Temporal fossa aneurysmal bone cyst. A case report

Alexis Román Matus M.D.
Juan Daniel Naranjo Hernandez M.D.
Carlos Ivan Amezcua Equihua M.D.
Paola Ramirez Carrillo M.D.

Tijuana, Mexico

Case Report

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Introduction: The Aneurysmatic Bone Cyst (ABC) is benign like lesion, infrequent and non-neoplastic in its nature, it occurs more frequently in people below 20 years of age (60-80%). It is uncommon in children with less than 5 years old and adults after 30 years old. About 2% of ABC localize in neck and skull. Until this day, there are few cases reported about this pathology.

Clinical case: Is a 12-year-old male. There was no clinical history relevant for current condition, clinical presentation of 2 months of evolution, characterized by commissure deviation to the left side, ipsilateral facial palsy, dizziness, earache and left otorrhea for approximately 1 month of evolution. Image studies showed an image of a tumor lesion with well-defined edges, heterogeneous densities, hypo intense predominance, 4-5 cm in petrous portion of temporal bone, affecting cerebral parenchyma, non-visible perilesional brain edema on cerebral magnetic Resonance Imaging. Patient was operated with complete resection of tumor lesion. Postoperative evolution occurred with no incidents or accidents.

Conclusions: The ABC, are benign tumor lesions that present very low incidence in cranial region, even with lower incidence are those localized in middle cranial fossa, existing few cases reported on literature, until today. Surgical resection usually curative and effort must be realized to avoid additional damage to delicate structures around the petrous portion of the temporal bone.

Key words: Aneurysmal Bone Cyst, Facial paresis, Cranial base

Introduction

Aneurysmal Bone Cyst (ABC) is a benign lesion, infrequent and non-neoplastic in nature, occurring most frequently in patients under 20 years of age, in 60-80% (rare before the age of 5 years and after 30 years). [1]

Usually, metaphysis of long bones and spine are affected, only approximately 2% of OAQs occur in the skull and neck region. [2,3]
Radical surgery is the gold standard as a treatment for ABC, however, it is not achieved in all cases, due to the presence of extensive tumors in complicated anatomical structures. [3]

To date there are few publications on the subject, therefore, we present the case of a 12-year-old boy with a diagnosis of left middle cranial base ABC.

Case report

A 12-year-old adolescent patient was admitted to the pediatric emergency department of the Hospital Gineco Pediátrico y de Medicina Familiar No. 31 (HGP MF 31), with symptoms of 2 months of evolution, characterized by deviation of the labial commissure to the right side due to left hemifacial weakness, dizziness, otalgia and left otorrhea of 1 month of evolution. She has no history of relevance to the presented condition.

On physical examination, normal brain functions were found. On examination of cranial nerves, with hypoesthesia in the territories of the three branches of the trigeminal nerve. Hyperesthesia and pain on palpation of the temporomandibular joint on the left side. Left facial paresis grade III of House and Brackman. Left conductive hearing loss. Left otoscopy with stenosis of the external auditory canal with purulent secretion and erythema of the walls of the canal, without being able to observe the tympanic membrane. The rest of the cranial nerves were unaltered. Motor examination, with slight right body hemiparesis (4/5).

Sensory exploration without alterations as well as cerebellar exploration, and absence of atavistic and meningeal. A simple CT scan of the skull and ears showed a tumor of extra axial characteristics, left

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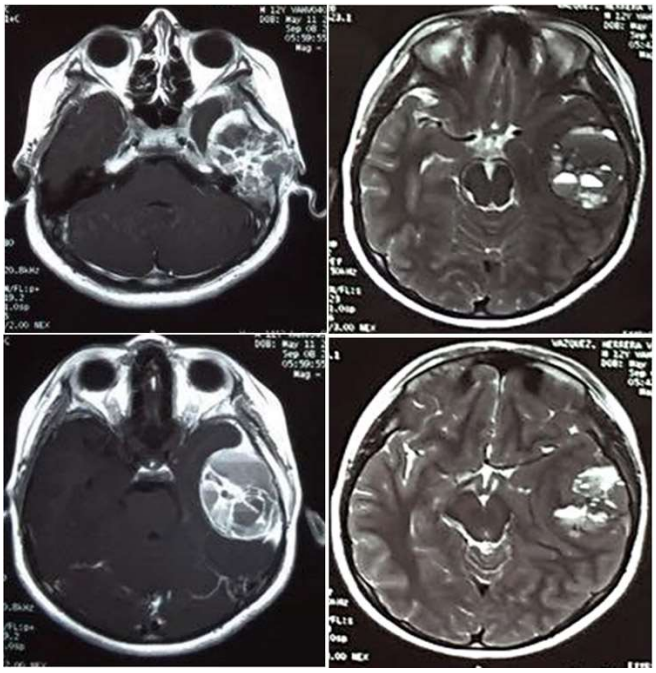

Figure 1. MRI of the brain showing a rounded image, with defined borders, 4-5 cm in the petrous portion of the temporal bone compressing cerebral parenchyma, heterogeneous, predominantly hypointense, without visible perilesional edema and with apparently

temporal, 45x32 mm anteroposterior, 41 mm transverse and 32 mm in larger diameters, with bony destruction of the left petromastoid region and temporal bone (temporal fossa), introduced into the external auditory canal (EAC) and left middle ear; lesion has calcifications inside conditioning mass effect on the temporal parenchyma and herniation of the uncus.

Simple and contrasted magnetic resonance imaging of the brain was performed (**Figure 1**).

Laboratory results with complete blood biometry and blood chemistry showed no alterations.

Based on the clinical and radiological findings, the diagnosis of ABC was considered, and it was decided to perform surgery.

Surgical intervention was performed, with tumor resection plus exploration of the external auditory canal and middle ear, in conjunction with pediatric neurosurgery and otorhinolaryngology. During the intervention a mixed tumor was observed at the base of the skull formed by solid portions circumscribing cystic areas with dark liquid material inside, it presented strong adherence to the dura mater and middle fossa, during the resection the dura mater was not injured and the lesion was completely resected in a fractional way. Curettage of the middle cranial fossa was performed to completely remove the residual tumor.

Subsequently, the middle ear was explored, where the ossicles were splinted. Otoscopy showed the decompression of the CAE and the integrity of the tympanic membrane. There were no incidents and/or accidents during the surgical-anesthetic procedure.

The histopathological report referred to a lesion composed of cystic spaces with abundant erythrocytes inside and separated by fibrous tissue, which was compatible with ABC. Histologic diagnosis corresponds to ABC.

After surgery, there was evident improvement of the hypoacusis and facial paresis, for which he was discharged.

In the postoperative follow-up after 8 months, a remarkable improvement is observed with absence of neurological signs or symptoms, and a control cranial CT scan is pending.

Discussion

Aneurysmal bone cysts were first described in 1893 by Van Arsdale, who called them "ossifying hematomas of the humerus". In 1940 Ewing used the term "aneurysmal" to describe these lesions. Jafeé and Lichtenstein first coined the term "aneurysmal cyst" in 1942. In 1950, they modified this term to "aneurysmal bone cyst". [1]

They are tumor-like expansions of the diploic space, which distort and diminish the cortical layer of the bone, are filled with blood and delineated by connective tissue composed of trabecular bone and giant cells. [4]

According to the definition of the World Health Organization, which defines them as a benign, expansive and osteolytic tumor, consisting of hematic cysts limited by connective tissue septa, bone trabeculae and osteoclast-like giant cells. [5]

Its age incidence is in people younger than 20 years, about 80% of cases, and there seems to be a 2:1 prevalence predominantly in women. 4,5 Only approximately 2-6% of ABC belong to the cranial bones, and their presentation is usually as a painful and progressively expanding mass. Two thirds of cranial ABC are usually located in the maxilla and mandible, leaving the remaining percentage to frontal,

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parietal, temporal and occipital bone areas, where cases have also been reported. [6,7]

Its etiopathogenesis is not yet known, however, the hypothesis of greater acceptance so far, speaks of an alteration in the local circulation, generating thrombosis of an aneurysmal vein or formation of arteriovenous fistulas, leading to increased venous pressure. Biesecker et al, in 1970, suggests that they are the origin of precursor lesions, probably arteriovenous bone malformations, which generate by hemodynamic forces, a reactive lesion in the bone. [8]

In cytogenetic studies, it has been observed that chromosomal rearrangements 17p11-13 or 16q22 are involved in the occurrence of ABC, and it appears that these rearrangements are always present, regardless of subtype or location.[9]

Its etiology has been associated with cranioencephalic trauma, however, this cause and effect relationship is not yet fully accepted, and theories that may reject it are currently being debated. [10] In this case there was no history of previous cranioencephalic trauma.

The clinical data, depending on the affected site, are usually local swelling, cranial neuropathies, proptosis and symptoms of intracranial hypertension. In this case, the patient presented with involvement of the V, VII and VIII pairs on the left side, as well as local swelling. [11]

Its appearance in imaging studies (cranial computed tomography and magnetic resonance imaging) has been described as an image of soap bubbles or explosion. The main characteristics are growth of the diploic space, visualization of an encapsulation with well delimited peripheral sclerosis, this indicates slow growth and predominantly benign character,[12] similar to what was found in this case.

Three evolutionary stages have been identified according to their radiological appearance: an osteolytic or early phase, a second phase in which a peripheral halo of greater density appears and is associated with internal walls and, finally, a calcified or late phase. [13]

Cranial tomography may show a multiloculated lesion, which may be enhanced after contrast administration.[14] Brain magnetic resonance imaging is the study of choice for diagnosis and may show multiple fluid levels within the cysts, representing sedimentation of red blood cells within the cavities filled with serohematic content. In T1 it is usually seen as a hypointense lesion, while in T2 it can be seen as a hyperintense lesion,[15] such images similar to those presented in the studies of this patient.

The differential diagnosis is based on imaging findings and can be compared with benign lesions such as fibrous dysplasia, giant cell tumor, non-ossifying fibroma, unicameral bone cyst and histiocytomas, as well as malignant lesions within which we find osteosarcoma, fibrosarcoma and telangiectatic osteosarcoma.[16]

Treatment is usually surgical, the goal being complete resection of the cyst, followed by primary reconstruction of the area, coinciding with what was performed on this patient. If the tumor is not completely resected, curettage of the abnormal debris in the bone is considerable, combined with pre- and postoperative adjuvant treatments. [13,16]

Radiotherapy should be considered for incomplete resections or deep lesions of dural involvement; however, cases of sarcomatous transformation have been reported after radiotherapy, so it is contraindicated in patients with associated fibrodysplasia.[17]

To date there is a very small number of patients with postoperative follow-up reported in the literature, so it is not possible to establish an exact percentage with respect to recurrences, however, it is currently recommended to follow the patient for a minimum period of 5 years, mainly in pediatric patients, who should be followed up until puberty. [18]

Conclusions

ABC are benign tumor lesions that present a very low incidence in the cranial region, even more the cases located in the middle cranial fossa are even rarer, and there are few reports found in the literature. The neurological manifestations expected for a ABC of this location are mainly affections of the cranial nerves V, VII and VIII, in addition to favoring recurrent otitis media. Resection is usually curative and efforts should be made to avoid further damage to the delicate structures that have their seat in the petrous portion of the temporal bone.

After an extensive literature search, this may be the first case of a temporal fossa ABC reported in the national literature.

Conflicts of interests

There are no potential conflicts of interest for any of the authors in this scientific report.

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Alexis Román Matus
Department of General Surgery
Instituto Mexicano del Seguro Social
Hospital General Regonal No. 1
Tijuana, Mexico
alexisrm188@gmail.com

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