Intraorbital Expansion of an Intradiploic Frontal Epidermoid Cyst: A Case Report and Short Review of the Literature

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Patient: Male, 58-year-old
Final Diagnosis: Epidermoid cyst
Symptoms: Exophthalmos
Medication: —
Clinical Procedure: —
Specialty: Neurosurgery

Objective: Rare disease
Background: Calvarial epidermoid cysts (EC) are encased remnants of ectoderm at the third week of gestation. There are also reports which consider them sequelae of head trauma. They are benign lesions. As they develop, they exert a mass effect to adjacent anatomical structures.

Case Report: We report the case of a 58-year-old male patient with left-sided exophthalmos. His radiologic examinations depicted an oval cystic lesion (7 × 5 × 5.5 cm) arising from the left frontal bone and abutting the ipsilateral orbital roof.
Our patient underwent a total extirpation of the lesion through a frontal craniectomy. Cranioplasty was then performed with a Porex® graft. The pearl-hued lamellae of the lesion macroscopically resembled keratin tissue.
Histopathological findings supported the diagnosis of an epidermoid cyst. Postoperatively, our patient had no neurologic deficits and a computed tomography scan showed no residual effects.

Conclusions: Large calvarial EC with intraorbital expansion in adults are rare clinical entities. Gross total resection with the infiltrated bone and cranioplasty is the treatment of choice, which also establishes the diagnosis.

Keywords: Exophthalmos • Methylmethacrylate • Mucoepidermoid Tumor

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Background

Intradiploic epidermoid cysts (EC) are benign congenital lesions. They occur when the ectodermal enfolding takes place prior to the 21st day of embryogenesis. The intradiploic cysts arise from these enclosed remnants after the neural tube joins. The main components are squamous epithelial tissue, with keratin and cholesterol. Paradoxically, they also occur in aging adults [1]. These lesions warrant prompt extirpation due to their tendency to grow, even in cases where no significant neurological symptomatology exists.

Case Report

A 58-year-old white man was electively admitted due to deteriorating exophthalmos of his left orbit. He reported that this situation had been developing during the last 3 months. His medical history included arterial hypertension and tobacco smoking. The MRI scan depicted an oval cystic lesion (7×5×5.5 cm) arising from his frontal bone and abutting the left orbital roof. It showed low intensity on T1 sequence and hyperintensity on T2 sequence (Figure 1). In diffusion-weighted imaging (DWI), restriction was prominent (Figure 2). In addition, on a skull computed tomography (CT) scan, the lesion had caused destruction of the inner table, with calcified...

Figure 1. T1 sequences after contrast medium administration. (A) Extradural lesion of frontal region with midline shift and no contrast enhancement. (B) Upper orbital wall emaciation, compression of periorbita and exophthalmos. Red arrows show the tumor periphery.

Figure 2. Typical diffusion restriction in diffusion-weighted sequencing (DWI) of the cystic tumor.
rims (Figure 3). The patient also had impaired upward gaze ipsilaterally due to immediate mechanical hindering. His overall neurological status was intact.

Surgery

Our patient was placed in supine position with his head resting on a horseshoe. Using a bicoronal incision, cutaneous and muscular flaps were retracted. Afterwards, the left frontal bone was exposed, revealing a soft, protruding, encapsulated and well-demarcated mass comprising the superficial part of the intracranial lesion. The lesion was initially resected via curettage. In addition, a left frontal craniotomy was performed around the lesion (Figure 4). The orbital roof was thinned due to the tumor, whereas periorbital tissue was not infiltrated. The pearl-hued lamellae macroscopically resembled keratin tissue. Thus, the suspicion of an epidermoid cyst was raised. The rest of the tumor was also excised, involving its capsule adherent to the dura. No remarkable hemorrhaging ensued during the excision.

The periorbita was then covered with a TachoSil® patch (human fibrinogen 5.5 mg and human thrombin 2.0 IU/cm²) and artificial meninx. The frontal bone defect was replaced by a Porex® graft (a porous polyethylene implant) manually formed and fixed using screws and plates (Figure 5). The wound was sutured under suction drainage.
Figure 6. (A) Gross findings revealed soft white keratin contents. (B) Histological examination demonstrated abundant keratin flakes and (C, D) heavily keratinized squamous epithelium (bold arrows) adjacent to osseous tissue (star). The epithelial cells were positive for keratin 5/6 (E) and p63 (F). (B: HE ×100; C: HE ×400; D: HE ×100; E: IHC ×400; F: IHC ×400).

Figure 7. (A) Postoperative CT control. Green arrow shows the anatomic region of the resected tumor at the convexity. (B) Yellow arrow shows the tumor-free intraorbital space.
Histopathological findings

Gross findings resembled soft white keratin contents (Figure 6A). Microscopically, representative sections of the specimens showed numerous keratin flakes and parts of osseous tissue that were adjacent to heavily keratinized stratified squamous epithelium (Figure 6B-6D). The granular layer of the epithelium was easily identifiable. There were no skin appendages (eg, hair follicles, sebaceous or eccrine glands). Immunohistochemically, the epithelium showed positivity to keratin 5/6 and p63 (Figure 6E, 6F).

The morphological and immunohistochemical findings supported the diagnosis of epidermoid (epidermal) cyst.

During the postoperative period, our patient developed no neurologic deficits. His wound healed by primary intention and the postoperative CT confirmed the total resection (Figure 7A, 7B).

Discussion

Calvarial epidermoid cysts comprise 1% of all intracranial tumors [2]. Their classical origin is attributed to the encased remnants of ectoderm in the third week of embryogenesis within the mesoderm of bone tissue [3]. The etiology of these lesions, though, has been advocated by some authors to be acquired, sometimes as a sequel of penetrating skull trauma and invasion of cutaneous tissues within the diploë [4]. In the history of our patient, no report of head trauma was referred. The course was a typical one: an indolent, slow-growing mass of the frontal region in close proximity to the orbital walls [4]. In 46% of intradiploic epidermoid cysts, both tables of the cranium are involved and they usually occur in adults [3].

With aging, some factors related to connective tissue are expressed at an increased rate. Matrix metalloproteases, connective tissue growth factor, bone morphogenetic protein, and inflammatory cytokines are included in that group [5]. Our patient was age 58 years when the cyst manifested, corroborating the aforementioned conclusions.

Akar et al presented a case series of 28 patients in whom the most frequent location was the cerebellopontine angle, and only 1 was intradiploic within the temporal bone [6]. The intradiploic extradural ECs found in the cranial vault erode the inner cortex and thin the outer cortex [7].

Intradiploic ECs show as hypodense and non-enhancing tumors on CT scans, similar to the fat density [8]. MR images reveal heterogeneous hypointensity on T1 sequences and heterogeneous hyperintensity on T2 fluid-attenuated inversion recovery (FLAIR) signal sequences. They are usually non-enhancing, with occasional minimal rim enhancement. Hyperdensity usually implies an intracystic hemorrhage. Diffusion-weighted imaging (DWI) is the proper sequence for use in diagnosing ECs, showing restricted diffusion with higher signal intensity than that of cerebrospinal fluid (CSF) on DWI [8].

Differential diagnosis first of all involves dermoid and sebaceous cysts [9]. Dermoid cysts are often associated with the suture lines. They also commonly appear periorbitally involving the midline. Their incidence is highest in childhood and they often contain fat and cutaneous appendages. Sebaceous cysts have fat density due to sebum, and are a palpable, non-fixed part of the cutis.

Apart from that, eosinophilic granuloma may be also misdiagnosed as an epidermoid cyst. However, they are more homogeneous, succulent in palpation, and have a square-edge appearance without calcifications on CT scans [9].

In addition, skull vault hemangiomas have a geometric pattern of expansion like honeycomb or radiating sunburst. Giant cell reparative granulomas present with lytic osseous erosion lacking calcifications. On MRI, they are slightly hypointense, with vivid gadolinium enhancement [9].

Plasmacytomas are a malignancy of plasma cells. They are either solitary or in the context of systemic multiple myeloma. They are hypodense on non-contrast CT and hyperenhancing on contrast CT. They lack a sclerotic rim, but they may include peripheral osseous fragments [9].

Calvarial metastases show a variable appearance. They are also either single or multiple masses with symmetrical or asymmetrical periphery. They are hyperintense tumors on CT owing to increased neoangiogenesis [9].

A literature review by Arko et al concluded that the commonest locations of the intradiploic ECs are on the frontal, parietal, and occipital bones, with approximately equal involvement [10], suggesting that they are associated with skull fractures, most often occurring in the parietal bone, followed by the temporal, occipital, and frontal bones. However, additional mechanisms of pathogenesis exist, mainly congenital causality of cyst formation. Parallel to that, they postulated a 2-peak model of manifestation, early (within the third decade) and again later (within the sixth decade) in life, supporting the 2 coexistent mechanisms. They also found they are mostly benign lesions and emphasized the role of capsule excision as the most significant factor preventing recurrence [10].

Interestingly, only a few cases with concomitant orbital expansion have been so far reported.
Ormond et al reported the case of a 67-year-old man with left-sided headache and ocular pain. Radiological examinations were indicative of a hemorrhagic tumor causing osteolysis of the wings of the sphenoid bone, shifting the frontal and temporal lobes and compressing the globe as well. He underwent a total surgical excision followed by cranioplasty and orbital reconstruction. The patient achieved remission, with no residual symptoms [11].

Samdani et al reported a female patient with intradiploic EC of her left frontal bone that developed after childhood trauma. The cyst led to osteolysis of the frontal sinus, culminating in exophthalmos and restricted oculomotion with frontal headache. Total excision of the cyst and its capsule was performed. The periorbita of the ipsilateral globe was not infiltrated. Costal grafts were used for the cranioplasty, along with titanium screws [12].

Sauaia Filho et al presented the clinical course of a 23-year-old woman with headache, ocular pain, exophthalmos and impaired oculomotor [13]. On CT scan, a lesion located in her left frontal sinus was noticed. The latter also had signs of osteosclerosis and extended into the ethmoidal cells and the orbital roof. They performed a total resection followed by calvarial reconstruction. Histopathology confirmed the diagnosis of an EC.

Although there is a consensus concerning the extirpation of the capsule, there is still exists disagreement regarding the extent of resection of the adjacent osseous tissue [3]. Recurrence rates can be high and have been reported to range from 8% to 25% [14,15]. Complications are rare and include infection and abscess formation, focal deficits due to mass effect, hemorrhage, and malignant transformation [16].

Conclusions

We presented an unusual case of intracranial intraorbital expansion of a large frontally located EC, which is one of the largest in the literature. These benign cysts should be included in the differential diagnosis of osseous lesions in adults. Gross total resection along with the capsule is the criterion standard treatment and establishes the diagnosis. Surgeon should not be misled by patient age and the lesion’s inconspicuous course.

Declaration of Figures Authenticity

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.

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