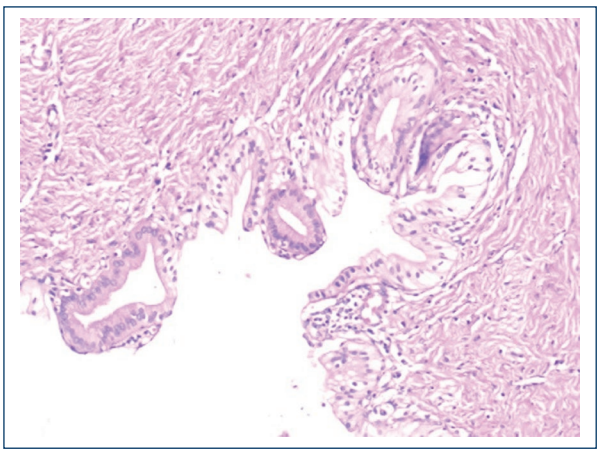

Figure 3. Histopathological report of cystic lesion with cuboidal cells without atypia.

duct, which is abnormally long and, added to the dysfunction of the sphincter of Oddi, allows the reflux of pancreatic contents, generating dilation of the common duct. This theory loses strength since the anomalous union occurs in between 50% and 80% of cases of choledochal cysts. Kusunoki made a theory of pure congenital origin, relating a decrease in the number of ganglion cells present in the common bile duct, this produces proximal dilation, similar to conditions such as achalasia or Hirschsprung's disease[1-3]. Recently, chromosomal anomalies have been detected that could play a role in etiopathogenesis, specifically the duplication of chromosome 17q12, which is associated with type Ia choledochal cysts. This chromosome encodes the HNF1B gene (Hepatic Nuclear Factor 1B), which is involved in the formation of the bile duct and whose alteration is related to jaundice and morphological anomalies in the gallbladder[2].

The first classification was proposed in 1959 by Alonso-Lej, which describes extrahepatic cysts (I-III); in 1977, Todoni included the intrahepatic forms (IV and V). Type I cysts (50-80%) are fusiform or cystic dilations of the common bile duct, which in turn are subdivided into three according to their morphology. Type II (2%) are true diverticula. Type III (1.4-4.5%) is choledococele. Type IV (15-35%) are multiple intra and/or extrahepatic dilations and are subdivided into Type IVa and IVb. Type V (20%), known as Caroli's disease, is the fusiform or saccular dilation of the intrahepatic bile duct. When associated with congenital liver fibrosis, it is called Caroli's syndrome[1,4-6] (Fig. 4). Despite remaining the most accepted classification, modifications to it have recently been suggested. In 2004, Visser established that the Todani classification has no clinical use and can generate confusion by not taking into account the differences between the pathogenesis, prognosis, and treatment of each cyst. It groups four completely different conditions in an alphanumeric way; instead, it suggests a classification. descriptive: common bile duct cysts, common bile duct diverticulum, choledochocele, and Caroli disease (Fig. 5)[7] Type I and IV cysts are variations of the same condition, since the majority of Type 1 cysts have intrahepatic dilations; likewise, their etiology is secondary to an anomalous union of the pancreato-biliary junction, thus allowing the reflux of pancreatic enzymes. These have the highest risk of malignancy. Type II are secondary to common bile duct duplication, and their malignant conversion is extremely rare. Type III comes from obstruction of the ampulla of Vater and dysfunction of the sphincter of Oddi (Table 1)[7-9].

Eighty percent of cases are detected during childhood, with prenatal detection being possible; the classic triad of jaundice, pain, and abdominal mass occurs in only 20% of cases. Eighty-five percent of infants present two of these symptoms. In adults, manifestations are generally secondary to complications such as abdominal pain (61%), pancreatitis and jaundice (18.8%), and malignancy (7.5%). Chronic inflammation can produce distal stricture, leading to biliary stasis, stone formation, infections, cholangitis, fever, obstructive jaundice, and even secondary biliary cirrhosis[1,5].

The most serious complication of choledochal cyst is cholangiocarcinoma (2-17%), reaching as high as 30% in some case series. The risk is mainly associated with age and type of cyst, being more common in patients over 20 years of age and in those with type I and IV cysts. The global incidence of malignant transformation is 7.5%, which represents a risk between 1000 and 2000 times greater than the general population[1,2].

The malignancy originates in the extrahepatic bile duct in 50-62% of cases, in the gallbladder in 38-46% and intrahepatically in 2.5%[1]. The most common histological type is adenocarcinoma, representing 73-84% of cases, followed by anaplastic carcinoma (10%), undifferentiated cancer (5-7%), squamous cell cancer (5%), and other types of neoplasms (1.5%)[5].

The pathogenesis of malignancy in the choledochal cyst is due to the presence of the common pancreatobiliary duct, which generates a sequence of chronic inflammation, cell regeneration, and DNA rupture. Furthermore, K-ras mutations, overexpression of p53, as well as Ca19-9 and Ki-67, have been detected as markers of epithelial instability. Therefore, adequate monitoring and treatment of choledochal cyst is essential to prevent this complication[2,10].

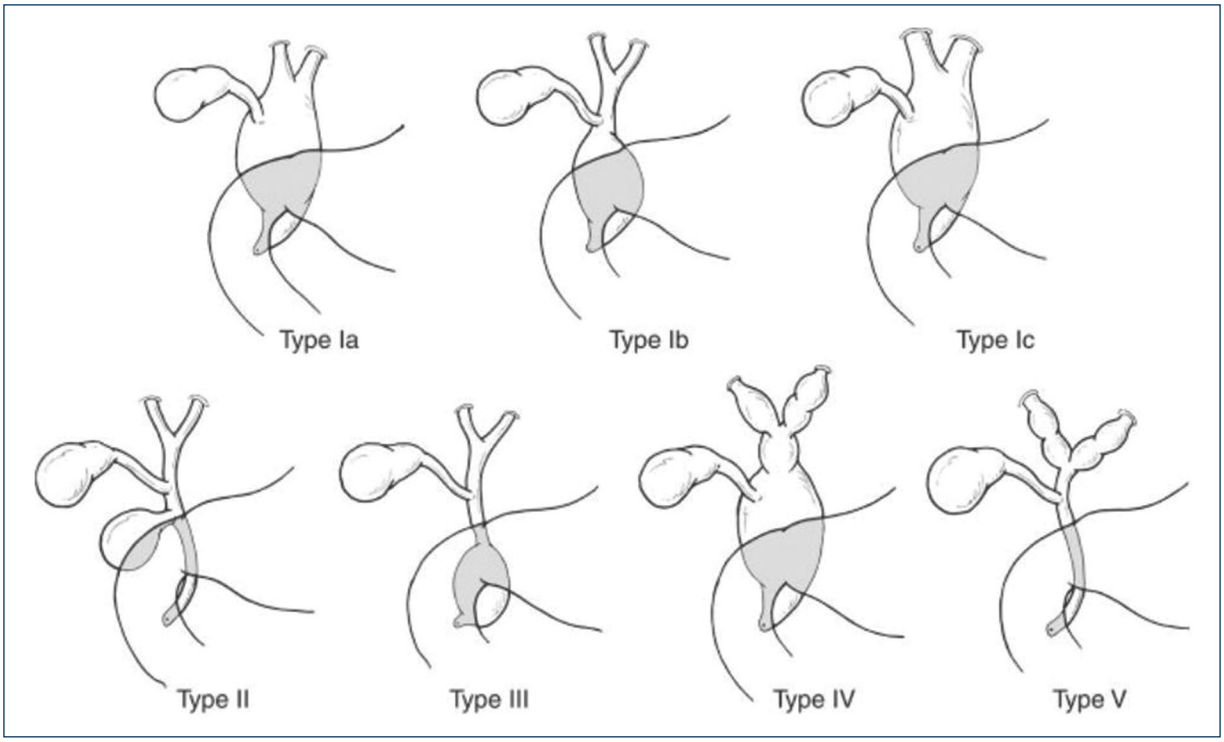

Figure 4. Todani classification[6].

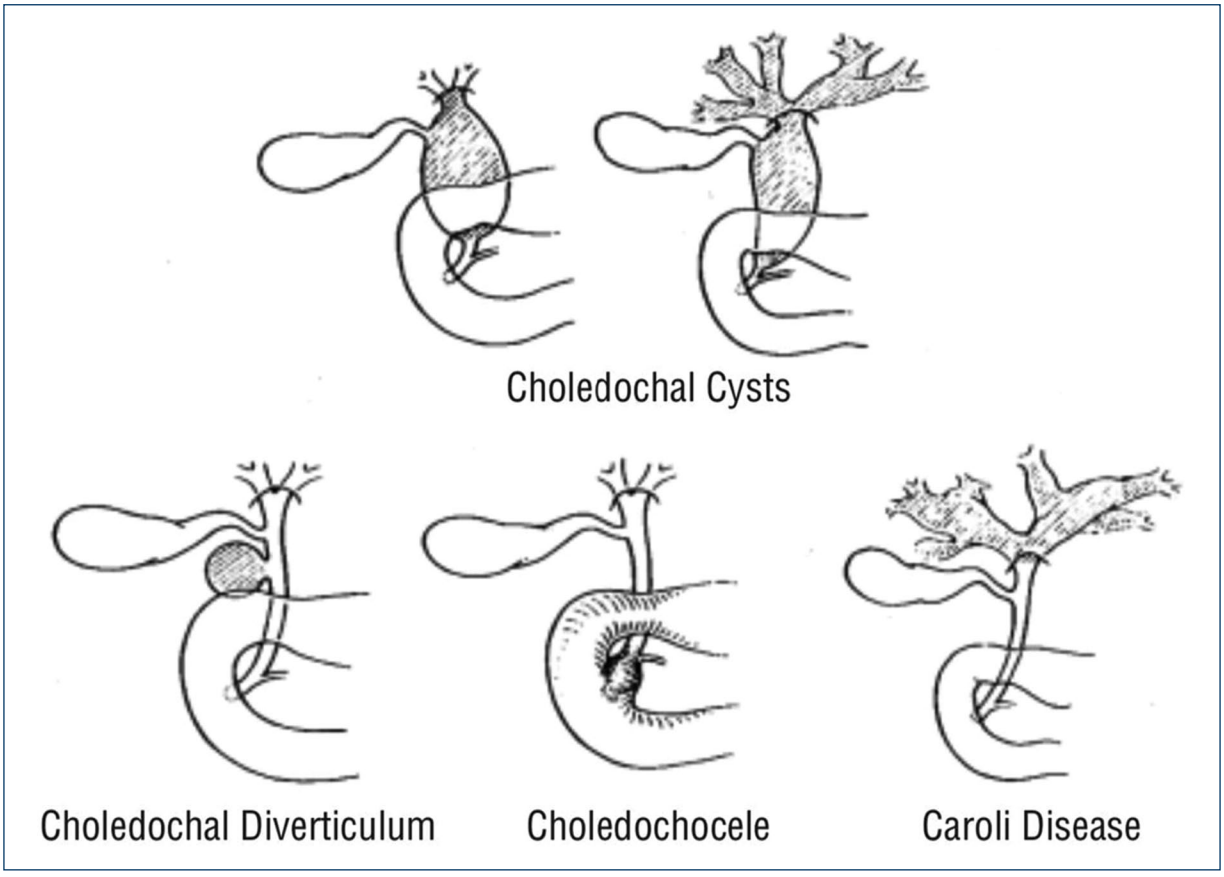

Figure 5. Descriptive classification proposed by Visser.

Table 1. Differences between choledochal cyst classifications

Anatomical and radiological features	Alonso-Lej (1959)	Todani (1977)	Visser (2004)
Cystic dilatation of the entire extrahepatic bile duct, sparing the intra-hepatic ducts, cystic duct and gallbladder	-	IA	Choledochal cysts
Focal/segmental dilatation of the extrahepatic bile. Duct, most frequent distal to cystic duct	-	IB	
Fusiform dilatation of the entire extrahepatic biliary tree	I	IC	
Diverticulum of the extrahepatic duct with narrow stalk connection to the common bile duct	II	II	Choledochal diverticulum
Dilatation of the intraduodenal part of the common bile duct (Choledochocele)	III	III	Choledochocele
Multiple intra and extrahepatic bile duct dilatations	-	IVA	Choledochal cysts
Multiple bile duct dilatations confined to the extrahepatic part	-	IVB	
Multiple bile duct dilatation of the intrahepatic part	-	V	Caroli desease

Ultrasound is the first study for the diagnosis of choledochal cysts, with a sensitivity of 71-87%. It is also useful for surveillance. After the initial assessment, it is necessary to complement other imaging studies to evaluate the pancreatobiliary system. Endoscopic retrograde cholangiopancreatography (ERCP) is considered the study with the highest sensitivity, reaching almost 100%. However, its sensitivity decreases due to the presence of inflammation and scarring. At present, cholangioresonance is considered the gold standard for the diagnosis of choledochal cysts, with a sensitivity of 90-100%, making it possible to delineate the pancreatobiliary anatomy, adjacent structures, stones, and malignancy. Likewise, it is preferred over computed tomography and ERCP due to its non-invasive nature as well as the absence of radiation[1].

The objectives of treatment focus on avoiding acute complications: cholangitis, liver abscess, pancreatitis, and sepsis, as well as chronic complications: portal hypertension, chronic pancreatitis, and the most serious, cholangiocarcinoma[5,11]. Initially, internal drainage was focused on relieving symptoms, being abandoned due to the high rate of complications, mainly malignancy[4]. At present, the treatment of choice is complete resection of the cyst from the bifurcation of the hepatic ducts to the pancreatic parenchyma associated with bilio-enteric reconstruction in Y of Roux. When the cyst extends to the head of the pancreas, the risk of residual cyst versus pancreaticoduodenectomy should be evaluated. The procedure is associated with complications such as cholangitis, biliary pancreatitis, and malignancy. They usually occur in older patients due to fibrosis and inflammation. When the excision of the cyst is complete, the incidence of cholangiocarcinoma is < 1%[1,3,5].

As there are no unified criteria for follow-up, a general review, Ca19-9, and an imaging study (ultrasonography or cholangioresonance) are suggested semi-annually for 5 years, followed by once a year for life. The cutoff values of Ca19-9 at 186 U/mL confer a sensitivity of 100% and a specificity of 94% for cholangiocarcinoma. However, these values can be altered by gastric, pancreatic, or hepatic neoplasms, as well as by benign conditions such as cholestasis[10,11].

Pregnancy represents a stage characterized by important hormonal, physiological, and anatomical changes. During this stage, the increase in estrogen values alters the emptying capacity and concentration of bile in the gallbladder, which is reflected in a tendency to present symptoms of biliary colic[12]. In the context of choledochal cysts, this is a rare pathology but represents an increased risk for the mother, presenting complications such as cholangitis, pancreatitis, peritonitis, and even malignancy, pregnancy loss, and pre-term delivery[13]. Diagnosis is limited to methods free of ionizing radiation due to the risk of congenital malformations, with ultrasound and magnetic resonance being the studies of choice. It is important to take into account the high possibility of variations in normal anatomy derived from the compression generated by the pregnant uterus. Once diagnosed, therapeutic options are limited to conservative management due to the increase in maternal-fetal morbidity and mortality derived from surgery. Diagnosis is limited to methods free of ionizing radiation due to the risk of congenital malformations,

with ultrasound and magnetic resonance being the studies of choice. It is important to take into account the high possibility of variations in normal anatomy derived from the compression generated by the pregnant uterus. Once diagnosed, therapeutic options are limited to conservative management due to the increase in maternal-fetal morbidity and mortality derived from surgery[12-15]. A report of 31 cases shows that the most frequent symptoms were abdominal pain (90%), jaundice (57%), fever (40%), palpable mass (19), childbirth (39%), cesarean section (48%), and abortion (14%). Only 48% reached 36 weeks of gestation. About 50% of the cases did not present complications, so their management was conservative. Drainage procedures were performed in patients who did not respond to the antibiotic, with positive results in all the mothers having only two abortions. Only three maternal deaths were recorded due to abdominal sepsis[14].

Conclusion

This is a unique clinical case due to the low incidence of choledochal cysts in the Western population (1:100,000-150,000)1 with an atypical presentation, occurring asymptomatically until adulthood and being detected incidentally. In < 20% of cases[5], el embarazo limita los métodos diagnósticos sin tomografía ni CPRE debido a la radiación ionizante, la cual aumenta el riesgo de efectos negativos en el producto[1]. The definitive surgical management had to be delayed until the pregnancy was resolved, which ended without complications, managing pain episodes symptomatically. Regarding surgical management, international recommendations were followed, including performing cyst resection, cholecystectomy, and Roux-en-Y bilioenteric anastomosis. Due to the involvement of the head of the pancreas, it was decided to maintain 1 cm of remnant since pancreatectomy is related to high morbidity. The presence of a remnant makes semiannual surveillance mandatory, quantifying Ca19-9 values associated with an imaging study (Ultrasound vs. cholangioresonance) and evaluating the need for a new surgical intervention if the appearance of cholangiocarcinoma is suspected[7,8].

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CLINICAL CASES

Pulmonary teratoma, an exceptional presentation: case report

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Abstract

Introduction: *Teratomas are most frequently located in the thoracic anterior mediastinum.* **Case presentation:** *A 26-year-old female patient who was admitted after respiratory distress was protocolized with a tomography that reported data of cyst tumor involving the upper left pulmonary lobe, a thoracotomy and tumor resection were performed, the pathology report mentioning data from mature cystic teratoma. The presence of pulmonary teratoma has a few reports in the world literature, the thomography provides suggestive findings of cystic tumor and allows surgical planning for its resection.* **Conclusion:** *Pulmonary teratoma is a rare and infrequent entity, its finding requires surgical resection due to the risk of rupture and malignancy.*

Keywords: *Pulmonary teratoma. Mature teratoma. Pulmonary cyst tumor. Thoracotomy.*

Introduction

Pulmonary teratoma is a type of extragonadal germinal tumor whose frequency in the thorax is higher in the anterior mediastinum. Its intrathoracic presence, mainly mediastinal, comprises 80% of this type of tumors[1]; however, its intrapulmonary presentation is exceptionally rare, there have been reports of this entity since 1839 by Mohr[2] and up to 2010 only approximately in 81 cases in the literature[3]. It is usually asymptomatic and is found as a finding due to another condition or in advanced stages there may be symptoms due to compression or invasion of neighboring organs. The importance of making a pre-operative diagnosis involves minimizing surgical risk. If teratoma is not treated it can cause life-threatening complications such as hemoptysis, airway compression, and malignant transformation.

Clinical case

This is a 26-year-old female patient, non-smoker and with no significant history, who attended the emergency department after a 15-day history of non-productive cough, pain in the left hemithorax radiating to the subscapular region, and dyspnea. On moderate exertion, on physical examination, he was afebrile, with the presence of tachypnea and on auscultation crackles were found in the left apical region. He was admitted to the emergency department with the following laboratories: hemoglobin 11.7 g/dL, leukocytes 9.31, 8.35% neutrophils, kidney and liver function within normal limits. A protocol was carried out with a computed tomography that showed the presence of a heterogeneous left apical lesion with a cystic component and suggestive of a lung abscess (Figs. 1 and 2). Antimicrobial therapy is started, and evaluation is requested by the thoracic surgery

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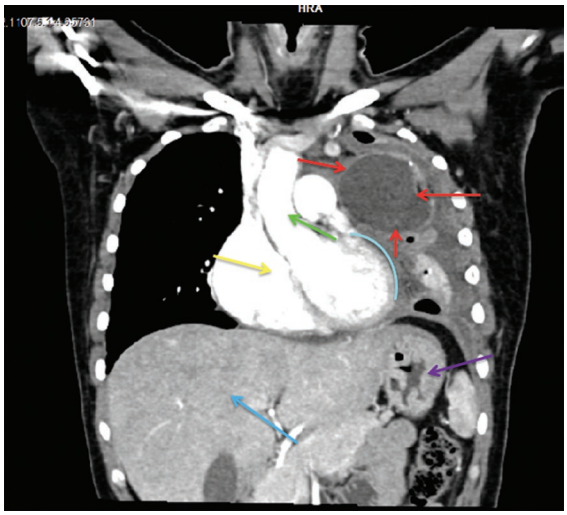

Figure 1. Coronal section contrasted chest tomography. Yellow arrow: cardiac silhouette. Red arrows: tumor mass in the left apical lobe. Blue arrow: liver. Green arrow: ascending aorta. Purple arrow: left kidney. Blue curve: heart delimitation and tumor lesion.

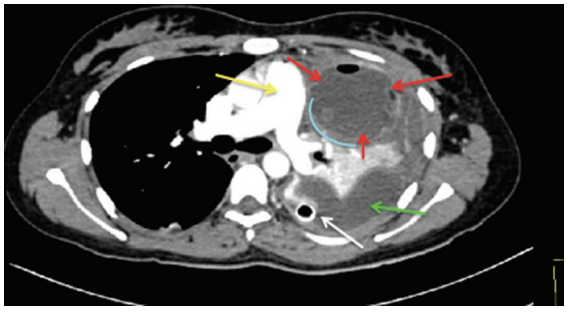

Figure 2. Contrasted chest tomography axial section. Yellow arrow: cardiac silhouette, red arrows: tumor mass, blue arrow: liver, green arrow: ascending aorta, purple arrow: left kidney, blue curve: heart delimitation and tumor lesion.

service and surgical exploration is indicated, a left anterolateral thoracotomy is performed, which presents as findings pleural effusion, pachypleuritis and a 70 × 70 × 35 mm left apical cystic lesion (Figs. 3 and 4), macroscopically with the presence of hair and skin, decortication and tumor resection are performed. The patient was admitted to intensive care with the need for ventilatory support for 48 h, progressing satisfactorily, discharged without complications, and was followed up by the outpatient clinic. It is presented with a definitive histopathological report that reports mature cystic

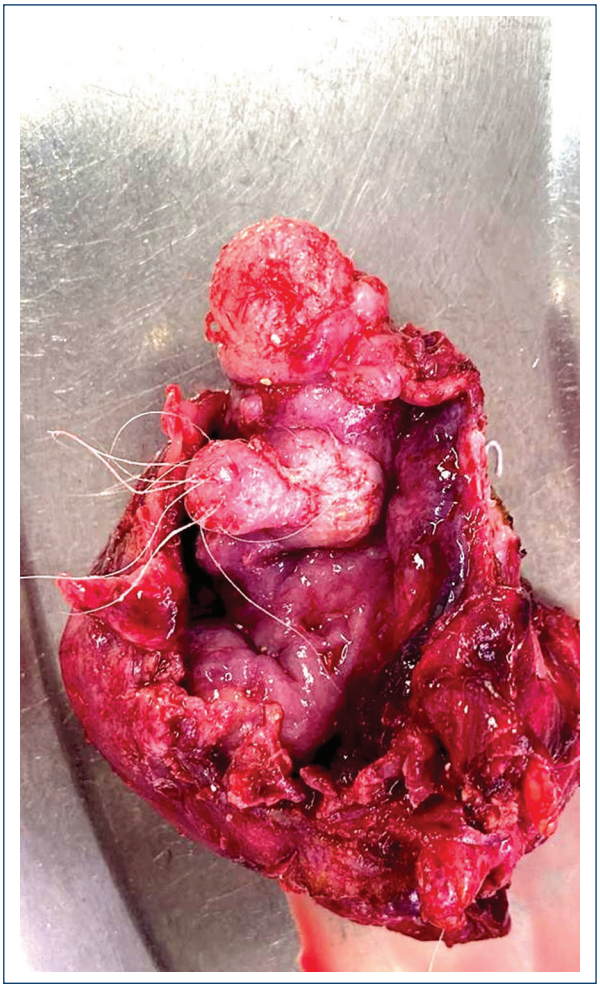

Figure 3. Surgical specimen: cystic teratoma with presence of ectodermal tissue (skin and hair).

teratoma and chronic eosinophilic pleurisy as a finding (Fig. 5). Due to this, in follow-up by external consultation, a tomography of the chest, abdomen and pelvis was requested, which reported chest with post-surgical changes, abdomen without data of tumor activity, intra-abdominal organs with normal characteristics, pelvis with uterus and annexes with normal characteristics, of In the same way, tumor markers are requested that report human chorionic gonadotropin beta fraction of 4 mUI/mL, alpha-fetoprotein (AFP) of 1.60 IU/mL, lactic dehydrogenase 125 IU/L, carcinoembryonic antigen (ACE) 1.49 ng/mL. Now with no recurrence data, she continues to be monitored by the outpatient clinic.

Discussion

The most common mediastinal tumors are lymphoma, thymoma, and teratoma. Teratomas are a type of

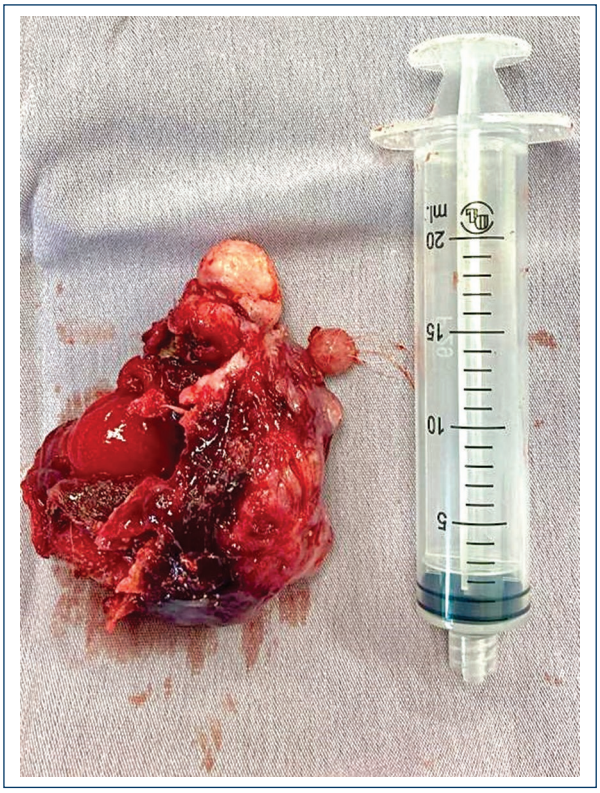

Figure 4. Surgical piece: mature cystic teratoma of 7 × 7 cm.

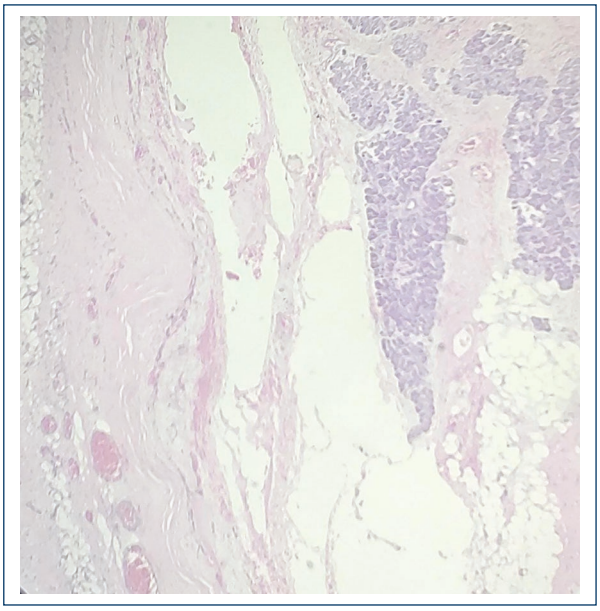

Figure 5. Teratoma components: mature adipose tissue, fibroconnective, monolayered glands, adjacent to the pleura (40×).

tumor that originate from pluripotent cells, with one or more endodermal, mesodermal or ectodermal components[4], to later differentiate into any type of tissue other than the site where they develop. It can involve different organs, most commonly it occurs in the ovaries, testicles, sacrococcygeal region, mediastinum, and other sites[5]. While the extragonadal form represents only 3%, anterior mediastinal teratoma corresponds to 15-20% of this percentage; however, its pulmonary presentation is rare, with few reports of this location[6]. Extragonadal tumors are only considered if there is no evidence of a primary tumor in the testicles or ovaries, which corresponds to the case of this patient. In this case, and due to the characteristics of the patient and the tumor, it is worth mentioning the existence of giant mediastinal tumors, which are defined as a tumor that occults half of the hemithorax or if it is > 10 cm, with a similar clinical picture. Despite the size, the prognosis will depend on the histological type with respect to the risk of metastasis and recurrence. Several studies suggest that both thoracic and mediastinal teratomas have a common origin from thymic tissue of the third pharyngeal arch[7]. They usually present as an imaging finding. The criterion is determined with exclusive pulmonary origin and the exclusion of gonadal origin or another primary extragonadal site[8]. There is a predilection for the left upper lobe, which corresponds to the case presented[9]. They usually present as mature, benign teratoma. Their age range varies from 10 months to 68 years, with no preferences between both sexes[10]. Up to 53% are usually asymptomatic[11], while in advanced stages, its clinical picture is insidious, it can present chest pain, hemoptysis, and cough, in advanced stages, the symptoms are the result of compression/obstruction of neighboring organs, there are even reports of trichoptysis in cases of invasion of the tracheobronchial tree[12]. The use of chest radiography, computed tomography, and magnetic resonance can be used to consider the diagnosis and assess its resectability[13]. Findings such as thin-walled cysts and calcifications are highly suggestive of teratomas, which in the case of the patient led to the presence of a cyst versus lung abscess. Tumor markers such as AFP and beta-hCG are usually at reference values; their elevation suggests the presence of a malignant tumor[14], the rest of the laboratories usually report within normal parameters. In the case of this patient, due to her urgent admission to the operating room, the protocol with tumor markers and extension studies was carried out after the intervention, with no data on tumor activity in another region and the tumoral markers in normal parameters. This kind of tumor is

generally benign, the size of the tumor is not related to malignancy. Malignant teratomas tend to be solid and nodular, while benign ones are frequently cystic and contain mature tissue. Macroscopically, your multicystic tumor is found that contains hair, teeth or skin. Its differential diagnosis depends on the location and includes bronchogenic cysts, pulmonary hamartomas, and cystic lymphangioma. Surgical treatment with complete resection by sternotomy, thoracotomy, or video-assisted thoracoscopy is the management of choice due to the potential risk of rupture and injury to other organs by proteolytic enzymes, as well as the risk of malignant transformation, which occurs in 1-2%[15]. In the case of presenting an immature teratoma, the combined approach with surgery and two circles of chemotherapy (BEP) is foreseen. Follow-up after resection is defined by histopathology, while malignant tumors require adjuvant treatment and close follow-up due to the risk of local or distant recurrence. While in benign tumors, local recurrence is not expected after en bloc resection with free margins[16].

Conclusion

There are some peculiarities in this case that require consideration, mainly due to the unusual location of the tumor, despite the fact that 5% of germ cell tumors have an extragonadal origin, the pulmonary location to date has few reports in the literature. The use of tomography for pre-operative diagnosis is important for surgical planning; however, the diagnosis is not always integrated due to the unusualness and non-specific symptoms of the condition. The definitive management is surgical with complete resection to avoid complications and malignant transformation. Close follow-up is important, even more so if the pathology result shows the presence of an immature teratoma, which, due to the risk of recurrence and metastasis, will require support with adjuvant treatment. The relevance of this type of cases is knowing about their existence, having a high index of suspicion and being able to carry out a complete diagnostic approach for adequate surgical planning, which will have direct repercussions on the evolution and outcome of the patient.

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CLINICAL CASES

Xanthogranulomatous cholecystitis. A rare case of chronic cholecystitis simulated gallbladder cancer

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Abstract

Introduction: *Xanthogranulomatous cholecystitis is a rare inflammatory disease of the gallbladder a variety of chronic cholecystitis. Some patients with this entity are misdiagnosed with gallbladder cancer, since the radiological findings and clinical manifestations are similar, which means unnecessary radical surgery, with increased morbidity and mortality.* **Case presentation:** *A 60-year-old male patient with a pre-operative diagnosis of cholecystitis plus dilation of the bile duct, and suspicion of a neoplastic process of the gallbladder in the post-operative period.* **Conclusion:** *The difficulty of an evident differential diagnosis after imaging techniques gives clinical relevance to the picture.*

Keywords: *Xanthogranulomatous cholecystitis. Gallbladder cancer. Chronic cholecystitis.*

Introduction

Xanthogranulomatous cholecystitis (CX) is a rare inflammatory entity of the gallbladder. CX is a variety of chronic cholecystitis with a very low frequency 0.7-13%[1,2] characterized by a destructive inflammatory process of a focal or diffuse nature, with an accumulation of layers of lipid macrophages, fibrous tissue, and acute and inflammatory cells. chronicles[3]. It is increasingly recognized by pathologists and clinicians, with an increase in its frequency identified. CX is more common than gallbladder cancer, in Japan it fluctuates from 0.7% to 1.8% of all cholecystectomy specimens, in the US from 1.2% to 10%, and 10% to 13.2% in India[4]. The occurrence in our environment of CX in surgically resected vesicles ranges from 1% to 2%.

CX was first described by Christensen and Ishak in 1970, as a pseudotumor of the gallbladder[5], but it was not until 1981 that Goodman and Ishak, from the Institute of Pathology of the United States Armed Forces, coined the term CX, due to its similarity to xanthogranulomatous pyelonephritis[6]. The diagnosis is histopathological; however, ultrasound and tomographic data that raise suspicion in this entity show the presence of gallstones (multiple or individual) most of the time[7]. Morbidity from this disease is 32% and no data are available regarding mortality[8].

The objective of the study was to describe the clinical, radiological, and surgical findings, as well as the histopathological results of a clinical case of a patient with CX diagnosed post-surgery as gallbladder cancer.

Case report

A 60-year-old male patient, with no pathological or surgical history, came to the emergency room due to abdominal pain of 2 days' duration in the epigastrium after eating; intensity of 8/10 on the pain scale, stabbing type that radiates to the lumbar region, associated with nausea without vomiting. The patient had scleral jaundice, abdomen with pain and resistance in the right

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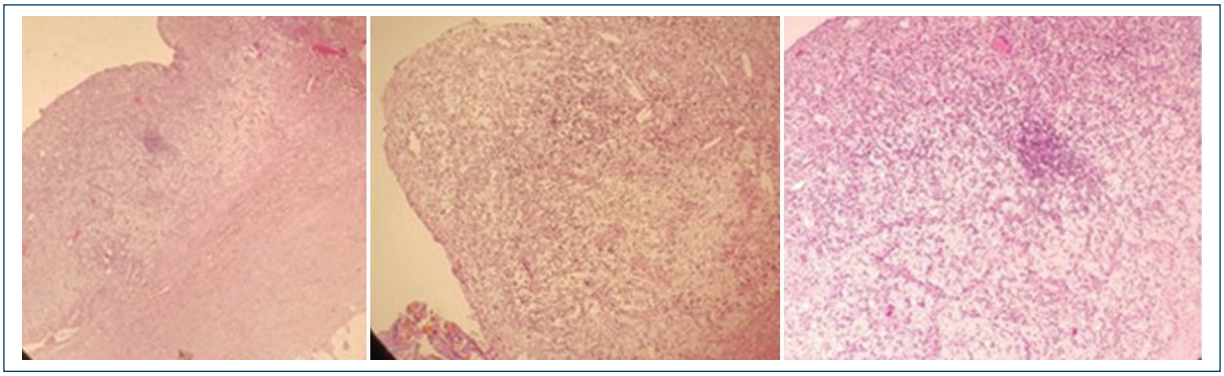

Figure 1. Histopathology of the gallbladder wall: typical macrophage histiocytes filled with debris and lipids; fibroblasts and accumulations of mononuclear lymphocytes with some giant cells.

upper quadrant, and a positive Murphy sign. The studies showed leukocytosis with left shift, preserved renal function, normal pancreatic enzymes, elevated bilirubin at the expense of direct bilirubin, and elevated liver enzymes with a cholestatic pattern. Abdominal ultrasound showed hepatomegaly with mild hepatic steatosis, dilation of the extrahepatic bile duct with a 9-mm common bile duct without evidence of an obstructive cause, gallbladder with WES complex, normal pancreas, without free fluid. Due to the worsening of the clinical picture and data of systemic inflammatory response, it was decided to take him to the operating room.

Open cholecystectomy shows a gallbladder with thickened hourglass walls with a stone fixed in the gallbladder fundus and in Hartman's pouch, abundant fibrosis containing thick bile, firmly adhered to the liver which made dissection of the bed difficult, Callot's triangle with marked fibrosis, thickened cystic duct with stenosis of its lumen, bile duct dilated approximately 10 mm, bile duct and intraoperative cholangiography without evidence of stones and adequate passage of distal and proximal contrast medium. The procedure is defined as extrinsic obstruction of the bile duct due to Mirizzi I Syndrome versus gallbladder neoplasia. The patient progressed adequately with progressive normalization of the liver profile and was discharged on the 4th day without complications. The pathological result was received with a CX report (Fig. 1).

Discussion

Various terms have been used to describe this process including ceroid granuloma, ceroid-type histiocytic granuloma, fibroxanthogranulomatous inflammation, and biliary granulomatous cholecystitis, although most authors now refer to it as CX[9]. CX is a pathology with a difficult diagnosis, frequently confused with gallbladder cancer; 40% of cases present intraoperative diagnostic doubts with this neoplasm. It is more common in women between 60 and 70 years old, generally with associated comorbidity, and in almost all cases, it is associated with long-standing cholelithiasis. Unlike the usual chronic cholecystitis, it is a process with a greater local infiltrative and destructive tendency[10].

Histologically, it is defined by the rupture of the Rokitansky-Aschoff diverticula, which are formed after a process of chronic cholecystitis, or by ulceration of the mucosa. This produces a severe inflammatory reaction in the interstitial tissue composed of fibroblasts and macrophages, which, in the process of inevitable phagocytosis of cholesterol and bile phospholipids, secondarily conditions a destructive enzymatic discharge of the tissue microenvironment of the gallbladder wall and its surroundings. This causes tumor-like thickening of the wall, with the appearance of yellowish nodules or striae that can extend to adjacent structures and organs with the formation of fistulas due to ulceration of the mucosa[11,12] and tissue fibrosis similar to the desmoplastic reaction typical of some tumors. epithelial, resulting in a macroscopic appearance similar to neoplastic[13]. Obstruction and chronic infection caused by the presence of stones produce degeneration and necrosis of the gallbladder wall, with the consequent formation of intramural abscesses. These are replaced by xanthogranulomas with exogenous foamy giant cell histiocytes, implicated as contributing factors similar to what has been experimentally demonstrated in xanthogranulomatous pyelonephritis[14].

Radiologically, CX can be confused with gallbladder cancer; one and the other can coexist in 10% of

cases[15]. Ultrasound shows hyperechogenic thickening of the gallbladder wall and the identification of intramural hypoechoic nodules representing foci of xanthogranulomatous inflammation or small abscesses, alteration of the mucosal lining, perivesicular fluid, stones, and bile sludge, and hyperechogenicity of the adjacent liver parenchyma[16,17]. Computed tomography shows, in addition to mural thickening, hypodense intramural nodules (5-20 mm), heterogeneous wall enhancement, and infiltration into the perivascular fat and adjacent liver parenchyma. Magnetic resonance imaging (MRI) shows xanthogranulomas with higher signal intensity on T2-weighted images. In-phase and out-of-phase sequences help identify intramural fat and diffusion sequences help differentiate CX from wall thickening due to gallbladder carcinoma[18]. In our case, MRI was not performed since it is not a routine diagnostic method in chronic cholecystitis.

Clinical symptoms, physical examination, and laboratory results are not useful to differentiate this pathology from other more common gallbladder or bile duct diseases. The clinical presentation does not differ from the spectrum of cholelithiasis and there are no specific characteristics of the disease. There are reports that describe the presence of a palpable mass in slightly less than half of the cases as well as mild jaundice in half of the patients who presented acutely. Some patients present perivascular or hepatic abscesses, gallbladder perforation, or enterobiliary fistulas, with figures ranging from 23% to 32%[19]. In our case, we found vesicle adhesions to loops of the small intestine, Mirizzi Syndrome I, and a decrease in the lumen of the cystic duct.

At present, cholecystectomy is preferable to initial medical treatment in acute cholecystitis; in the case of cancer, it should be performed by a surgeon with extensive experience in liver surgery[20]. When gallbladder cancer is suspected, it must be assumed that most cases are unresectable or have a poor prognosis and that laparoscopic surgery is contraindicated due to the increased risk of bile effusion, gallbladder rupture, peritoneal dissemination, and recurrence. Surgical management of gallbladder cancer is indicated in the earliest stages of the disease and it is important to evaluate therapeutic options in patients with advanced disease[21]. An expert consensus and the National Comprehensive Cancer Network guidelines recommend simple cholecystectomy in patients with T1a. For non-metastatic higher stages, extended oncological resection is recommended in patients with stages T1b, T2, and T3 who do not have disseminated disease. This surgery includes gallbladder fossa resection, IVb and V bisegmetectomy, and regional lymph node dissection; common bile duct resection and hepatectomies are considered in some selected patients. Patients with advanced stages (T4) treatment are palliative[22].

Many experts emphasize the need for general surgeons to become familiar with this pathology since CX can mimic hepatobiliary neoplasia in its clinical presentation, radiological findings, intraoperative appearance, and rarely, histological appearance, which often makes pre-operative distinction between these entities impossible[23].

Conclusions

CX is a rare pathology that usually affects patients with long-standing cholelithiasis. As it occurs in patients with greater comorbidity, post-operative complications also increase. CX can simulate a neoplasm or coexist with gallbladder carcinoma. Differentiation between CX and gallbladder cancer can be made intraoperatively by fine-needle aspiration biopsy or cold section. Due to the possible association between these two pathologies, the surgical procedure of choice should be total cholecystectomy. At present, it is impossible to macroscopically differentiate CX and gallbladder adenocarcinoma, so the definitive diagnosis is obtained until the histopathological analysis of the surgical specimen. CX can be a diagnostic challenge and a therapeutic dilemma due to its similarity to gallbladder neoplasia.

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CLINICAL CASES

Paget-Schroetter syndrome: case report

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Abstract

Introduction: *The Paget-Schroetter Syndrome is one in which the compression of the axillary and subclavian veins generates thrombosis, leading to blood stasis in the venous system of the upper limb and its respective complications. First-line treatment focuses on thrombolysis and anticoagulation of patients.* **Case presentation:** *A 23-year-old female patient attended the emergency department due to edema and erythema of the right thoracic extremity for 1 day, as well as pain in the axillary region when moving the arm and infraclavicular pain on palpation, and paresthesia in the fingers, that started 2 hours before consultation.* **Conclusion:** *However, the debate continues as to whether the resection of the first rib as a decompression method should be applied to all patients or whether, depending on the characteristics of each patient, it is possible to follow other regimes to avoid the recurrence of symptoms and even subsequent thrombosis episodes.*

Keywords: *Thrombosis. Anticoagulants. Thrombolysis. Decompression.*

Introduction

Paget-Schroetter syndrome is a compression of the subclavian and axillary veins that triggers thrombosis of the affected veins. This is generated due to the decrease in space between the first rib, the clavicle, the scalene muscle, the subclavian muscles, and the costocoracoid ligament[1,2]. Venous thrombosis in these regions accounts for 2-4% of all cases of venous thrombosis deep, with an incidence of 11 of every 1,000,000 hospital admissions[2]. This syndrome is responsible for 10-20% of deep vein thromboses of the thoracic extremities and 30-40% of spontaneous thromboses of the axillo-subclavian veins[3,4].

The pathophysiology is due to an injury to the vascular endothelium, which triggers the coagulation cascade until a thrombus forms within the lumen and the flow is obstructed[3-5]. This damage is generated when performing physical effort with the shoulder, which is why it is observed in young, previously healthy, and physically active patients[1,3-6]. Repeated damage generates perivascular fibrosis, which perpetuates the stenosis of these veins[4].

The clinical signs observed are a sensation of heaviness of the affected limb, pain, edema, erythema, cyanosis, collateral venous network, and visibly dilated veins in the shoulder (Urschel sign). Most patients present symptoms within the first 24 h after exertion[1,3,4,6].

The imaging study to be performed in the first instance is Doppler ultrasound, with a sensitivity of 78-100% and a specificity of 82-100%. However, the gold standard is 1.4 contrast venography. Subsequently, phlebography should be performed to observe thrombosis of the subclavian and axillary veins, as well as the collateral branches, and a provocation maneuver that consists of abducting the limb to observe flow obstruction 1 must be performed. Among the differential diagnoses are found: cellulitis, lymphedema, neoplastic compression, and superficial thrombosis[3]. It is advisable to evaluate other possible causes of hypercoagulability[3].

The first step of treatment consists of anticoagulation followed by catheter-directed intrathrombolysis using alteplase or reteplase within the first 10 days[1,4,6]. It is suggested that no more than 2 weeks have passed

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from thrombus formation to the procedure since this is associated with a success rate of 75-84%, while if it is performed between 2 and 12 weeks this rate decreases to 29%[4,7]. Likewise, the patient must be anticoagulated for 3-6 more months with low molecular weight heparin (LMWH) and subsequently with Vitamin K[1,6] antagonists. More than 90% of patients manage to return to their daily activities.

Resection of the first rib by transaxillary, supra, or infraclavicular resection is recommended by multiple authors, although a claviculectomy can also be performed, since thrombosis after thrombolysis is generated in up to a third of cases[1,6,7]. Finally, angioplasty is performed and a stent is placed if considered necessary[1,2]. Complications of thrombosis include pulmonary thromboembolism, post-thrombotic syndrome (associated with conservative treatment with anticoagulants), recurrent thrombosis and perivascular fibrosis, the latter occurring in 10-12% of patients[2-4].

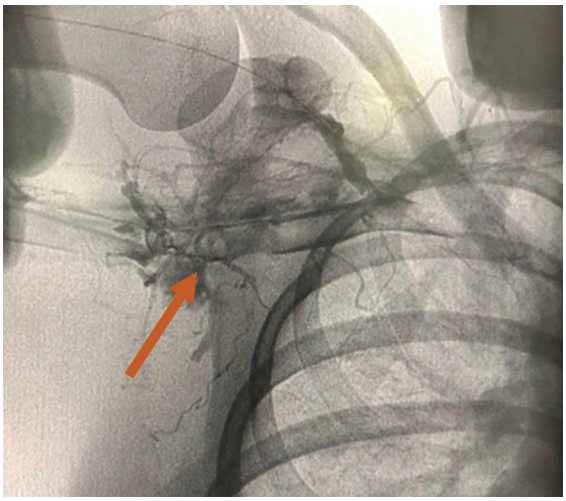

Figure 1. Obstruction of the passage of contrast medium during catheter placement for thrombolysis of axillary and subclavian veins (arrow) due to the presence of thrombus and its passage through the developed collateral circulation.

Case report

A 23-year-old female patient attended the emergency department due to edema and erythema of the right thoracic extremity for 1 day, as well as pain in the axillary region when moving the arm and infraclavicular pain on palpation, and paresthesia in the fingers, that started 2 hours before consultation. In previous days, the patient reported having a feeling of heaviness in her right thoracic extremity after lifting weights in the gym.

The patient reports having been lifting weights for 4 years, using an oral contraceptive with estradiol and dienogest for 1 year, changing this method to a dermal patch with ethinyl estradiol and norelgestromin 1 month before the development of symptoms. As a family history, his father developed deep vein thrombosis of the left pelvic extremity after spinal instrumentation.

On physical examination, the patient presented edema and erythema of the right thoracic extremity, collateral and dilated veins in the shoulder, pain along the brachial and axillary veins when compressing the forearm, decreased radial pulses, and capillary refill lasting 4 s. She was administered 1.5 mg/kg/24 h of LMWH as the first approach and the next day, a Doppler ultrasound was performed where thrombosis of the axillary and subclavian veins was found. Conservative therapy was started with rivaroxaban 40 mg per single dose and subsequently 20 mg every 24 h, however, the condition deteriorated with the onset of dull pain in the right suprascapular region which gradually increased in intensity and radiated through the body. The dorsum until it reaches the ipsilateral lung base and becomes transfection, preventing deep inspiration.

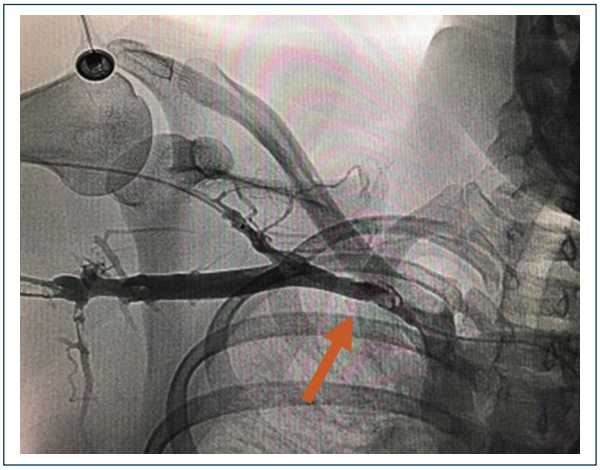

Figure 2. Passage of contrast medium through axillary and subclavian veins free of obstruction after thrombolysis. Tortuosity (arrow) is seen in the path prior to performing balloon angioplasty.

A Doppler ultrasound was performed again, which confirmed deep vein thrombosis of the subclavian and axillary veins with data of brachial, ulnar, and radial venous insufficiency. Subsequently, a computed tomography angiography was performed where pulmonary thrombosis of the right posterior basal segmental

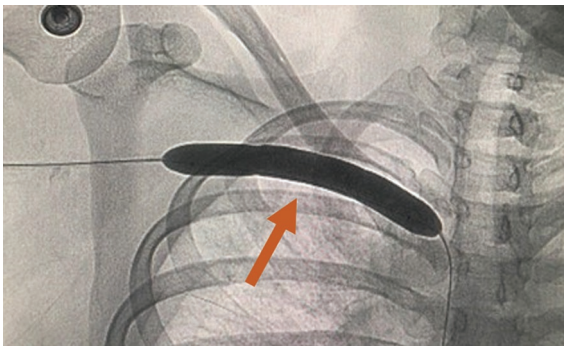

Figure 3. Balloon angioplasty (arrow) of axillary and subclavian veins after thrombolysis and verification of viability of venous circulation by passing contrast medium.

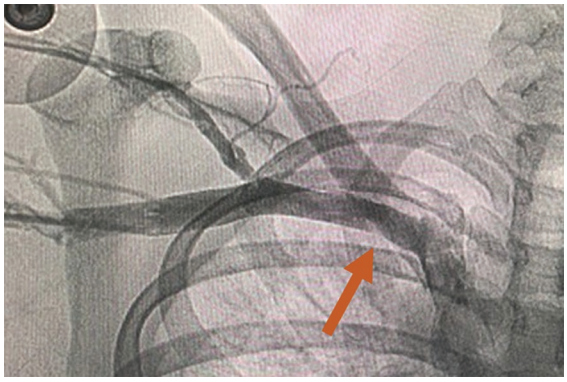

Figure 4. Axillary and subclavian veins after thrombolysis and angioplasty where complete viability (arrow) is observed when contrast medium is used.

branch was found, an area of condensation in the right posterobasal segment probably related to pulmonary infarction and posterobasal linear atelectasis.

As a treatment, catheter-directed intrathrombo thrombolysis with alteplase as seen in figure 1, was used. The veins affected were tortuous and narrow, as can be appreciated in figure 2. Subsequently, balloon angioplasty (Fig. 3) was performed where the absence of obstruction within the veins was confirmed, as shown in figure 4. Due to the favorable evolution of the treatment, she was discharged from hospitalization and a follow-up appointment was scheduled one month after the procedures. During this time, he did not present obstructive symptoms, and when a Doppler ultrasound was performed 30 days after hospital discharge, compressible veins were observed without compromised flow, so it was not necessary to perform decompressive surgery.

Discussion

Silverberg et al. determined that the exclusive use of oral anticoagulant therapy is associated with post-thrombotic syndrome in 44% of Cases[8]. In this retrospective study, 18 patients who did not receive surgical treatment to resect the first rib were followed, and determined residual symptoms using the villalta scoring system, in which it was observed that 94% had minor symptoms. Likewise, the Quick-DASH system was used to evaluate the impact on quality of life, where 83% of the patients did not present any alteration in their quality of life and all managed to return to their work activities, and an ultrasound was performed to evaluate the viability of the brachial, axillary and subclavian veins, demonstrating that 89% of patients maintained the patency of the affected vein, even years after thrombolysis[8].

An athlete presented thrombosis on two occasions treated with anticoagulation, due to having a factor V Leiden and PT20210 mutation. Anticoagulant treatment was maintained for 6-12 months in patients without thrombophilias and for life in those with a related condition. Therefore, they determined that the majority of patients who do not receive surgical treatment remain asymptomatic, although slight circulatory changes may be observed. It is important to consider that surgical decompression can promote brachial palsy, chronic pain, chylothorax, phrenic nerve paralysis, arterial injury, and hemorrhage in 5-15% of patients[8].

Lee et al. formulated an algorithm where only those patients who remained symptomatic upon subsequent visits received surgery. In their study, 23% of patients who did not receive surgical treatment developed thrombosis, but the time they remained anticoagulated was only 5 months, compared to the patients studied by Silverberg et al., where the average treatment time was 26 months[8,9].

Lee et al. conducted another study where he found that of 22 patients with upper limb thrombosis (only nine remained without surgical treatment), eight of these had minimal symptoms after anticoagulant treatment and only one persisted with moderate symptoms although he did not present obstruction. Eleven of 13 patients who underwent decompressive surgery continued to partially present the initial symptoms to a lesser extent and two other patients remained with the same degree of symptoms. Lee and his group point out that not all patients should be treated surgically and that they should be followed up while they are undergoing treatment to determine if decompressive surgery is required, as it exposes the patient to multiple risks[10].

However, in a meta-analysis carried out by Karaolanis et al., where the majority of patients received surgical intervention for resection of the first rib, complications associated with surgery were ≤ 6%, and are related to a greater rate of vascular viability and lower symptoms compared to patients who received anticoagulation and thrombolysis, which is why it is recommended in patients with low or intermediate surgical risk[11].

Likewise, Hangge et al. determined that anticoagulation is less effective than thrombolysis and decompressive surgery. The results of thrombolysis are superior if it is performed within the first 2 weeks, but without mechanical decompression, there may be a relapse in a third of patients, which is why surgical treatment is recommended in patients who are candidates[7].

Conclusion

Despite the great advances regarding the definitive treatment of thoracic limb thrombosis, it is still necessary to carry out more studies with a large number of participants that compare the effectiveness of the long-term use of oral anticoagulants and resection of the first rib after thrombolysis. Likewise, a study is considered necessary to evaluate different variables that patients may present, such as antiphospholipid syndrome, mutations, or protein deficiencies that generate procoagulant states, where the resolution of Paget-Schroetter syndrome also involves and considers the management of these states, to avoid surgical risks and use the advantages of oral anticoagulation.

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CLINICAL CASES

Endovascular repair of a complex ulcer of the descending thoracic aorta

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Abstract

Introduction: *Penetrating aortic ulcer is a focal lesion that develops in the region of an atheromatous plaque that penetrates the internal elastic lamina of the vascular wall. It can occur in any of the aortic segments and is the rarest entity of acute aortic syndrome, also composed of aortic dissection and intramural hematoma.* **Case presentation:** *It may be present together or isolated with dissection and intramural aortic hematoma, which, if not treated adequately and correctly, compromises the life of those who suffer from it.* **Conclusion:** *The study aimed to demonstrate the benefits of endovascular repair of the components of acute aortic syndrome: aortic ulcer and intramural hematoma.*

Keywords: *Aortic ulcer. Endovascular repair. Acute aortic syndrome. Descending thoracic aorta.*

Introduction

Penetrating aortic ulcer was first described by Sheenan T but differentiated from other aortic pathologies by Stanson in 1986 during his report on penetrating atherosclerotic ulcers of the thoracic aorta[1]. Penetrating aortic ulcer represents one of the entities of acute aortic syndrome, which develops in the area of an atheromatous plaque that penetrates the internal elastic lamina of the vascular wall or due to inflammatory changes in the aortic wall. Risk factors for its development include hypertension, male sex, smoking, atherosclerosis, previous aortic operations, and connective tissue disorders. These may be asymptomatic and only be detected incidentally in imaging studies, or they may cause symptoms of chest pain, mild abdominal pain, chronic low back pain, or dyspnea[2,3].

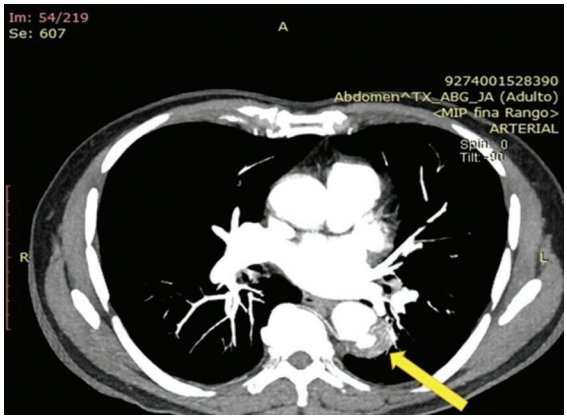

Figure 1. The ulcerated lesion of the descending thoracic aorta indicated by the yellow arrow.

The clinical tendency of penetrating aortic ulcers is variable; they may remain stable, increase in size, or progress to an intramural hematoma, dissection, pseudoaneurysm, or aortic rupture if the ulcer ruptures the adventitia.

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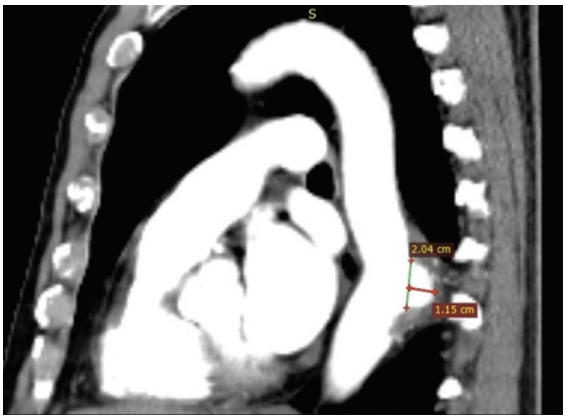

Figure 2. Ulcer measurements.

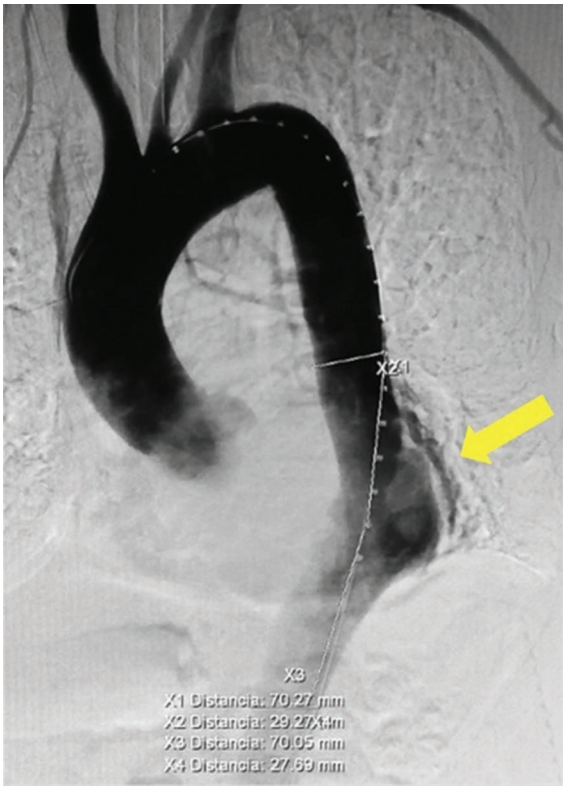

Figure 3. Aortography showing the aortic ulcer indicated by yellow arrow.

The development of multiple endovascular technologies (guides, endoprostheses, intravascular ultrasound, and catheters) has allowed a better understanding and resolution of this type of pathologies. Furthermore, endovascular aortic ulcer repair offers a less invasive approach to the treatment of affected patients, with very encouraging results in the short and medium term, thus reducing mortality and comorbidities for the benefit of those with this type of vascular disease[3].

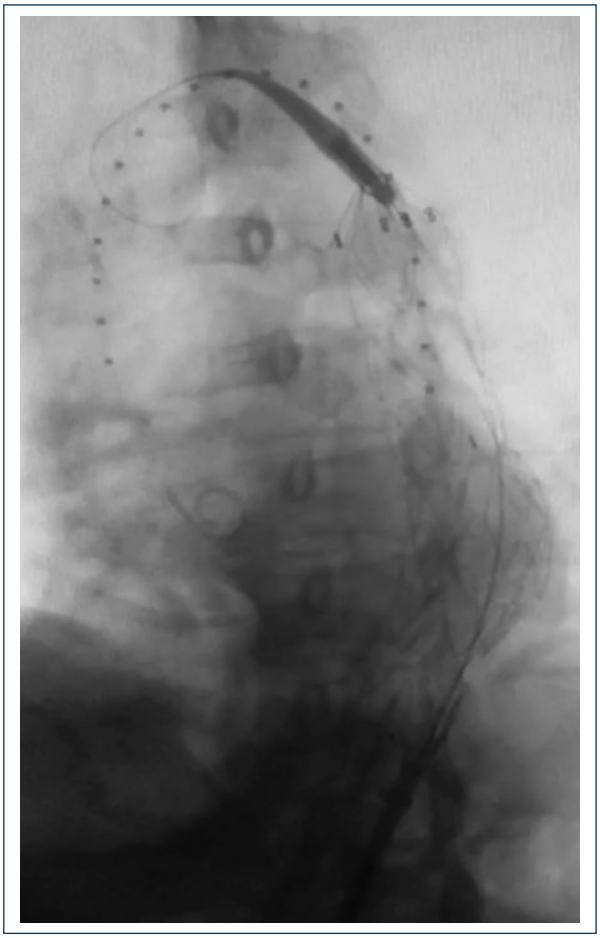

Figure 4. Endoprosthesis release to exclude aortic ulcer.

Case report

A 76-year-old male patient with a history of systemic arterial hypertension and a smoker, who came to the emergency department due to chest pain radiating to the back suddenly and progressively, without cardiac alterations. The angiotomography shows an ulcerated lesion at the level of the descending thoracic aorta measuring 2.04 cm deep by 1.15 cm long associated with intramural hematoma (Figs. 1 and 2). The patient is referred to our service for case evaluation and treatment.

Due to the clinical characteristics of the patient, the anatomical location for approaching the lesion and evaluating the high risk of aortic rupture, and endovascular repair with the Valiant™ Captivia Stent Graft

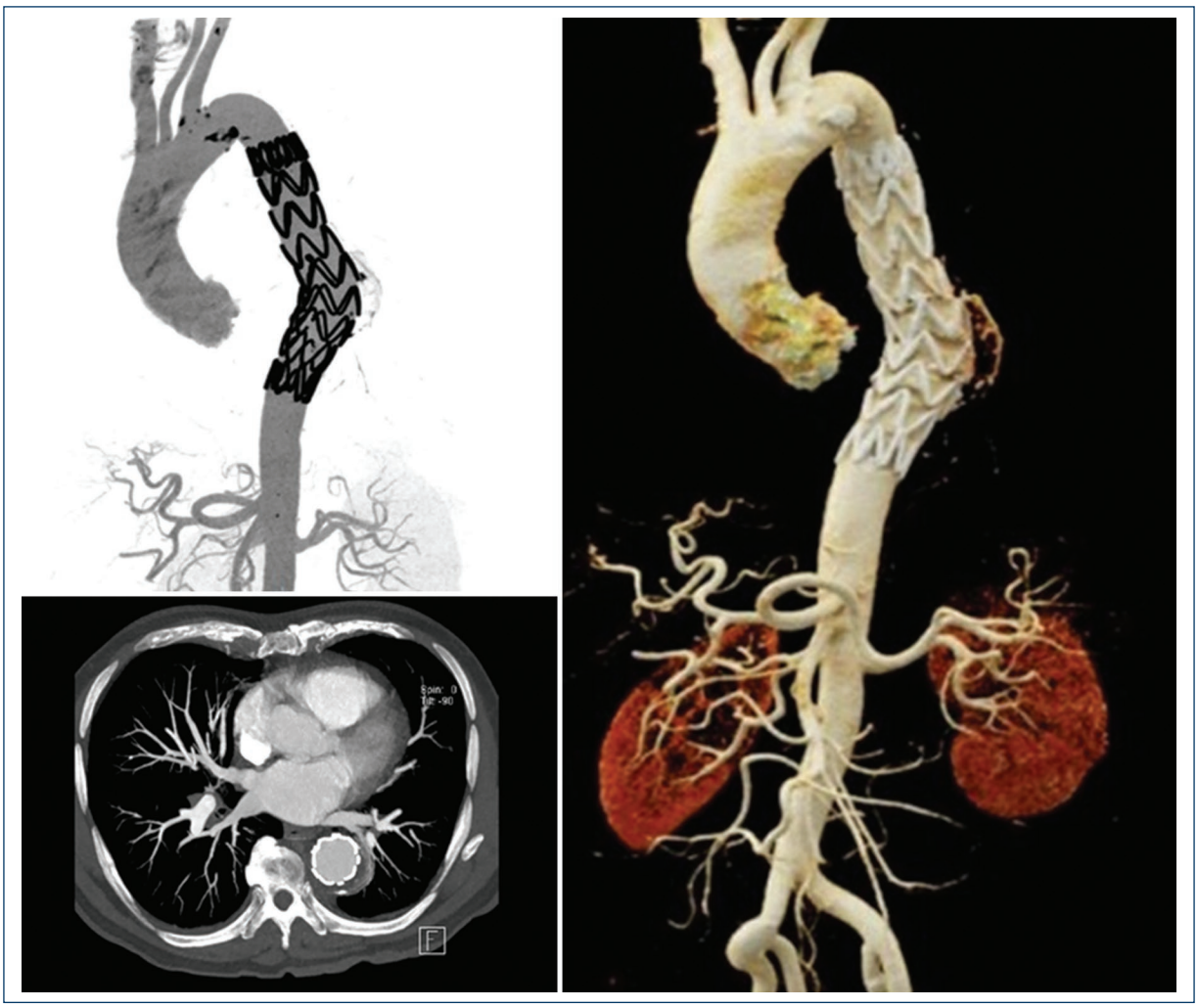

Figure 5. Post-surgical outcome.

stent graft with the Captivia™ Delivery System was scheduled, after meticulous planning surgery of the case. The lesion was excluded without complications (Figs. 3-5).

Discussion

An aortic ulcer diameter > 15 mm represents a high risk of disease progression toward aortic dissection, the appearance of pseudoaneurysm, or aortic rupture, with greater comorbidity. Patients benefit from early intervention in principle through aggressive blood pressure control with intravenous β-blockade and aggressive analgesia to control pain if present. In addition: Once the patient is stabilized, endovascular surgery should be performed. Exclusion of the aortic ulcer as long as adequate surgical planning is carried out and the aortic anatomy allows it[3-5].

In the endovascular repair of the aortic ulcer, we can obtain better data on the anatomical and topographic characteristics of the pathology, in addition, the accessibility to the affected area is faster, less morbid, and with excellent results.

One concern about performing endovascular repairs of this type of pathology lies in the potential risk of progression to aortic dissection. This iatrogenic complication could be caused by the oversizing of the graft through the aortic wall in the proximal or distal landing zones. To avoid this complication, the area of ulceration must be completely excluded and at a safe landing distance of the endoprosthesis > 20 mm from the edge.

Without a doubt, the open surgical repair procedure in these patients adds complexity and, in turn, greater

morbidity and mortality, which is why it is currently displaced by endovascular procedures. In Mexico, there is no database where the evolution results of this type of pathology can be compared.

We decided to repair this pathology using an endovascular technique to precisely prevent its progression and unfavorable repercussions for the patient's quality of life. With the procedure, we achieved complete exclusion of the injury, without complications for the patient and with a satisfactory recovery, in addition, after being discharged from the hospital 48 h after the procedure.

Conclusion

Endovascular repair of the pathologies that make up acute aortic syndrome is a highly resolving, reproducible technique that achieves the desired goal of improving the patient's quality of life and reducing major complications associated with open surgical interventions.

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Conflicts of interest

The authors declare no conflicts of interest.

Ethical disclosures

Protection of human and animal subjects. The authors declare that no experiments were performed on humans or animals for this study.

Confidentiality of data. The authors declare that they have followed the protocols of their work center on the publication of patient data.

Right to privacy and informed consent. The authors have obtained the written informed consent of the patients or subjects mentioned in the article. The corresponding author is in possession of this document.

Use of artificial intelligence for generating text. The authors declare that they have not used any type of generative artificial intelligence for the writing of this manuscript nor for the creation of images, graphics, tables, or their corresponding captions.

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