Intradiploic Epidermoid Cyst of the Posterior Fossa – Case Report and Review of the Literature

Cisto epidermoide intradiploico da fossa posterior – Caso clinico e revisão da literatura

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Abstract

Intracranial epidermoid cysts represent ~ 0.2 to 1.8% of all intracranial tumors. These tumors are most frequently encountered in the cerebellar pontine angle or in the parasellar region. Rarely, they arise from the cranial diploe, being able to affect every flat bone of the cranium.

We report a case of a 63-year-old male who presented with progressively worsening headache and vertigo with 6 months of evolution. Neuroimaging identified a probable occipital intradiploic epidermoid cyst with mass effect on the cerebellar hemispheres. This lesion was approached using a suboccipital craniotomy, followed by total resection of the tumor and cranioplasty with titanium plate placement. The histological evaluation confirmed the diagnosis of intraosseous epidermoid cyst. The patient had a successful recovery, without complications or neurologic dysfunction.

Being benign lesions, commonly remaining asymptomatic and rarely presenting as a bony lump in the skull bone, it could be straightforward to assume a conservative management with planned follow-up. On the other side, a more aggressive strategy with surgical excision has been advocated, especially in lesions that tend to enlarge and erode the cranial bone with possible consequent epidural extension and mass effect symptoms. A preoperative diagnosis is extremely helpful in proper surgical planning. Diffusion weighted imaging facilitates a straightforward diagnosis.

As was observed in our case, the largest reviews on intradiploic epidermoids available in the literature mostly demonstrated a benign clinical course. However, malignant transformation can occur. Some patients develop permanent neurologic deficits from mass effect or tumor infiltration. However, surgical approach of the tumor is curative in most cases. Nonetheless, from our experience, it is important to maintain clinical and imaging follow-up with regular monitoring to prevent possible tumor recurrences.

Keywords

- epidermoid cyst
- craniotomy
- cranioplasty
- titanium mesh

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Resumo

Os cistos epidermoides intracranianos representam cerca de 0,2 a 1,8% de todos os tumores intracranianos. Esses tumores são mais frequentemente encontrados no ângulo pontocerebeloso ou na região parasselar. Raramente surgem na diploe, no entanto podem afetar todos os ossos do crânio. Relatamos o caso de um homem de 63 anos que apresentou agravamento progressivo da cefaleia e vertigem e desequilíbrio com 6 meses de evolução. O estudo de imagem realizado identificou um provável cisto epidermoide intradiploico occipital com efeito de massa nos hemisférios cerebelosos. A lesão foi abordada por craniotomia suboccipital, seguida de ressecção total do tumor e cranioplastia com colocação de placa de titânio. A avaliação histológica confirmou o diagnóstico de cisto epidermoide intraósseo. O doente teve uma recuperação bem sucedida, sem complicações ou disfunção neurológica. Sendo lesões benignas, que geralmente permanecem assintomáticas, pode-se assumir um tratamento conservador com seguimento clínico. Por outro lado, uma estratégia mais agressiva com excisão cirúrgica tem sido preconizada, principalmente em lesões que tendem a aumentar e invadir as estruturas ósseas cranianas com possível extensão peridural e sintomas causados pelo efeito de massa. Um diagnóstico pré-operatório é extremamente útil no planeamento cirúrgico adequado. A imagem ponderada por difusão facilita um diagnóstico direto. Como observado no nosso caso, as maiores revisões sobre epidermoides intradiploicos disponíveis na literatura demonstraram, na sua maioria, um curso clínico benigno. No entanto, a transformação maligna pode ocorrer. Alguns doentes desenvolvem déficits neurológicos permanentes por efeito de massa ou infiltração tumoral. A abordagem cirúrgica do tumor é curativa na maioria dos casos. Consideramos importante manter o seguimento clínico e imagiológico regular para prevenir possíveis recidivas tumorais.

Palavras-chave

- ► cisto epidermoide
- ► craniotomia
- ► cranioplastia
- ► rede de titânio

Introduction

First described in 1838 by Müller, intracranial epidermoid cysts represent ~ 0.2 to 1.8% of all intracranial tumors.[3] Most of these tumors arise from the cerebellopontine angle or from the parasellar region. The diploe of the skull is an uncommon location for the presentation of an epidermoid cyst, although this type of lesion can affect every flat bone of the cranium. Primary intradiploic epidermoid cysts of the central nervous system (CNS) are considered rare, with a few more than 200 cases reported so far.[11] The nature of theses lesions is primarily congenital, presumed to occur due to inclusion of ectodermal cells in the bony tissue during neural tube closure in the embryonic life.

We share our experience on a case of primary intradiploic epidermoid cyst in the occipital bone with compression of the dura in the posterior fossa and review the literature on intradiploic epidermoid tumors.

Case Presentation

A 63-year-old male presented to our department with progressively worsening headache, intermittent dizziness, and imbalance, with 6 months of evolution. The vital signs were stable. Personal medical history included arterial hypertension on pharmacological control with aldosterone receptor antagonist and calcium channel blocker, benign prostate hyperplasia, and peripheral venous insufficiency. No relevant familial history was found, namely regarding neurocutaneous disorders.

Physical examination revealed an elevation of the occipital cranial bone region, hard and fixed on palpation, with no obvious redness, swelling or tenderness of the superficial scalp. A cranial computed tomography (CT) scan (►**Fig. 1**) showed a paramedian occipital osteolytic intradiploic expansive lesion with left lateral extension, with smooth bone remodeling and focal bony defects.

Complementary brain magnetic resonance imaging (MRI) was performed (►**Fig. 2**), documenting the large, well-delimited heterogenous intradiploic lesion, mostly hyperintense on T2/FLAIR and hypointense on T1, with no contrast enhancement and demonstrating a characteristic restricted diffusion pattern, suggestive of intradiploic epidermoid cyst. The lesion measured ~ $72 \times 30 \times 40$ mm (transverse, anteroposterior, and rostrocaudal axis, respectively). Intracranial extradural extension and mass effect on the cerebellar parenchyma were also evident.

Surgery was performed under general anesthesia with the patient in ventral decubitus, with the head fixed in the Mayfield head. A midline incision was performed for posterior fossa craniectomy with bilateral enlargement. The tumor eroded both the external and internal laminae of the cranial bone. It was solid, soft, tender, pale yellow, with poor blood

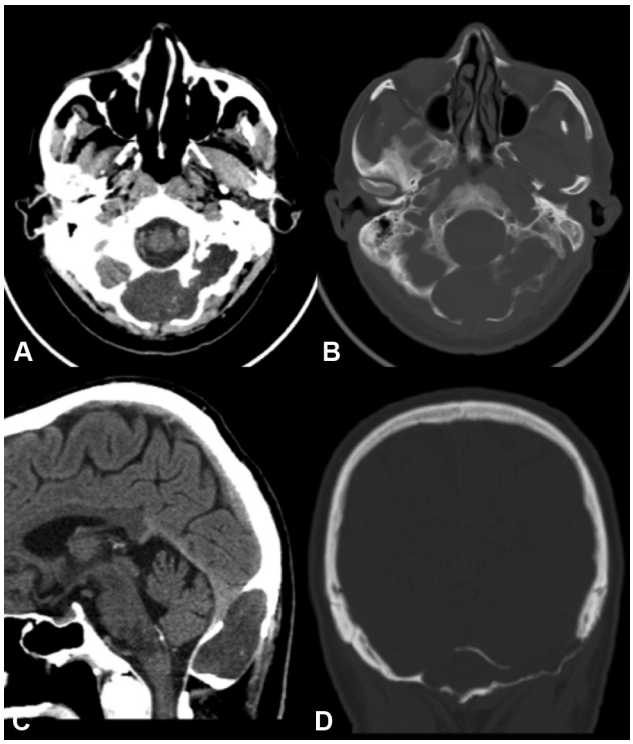

Fig. 1 Head computed tomography scan showing an occipital osteolytic hypodense intradiploic expansive lesion. Note the widening of the diploic space with smooth bone remodeling, thinning of the inner and outer tables, and associated bony defects.

supply, and presented clear margins. There was a slight adhesion to the underlying dura mater, still intact, protecting the underlying parenchyma of the cerebellar lobes.

During lesion resection, hemorrhage was satisfyingly controlled. We performed cranioplasty with a titanium mesh with good results, without complications up to follow-up data. A CT scan performed 1 day after the operation (►**Fig. 3**) showed complete resection of the lesion and absence of immediate complications.

The postoperative histological diagnosis reported epidermoid cyst, (►**Fig. 4**) the first with bone and lining of the cyst and the second with the lining (keratinized stratified squamous epithelium) and the content (keratin).

The patient was discharged 3 days after the operation, with no record of complications or neurological deficits. Headache and vertigo progressively improved during the period of hospitalization. At the 1-month follow-up consultation, the patient was asymptomatic.

A 6-month follow-up brain MRI (►**Fig. 5**) was performed, documenting lack of recurrence.

Discussion

Representing 5% of all intracranial epidermoid cysts, intradiploic epidermoid cysts are considered rare.[1] The nature of these lesions is mostly congenital, arising from sequestration of ectodermal cells, although acquired etiology, namely post-traumatic, has been reported.[2] The slow growth rate of the

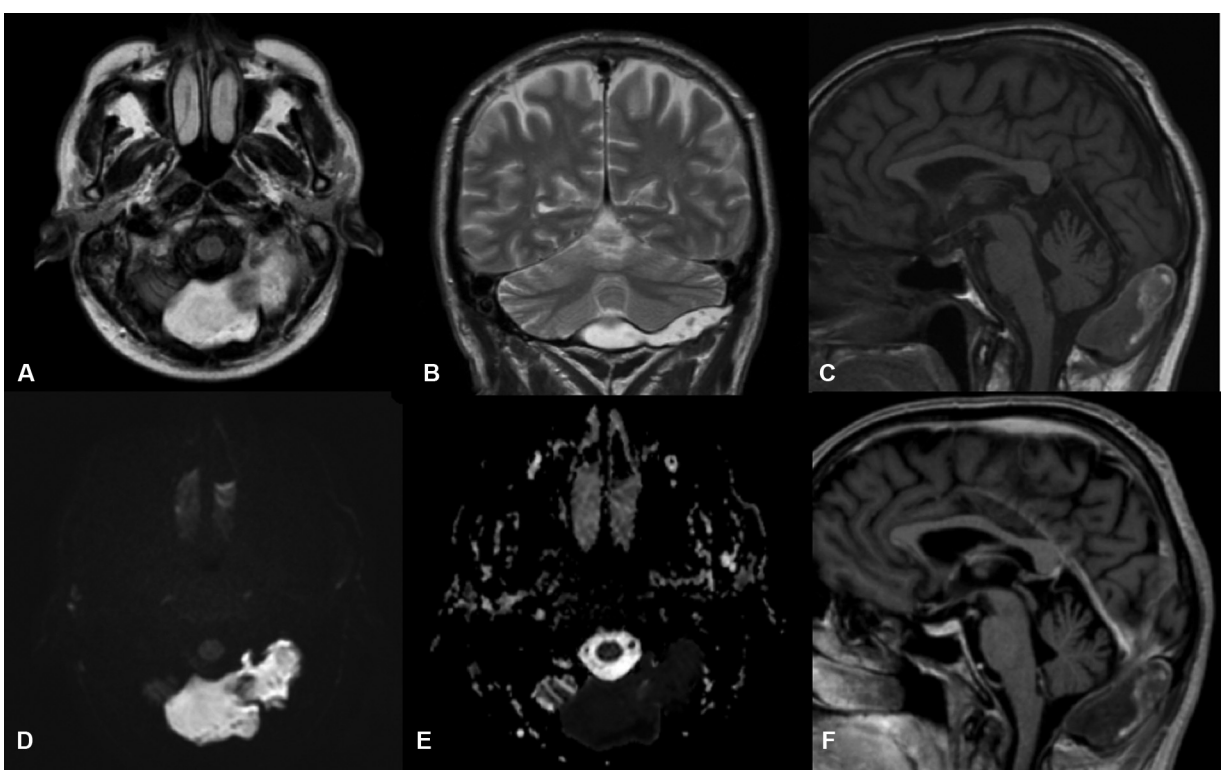

Fig. 2 Brain magnetic resonance imaging A. Axial FLAIR sequence; B. Coronal T2 weighted sequence; C. Sagittal T1 weighted sequence; D. DWI; E. ADC map. F. Sagittal T1 contrast enhanced weighted sequence showing the well-delimited intradiploic lesion in the posterior fossa, with intracranial extradural extension molding the cerebellar parenchyma. The lesion presents T2/FLAIR hyperintensity (images A and B), heterogeneous signal on T1, mostly hypointense with small areas of hyperintensity arrow head (image C), and no contrast enhancement (image F). Finally, note the characteristic restricted diffusion pattern (images D and E). This is an intradiploic epidermoid cyst.

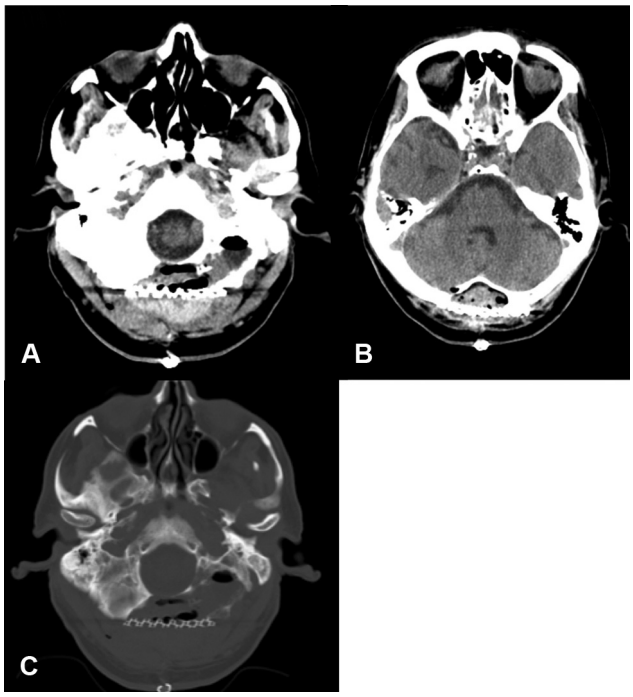

Fig. 3 1-day postoperative head computed tomography scan showing no major complications. In the bone window, we can observe the correct placement of the cranioplasty titanium mesh.

cyst causes expansion of the inner and outer tables of the skull and, through continuous enlargement, it may achieve gigantic dimensions, originating exuberant images, such as in our case. Giant intradiploic epidermoids, however, are exceptionally rare.

A preoperative diagnosis is extremely helpful in proper surgical planning. At times posing as a diagnostic challenge on neuroimaging, current developments in MRI sequences have assisted in obviating this problem[11].

On CT imaging, intradiploic epidermoids typically present as nonenhancing hypodense osteolytic lesions with smooth margins and associated bony defects, causing widening of the diploic space and preferential erosion of the inner table. Differential diagnoses of lytic bone defects with expansion of the diploic space include dermoid cyst, intraosseous hemangioma, eosinophilic granuloma, and fibrous dysplasia. Dermoid cysts usually present in childhood and are located in the midline along suture lines. Intraosseous hemangiomas show characteristic intralesional spicules or trabeculae and homogenous enhancement after contrast administration. Differently from epidermoids, they generally erode the outer table, with a relative sparing of the inner table. Eosinophilic granulomas have a characteristic appearance in the skull as punched-out lytic lesions without a sclerotic rim. Sometimes, a double contour or beveled edge appearance may be seen due to asymmetrical involvement of the inner versus the outer table (the classic "hole within a hole sign"). If there is a small sequestrum of devascularized bone, this will result in a typical "bull's eye" or "button sequestrum" appearance. Fibrous dysplasia lesions generally have more ill-defined margins. There is expansion of the diploic space, but usually both tables are intact, with no bony defects. There is loss of

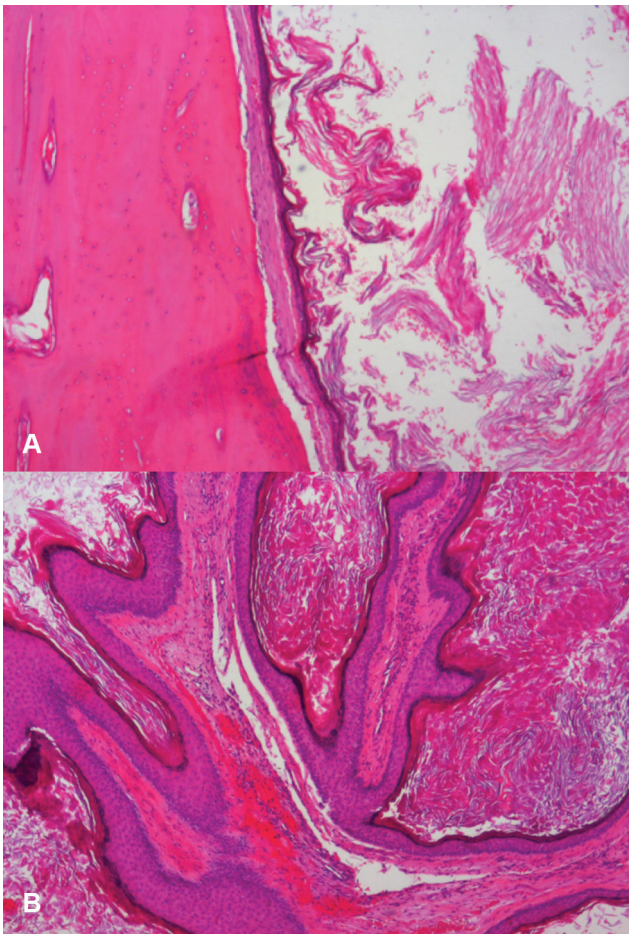

Fig. 4 A. Histologic examination, corresponding to coating (keratinized stratified pavement epithelium) and the content (keratin). B. Histologic examination, corresponding to bone and cyst lining.

the normal corticomedullary differentiation, with a typical homogeneous ground glass appearance.[3]

On MRI imaging, we usually find a mildly T1 hyperintense lesion, with iso/hyperintensity on both T2 and FLAIR images, restricted diffusion pattern, and no contrast enhancement. The differential diagnosis does not pose much of a challenge. Nonetheless, rare, atypical "white epidermoids" with high protein content that show reversed signal intensity on MRI images and have no restricted diffusion have been reported.[1,4]

A review of 8 cases of infratentorial giant intradiploic epidermoids found a mean age at the onset of symptoms of 55 years old and a predilection for males (male:female ratio 8:0).[2] Reported cases of giant intradiploic epidermoids usually course with cranial hypertension symptoms, probably explained by compression or thrombosis of the venous dural sinuses. Other expected signs and symptoms include headaches, which are the most common, cranial nerve deficits, cerebellar symptoms, and seizures. Several case reports found painless subcutaneous scalp swelling to be common. Our patient presented with occipital localized headache, dizziness, and loss of balance, explained by the mass effect on the left cerebellar hemisphere. Despite clear intradiploic expansion, thinning and disruption of both tables, the

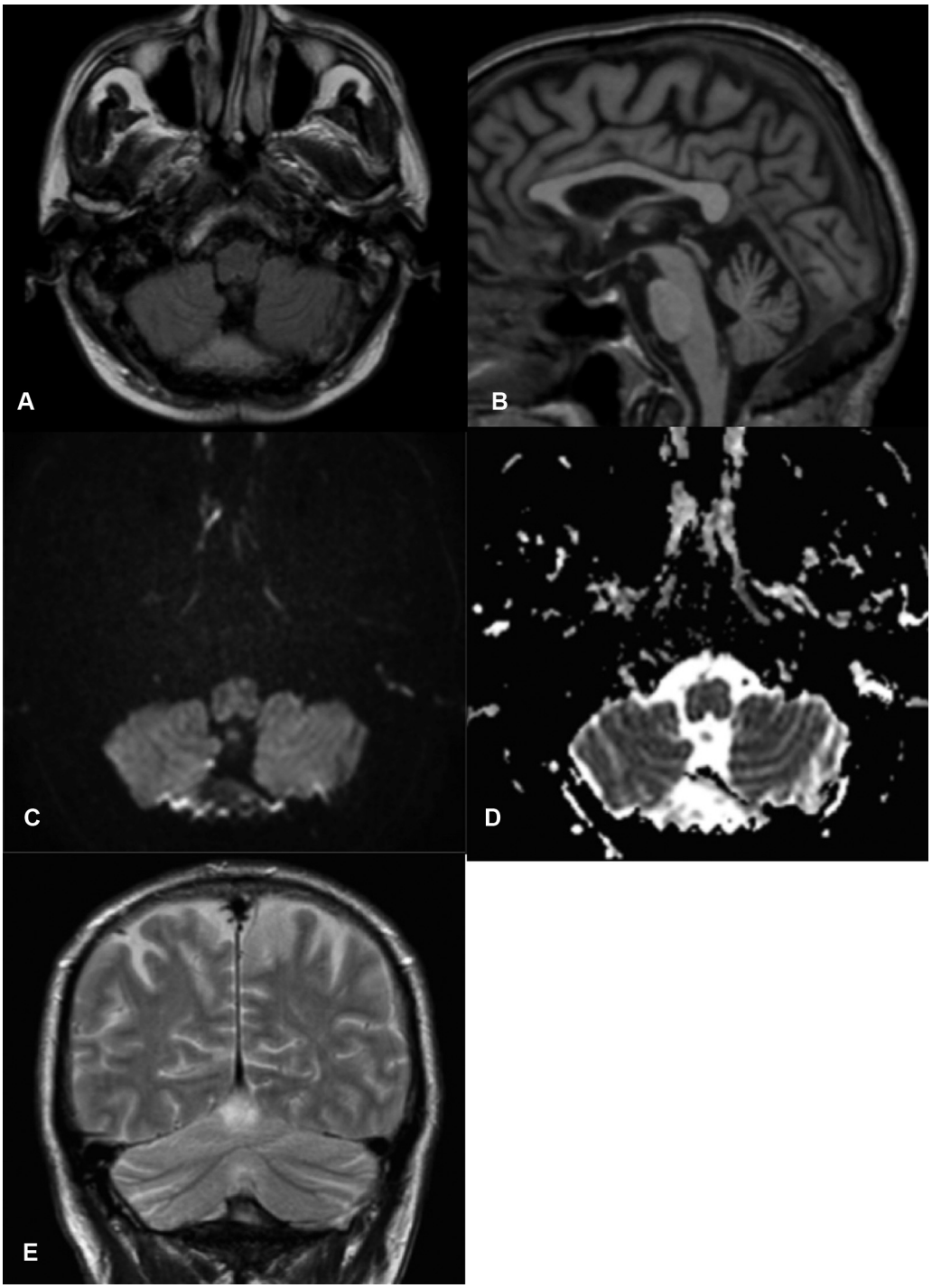

Fig. 5 6-month follow-up brain magnetic resonance imaging documenting lack of recurrence.

epidermoid mostly insinuated intracranially and no scalp swelling was noted. Despite achieving an exuberant size with intracranial expression, there was no significant affection of the cerebrospinal fluid pathways or venous system. These aspects are characteristic of our case, making it unique. Fortunately, there were no complications associated with the lesion itself, since, in particular, the rupture of the cyst, a possible complication, that can may worsen the case's evolution.

Spontaneous cyst rupture may complicate intradiploic epidermoid tumors, producing granulomatous meningitis.[4] Although very rare, malignization might occur, especially in cases of recurrence due to incomplete resection.[3,5,6]

Surgery with total removal of the epidermoid cyst is advocated.[7] Despite its size, total removal of the lesion along with its capsule leads to definite treatment with very low morbidity and mortality.[2,8–12] Conservative resection can also be considered given the slow growing nature of this tumor, especially when its localization near crucial neurovascular structures poses a threat. Excision of the eroded bone and cranioplasty, as performed in our case, may be necessary. As in other pathologies, the advantage of surgical

excision is the possibility of obtaining a sample to confirm the diagnosis.

Intraosseous epidermoid cysts must be included in the differential diagnosis for osteolytic of calvarian and skull lesions.[13]

Conclusions

Intradiploic epidermoid cysts are rare congenital lesions. They present a characteristic dormant clinical course. The slow growth rate of the cyst causes expansion of the inner and outer tables of the skull, being able to achieve gigantic dimensions and originating exuberant images, such as in our case. Neuroimaging shows typical findings, particularly the characteristic restriction diffusion pattern on diffusion weighted imaging (DWI), providing a straightforward diagnosis. Imaging is also extremely helpful in surgical planning.

Despite their benign histology, epidermoid tumors may enlarge and cause lytic destruction of the cranial bone, sometimes invading the dura mater and underlying structures. Fortunately, this was not present in our case. Surgical resection should be considered for selected patients as early as possible to avoid progression-related deficits and more extensive surgery with potential complications or sequelae. Surgical management with noncomplicated total removal of these lesions is the treatment of choice, which also establishes the diagnosis and can provide a good long-term outcome. Nonetheless, from our experience, it is important to maintain clinical and imaging follow-up with regular monitoring to prevent possible tumor recurrences.

Conflict of Interests

The authors have no conflict of interests to declare.

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