Giant Intradiploic Epidermoid Cyst of the Skull: A Case Report and Management of Large Cystic Scalp Masses

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Epidermoid cysts of the skull are very rare, benign, slow-growing tumors. Only a very few cases have ever been reported and they can be found in the diploë, the cranial cavity, or in the spinal canal. Intradiploic epidermoid cysts are even rarer and can be found in any part of the skull. There are 2 theories regarding the derivation of these cysts. One theory suggests that they are derived from ectodermal tissue misplaced in the cranial bones during embryonic development. The alternate theory suggests possible traumatic implantation of these cells. They often reach a large size (3–5 cm) without creating neurologic symptoms. A 34-year-old man was seen for resection of a large melanoma on his right arm, and in the workup he was noted to have a large, long-standing lesion on the posterior scalp. After radiologic assessment was completed, the right parietooccipital lesion was removed with a subsequent cranioplasty. The management of this lesion was handled appropriately but could have led to disastrous consequences if a more cavalier approach had been taken. The clinical presentation, pathologic findings, radiologic features, and treatment of the lesion are reviewed.

A 34-year-old male presented to the clinic for follow-up status postremoval of an amelanotic melanoma from his right arm. He stated he also had a very large scalp mass in the right parietal region that had been diagnosed at another clinic as a sebaceous or pilar cyst 7 or 8 years prior to presenting. He was told removal of the cyst would not be a problem. Further, there was a history of trauma to the area prior to the development of the lesion. The patient stated he struck his head on the hood of his car while working on the engine and shortly thereafter he noted a small, nonpenetrating lesion. The lesion did not require medical intervention at that time. Since that time he has remained asymptomatic and, upon questioning, denied headache, loss of consciousness, seizure activity, visual disturbances, memory loss, speech dysfunction, extremity weakness, numbness, parasthesia, loss of balance, or loss of coordination. The patient found this lesion bothersome aesthetically and wanted it removed. He stated that one practitioner previously suggested in-office aspiration and removal. Past medical history was positive for the aforementioned melanoma and cardiac arrhythmia. The patient denied any pertinent family history.

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The authors report no conflicts of interest in relation to this article.

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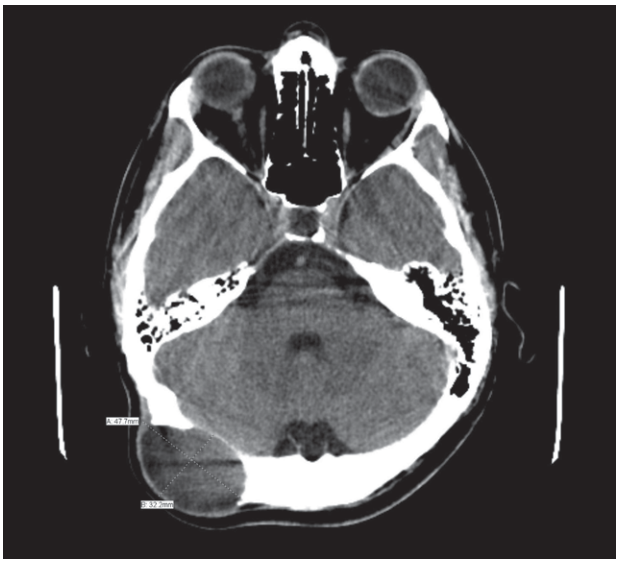

Figure 1. Computed tomography of the head showing a 4.7×3.2×4-cm cyst extending above the right parietal scalp.

Physical exam revealed a healthy male, appearing to be the age he stated, with no notable asymmetry. A 3.5×5-cm, semifirm, nodular lesion with smooth borders was noted above the right parietal scalp. No erythema or fluctuance was noted. The lesion was fixed and nontender on palpation. Other than the noted lesion, he was normocephalic and his mental status was not altered. Testing showed intact cranial nerves, and pupils were equal, round, and reactive to light with accommodation.

The decision to perform a computed tomography scan with and without contrast was made due to the fixed nature of this large lesion. The radiologist suggested this test would help enhance the visualization of the extent of the lesion. The radiologist noted that the posterior scalp housed a lesion 4.7×3.2×4 cm in size (Figure 1). The eroding of the calvarium above the right parietal region can be appreciated with blurring of the dural margin (Figure 2). Typically, benign scalp lesions do not erode the calvarium, thus a neurosurgical evaluation was performed. At the suggestion of the neurosurgeon, the patient elected for resection and cranioplasty. Using primarily blunt dissection, the surgeon followed the capsule around, dissecting the subcutaneous plane down to where it entered the bone. With a curette, it was separated from the inner table of the skull and then the tumor was extirpated in toto. When the capsule was removed, it appeared to contain typical sebaceous material, yellow-gray and flocculent in character. The dura was then carefully examined, and residual capsule and sebaceous material was carefully curetted. A cranioplasty was performed without complication to repair the defect. The entire procedure was well tolerated by the patient.

The specimen was sent for pathologic evaluation. The pathology report indicated that the cyst was deeply invasive due to the presence of skeletal muscle (Figure 3). It was lined with squamous epithelium, including a granular layer, and was filled with keratinaceous material (Figure 4). The cyst was entirely surrounded by a fibrous capsule. Chronic inflammation and foreign-body reaction were noted, as evidenced by giant cells. After discussion of the presentation and radiological/surgical findings, a diagnosis of intradiploic epidermoid cyst (IEC) was made.

Because the cyst was removed completely, recurrence is highly unlikely. The patient recovered without incident and is monitored routinely for signs of recurrence.

Extremely rare, IECs are congenital intracranial tumors and, to date, relatively few cases have been reported. They were first described by Cushing[1] in 1922 and can occur in any part of the skull. As a congenital tumor, they are derived from ectodermal remnants that persist or sequestered within the cranium along embryonic fusion planes during embryonic development.[2] An alternative theory is that they may also develop after trauma.[3,4] A number of case studies describe head injuries as the cause of inclusion of epidermal cells into the diploë of the bone. This typically leads to an intradiploic pseudomeningocele, which is of traumatic origin and should be distinguished from an epidermoid cyst, which is of congenital origin. The histopathology of the 2 lesions is notably distinct.

These rare tumors have been reported to be found in individuals ranging in age from 1 year to 70 years with a mean age of 32 to 38 years.[5,6] No gender preference has been found. It is typical for clinical symptoms to be delayed.

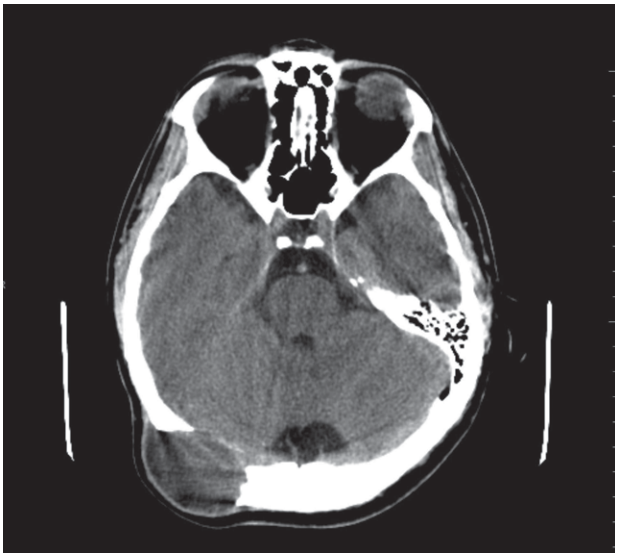

Figure 2. Computed tomography of the head showing eroding of the calvarium by an intradiploic epidermoid cyst and its projection onto the dura.

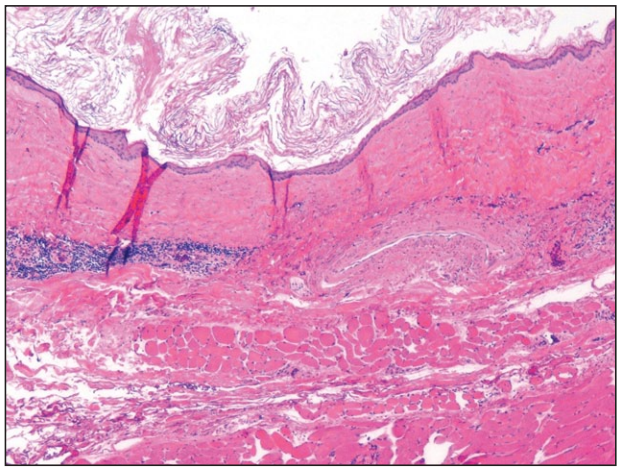

Figure 3. The aggressive nature of an intradiploic epidermoid cyst and its surrounding capsule (H&E, original magnification ×40).

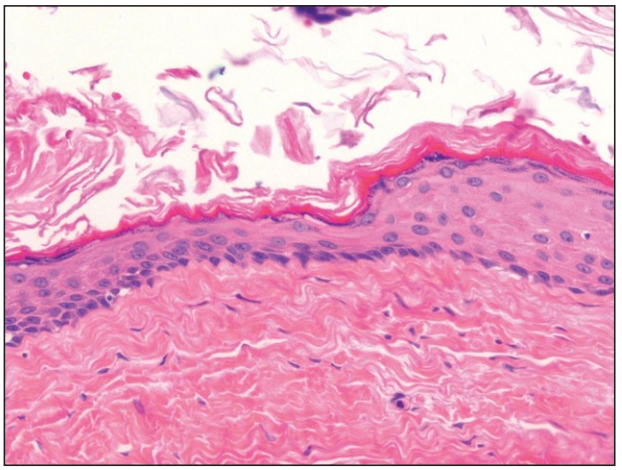

Figure 4. An intradiploic epidermoid cyst lined with squamous epithelium including a granular layer (H&E, original magnification ×200).

In addition, IECs are thought to be due to the sequestration of keratin and cellular debris as the cyst gradually enlarges. They typically present as a painless swelling under the scalp, and the most frequent symptom is headache.[7] Arana et al[8] reported the averages of presenting symptoms and signs in 37 histologically-proven IECs as follows: symptomatic, 89.2%; lump alone, 45.9%; lump and pain or headache, 21.6%; exophthalmos, 16.2%; asymptomatic, 10.8%; and mixed symptoms, 5.4%. Other symptoms encountered include seizures, focal neurological deficits, pneumocephalus, and intracranial hypertension.[7,9]

Computer tomography of IECs allows for thorough assessment of involvement and intracranial extension. There is no contrast enhancement. They typically present as large, homogenous, hypodense areas with erosion and expansion of both the inner and outer tables of the skull, with sharply defined sclerotic borders and calcification.[7] This sclerotic margin is primarily due to the slow, steady pressure exerted by the tumor over time. Simultaneous erosion of both the inner and outer table of the skull has been seen in up to 72% of cases.[8] Magnetic resonance imaging (MRI) is preferred, although computer tomography may be more readily available. However, an MRI is best utilized for determining the extent of intracranial expansion of the epidermoid tumor due to the signal difference between the tumor and the neural tissue.[10] Our patient's radiologic findings were similar to cases presented in the literature.

The differential diagnosis for these tumors is extensive and includes dermoid cyst, pilar cyst, hemangioma, lipoma, eosinophilic granuloma, fibrous dysplasia, aneurismal bone cyst, and meningeoma.[11] Coming to the correct diagnosis relies on an appropriate history and examination. A frequent misdiagnosis is a hydatid cyst because it mimics an IEC in many ways but lacks bony erosion or calcification.[7] Radiologic studies are helpful in making the diagnosis, but surgical removal and histopathologic confirmation are definitive; hence, IECs should be removed with great care. Cushing[1] stated the aim of surgery is the total removal of the capsule, which must be carefully dissected from the bone and dura mater. Complete removal of the capsule is essential for a good long-term prognosis, and once the epithelial lining is removed the cysts do not recur.[12,13]

Malignant changes in the epithelial lining of epidermoid cysts have been reported. Yanai et al[14] found 5 cases with malignant transformation out of a series of 100 IECs collected from medical literature. When epidermoid cysts have undergone several operations with repeated bouts of inflammation, they are more likely to make a transformation into a squamous cell carcinoma.[15] Another case in the literature describes a malignant change leading to melanoma in situ.[16] Due to the positive history of our patient for melanoma, we were concerned that a malignant change was possible, and it warranted complete pathologic examination of the epithelium around the cysts so that no malignant process was overlooked.

Determining the anatomic origin of an IEC can be difficult and often requires an intraoperative examination. In this case, the lesion seemed to originate from the inner table of the skull and therefore qualifies as a true IEC of congenital or traumatic origin. More important is that regardless of the origin, this typically benign lesion had behaved more aggressively. On physical examination, this lesion looked much like a pilar cyst in location and feel with the exception of the fixed nature of this lesion. Pilar cysts are typically smooth, mobile, with or without inflammation, and devoid of a punctum. Furthermore, histologically pilar cysts lack a granular layer. Epidermoid cysts frequently contain a punctum, are not inflamed, and may or may not

be mobile depending on the depth of involvement. They can be pigmented in darker skinned individuals.

Most practitioners are cautious about midline scalp lesions but many may not know to be cautious about large cranial lesions in other locations. Large tethered cystic lesions of the face and scalp should be examined thoroughly and possibly with imaging studies before excision is undertaken. In our case, the presenting lesion was 3.5×5 cm and constituted a large lesion. Currently, there is no consensus for when a scalp lesion can be considered large, but most IECs present around 3×5 cm. Obviously, every tethered scalp lesion does not require an MRI. Knowing the entire clinical picture is essential. A smaller infected scalp lesion could very well be tethered to the underlying scalp and imaging would be unnecessary. In our case, the lesion grew slowly for many years, which became fixed and quite large (>3 cm). In-office removal of the lesion could have had disastrous consequences.

Even though IECs are benign, slow-growing tumors, they can reach enormous size. If a patient presents with a slow-growing scalp lesion that has attained substantial size, radiologic imaging should be obtained, especially if there are any associated symptoms. These tumors should be removed prophylactically with complete removal of the tumor and capsule. Long-term prognosis is excellent.

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