We report a case of an intradiploic epidermoid cyst of the lateral orbital wall that resulted in proptosis and downward displacement of the left eye. Computed tomography and magnetic resonance imaging revealed a bone-destroying mass involving the frontal, sphenoid, and zygomatic bones that extended into the orbit, temporalis, and anterior cranial fossa. Complete surgical removal was performed through a lateral orbitotomy. The lateral wall was reconstructed with a porous polyethylene sheet. The cyst has not recurred after 18 months of follow-up.

Dermoid and epidermoid cysts are among the most common orbital tumors, comprising 3% to 9% of all orbital masses; diagnosis usually occurs during infancy or early childhood. These tumors are choristomas that arise from epidermal tissues trapped along bony suture lines during embryologic development and more rarely after trauma. Epidermoid cysts account for less than 1% of cranial tumors; 25% are intradiploic and 75% are intradural. Fewer than 20 well-documented cases of intradiploic epidermoid cysts of the bony orbit have been reported in the literature and the involvement of multiple orbital bones is exceptional.

We report a case of a giant epidermoid cyst involving 3 orbital bones (sphenoid, zygomatic, and frontal) that extended into the orbit, temporalis, and anterior cranial fossa. The cyst manifested clinically as progressive exophthalmos.

A 22-year-old healthy woman was referred to our clinic with a 6-month history of progressive downward displacement of her left eye (Figure 1). The visual acuity was 20/20 bilaterally and the intraocular pressure was 16 mm Hg bilaterally. A left 3-mm proptosis and a 7° hypotrophy that did not induce diplopia also were noted. Slitlamp and fundus examination findings were normal. The workup included computed tomography (CT) and magnetic resonance imaging (MRI). The CT scan revealed a bone-destroying mass involving the lateral wall and orbital roof. The mass was isodense with the brain parenchyma, had well-defined sclerotic borders, and had invaded the orbital cavity and temporalis fossa (Figure 2). No enhancement with intravenous contrast was observed. The MRI scan showed a relatively hypointense signal on T1-weighted images and hyperintense signal on T2-weighted images. Coronal sections showed invasion into the anterior cranial fossa without affecting the brain parenchyma (Figure 3). A biopsy of the mass was performed through a lateral orbitotomy. Histopathologic study revealed abundant degenerated keratin debris and occasional fragments of a stratified and keratinized epithelium without skin appendages, which led to the diagnosis of epidermoid cyst (Figure 4). The same surgical approach was used to completely excise the orbital mass, but this time a bone flap was obtained. The lesion was removed piece-meal with extensive dissection, excision, and profuse irrigation to ensure complete removal of the cyst. A large defect in the lateral wall was repaired with a 1.5-mm
porous polyethylene sheet (Medpor; Porex Surgical Inc, College Park, Ga). Exhaustive histopathologic study of serial sections from all the excised fragments disclosed the same findings described on the biopsy specimen. Since no skin appendages were detected, the diagnosis of epidermoid cyst was confirmed. Eight months after surgery, the aesthetic results were satisfactory (Figure 5) and no recurrence was detected on CT (Figure 6). No clinical sign of recurrence has been observed during an 18-month follow-up.

COMMENT

The term “dermoid” has been used extensively to describe dermoid and epidermoid cysts in the orbit; however, they are different entities. Both are cystic choristomas filled with keratin, cholesterol clefts, or degenerated blood components, and produced by keratinizing squamous epithelium; but whereas true dermoids have skin appendages on their walls, epidermoid cyst walls do not have these appendages. Interestingly, their clinical behaviors usually differ. Dermoid cysts are common, diagnosed during infancy or early childhood, located superficially or in the anterior orbit, commonly mold bone, and rarely induce bone lysis (dumbbell-dermoids). Conversely, epidermoid cysts are rare lesions that are usually diagnosed later in life, likely because they are deeper in the orbit, and they typically develop within orbital bones diploe. In fact, intradiploic epidermoids are rare tumors more commonly located in the skull bones, particularly in the temporal or frontal bone. Only about 100 cases have been described in the literature, with fewer than 20 well-documented cases located in the bony orbit.

As in our case, orbital intradiploic epidermoid cysts are commonly located in the lateral wall. They may invade other orbital walls and extend into the orbit, temporalis fossa, or cranial cavity. In our patient, all these growth patterns occurred, which, combined with its significant size, accounted for the name “giant” epidermoid cyst. A similar case with middle cranial fossa involvement was reported by Rumelt et al.

The clinical characteristics of orbital intradiploic epidermoid cysts were well described by Eijpe et al. The most common clinical manifestation is unilateral proptosis, which
is eventually associated with extraocular muscle impairment or diplopia. Nonophthalmologic signs include headache and other neurologic symptoms that are especially common when intracranial extension occurs. Cranial or significant temporalis fossa invasion may induce pulsatile proptosis or proptosis associated with mastication, respectively. In the present case, exophthalmos and downward displacement of the left eye were the main symptoms that were only associated with ipsilateral headache.

Both CT and MRI play an essential role in diagnosing epidermoid cysts and determining the surgical excision strategy. Three types of secondary bone changes have been described: sclerosis, erosion, and fossa formation with bone attenuation. Characteristic bone destruction with sclerotic borders was the CT finding in our case. Despite its limited ability to define bone changes, MRI has a definitive role in further characterization of these lesions: both dermoid and epidermoid cysts display a highly suggestive hyperintense signal on T2-weighted images. Moreover, MRI may more accurately demonstrate intracranial extension, as in this case. Differential diagnosis in our case included eosinophilic and cholesterol granuloma.

Most of these tumors are surgically managed through a coronal approach to provide wider exposure, and a craniotomy is performed to deal with intracranial extension. In our case, because of the low probability of brain involvement, a lateral orbitotomy with a bone flap was preferred, which allowed for good exposure and excision of the cyst through a small cutaneous incision. The importance of complete excision must be emphasized because incomplete excision may lead to chronic granulomatous inflammation or recurrence; moreover, recurrence is associated with increased risk of malignancy. In the present case, the cyst has not recurred after an 18-month follow-up.

In summary, the present case is an exceedingly rare occurrence of unilateral proptosis resulting from an intradiploic epidermoid cyst primarily located in the bony orbit and secondarily extending to the cranial cavity and temporalis fossa. Its clinical presentation, imaging characteristics, and management rationale are also stressed. Intradiploic epidermoid cyst must be considered in the differential diagnosis of benign bone-destroying lesions affecting the orbit.

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