

Intradiploic epidermoid cyst of the skull. Case report

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Abstract

Authors describe a giant intradiploic epidermoid cyst of the cranial vault with massive intra- and extracranial extension and analyze the clinicopathological, imaging features and treatment of these lesions in the light of the most important published data. A 38-years old male patient with a history of chronic headache reported a painless subcutaneous swelling 4 months ago on the right frontoparietal scalp. CT and MRI evaluation revealed a large well-defined extracerebral mass in the frontoparietal scalp with destruction of both the inner and outer tables of the bone. The patient underwent a total microsurgical resection of the tumor and the cranial defect produced by the resection of the space-occupying lesion was repaired with titanium wire mesh and methyl methacrylate cranioplasty. The histological examination has shown a cystic structure lined by squamous epithelium and containing laminated keratin material, aspects that are suggesting a epidermoid cyst. The patient post-operative course was uneventful, without any neurological deficit and was discharged after 48 hours of hospitalisation in a good medical condition. The review of the literature shows that the parietal bone location for intradiploic epidermoid tumors is rare. Complete surgical resection represent the gold

standard of treatment, and no adjuvant therapy is needed. Recent advances in diagnostic imaging and treatment of these lesions are reviewed.

Keywords: dermoid, epidermoid, intracranial tumor, radical removal, skull tumor

Intracranial epidermoid cyst, although rare, is well documented. Also known as epidermoid tumor or ectodermal inclusion cyst, or “pearly tumor”, epidermoid cyst is a benign congenital inclusion cyst that account for 1% of all intracranial tumors (1, 11). It is thought to arise from defects in the separation of the neuroectoderm during the formation of the neural tube, leading to sequestration of ectodermal remnants within the cranial bones (5). Intracranial epidermoid cysts are well circumscribed and most often occur within the diploe (1), in the parasellar region, sylvian fissure, cerebellopontine angle, posterior fossa, and fourth ventricle. Intradiploic epidermoid cysts are less common than the intradural varieties (9).

In general, epidermoid cysts typically appear on CT scans as low-density, hypodense, nonenhancing lesions, similar to the fat density. On MRI this lesion demonstrates high signal intensity in T1-weighted and variable T2-weighted signal. Sometimes, the cyst contents can be hyperdense, mimicking a haemorrhage.

Case Report

History. A 38-years old man was admitted with a history of chronic headache, mainly in the right parietal region, which had recently increased in severity and frequency. For 4 months he had noticed a small painless bulge under the scalp.

Examination. Neurological exam, didn't reveal any deficits but physical examination found a right frontoparietal soft, non-tender lump, which had no mobility over the underlying bone and the skin covering the lesion could not be retracted.

Contrast-enhanced computed tomography showed destruction of the right parietal bone by a large extracerebral hypodense intradiploic tumor. In bone windows the CT scan shows erosion of both inner and outer table of the calvarium and compression on the right parietal lobe (Figure 1). On the MRI examination the mass demonstrated homogenously high signal intensity during both T1- and T2-

weighted sequences, and no perifocal cerebral edema was detected on the T2-weighted sequence (Figure 2).

Operation. The patient underwent a right parietal craniotomy. When the scalp flap was reflected, a white encapsulated, mostly intradiploic large tumor was identified.

The 7 cm tumor with irregular calcified borders was herniating through bone defect (Figure 3)

The tumor was resected within the apparently normal bone followed by the excision of the markedly thickened dura.

A duroplasty with a galeal flap and a reconstruction of the skull bone with a titanium mesh were performed (Figure 4).

Postoperative course. After the intervention, the patient's neurological status remained stable with intact motor strength and sensation.

Histologically, a diploic epidermoid cyst was established.

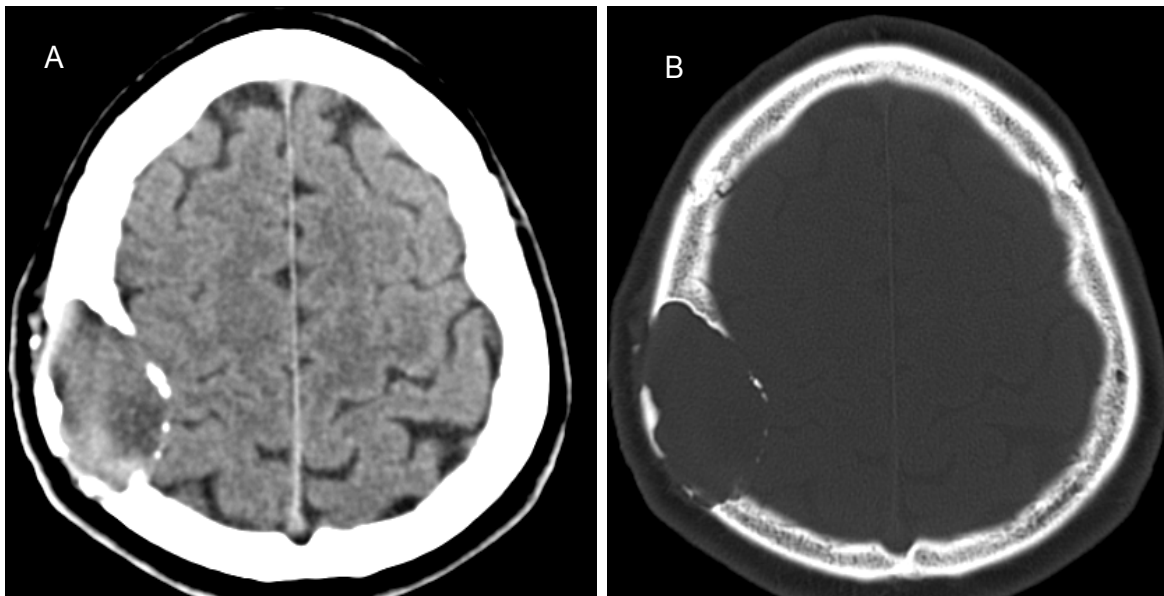


Figure 1 A Computerized tomography scans demonstrating a well circumscribed extracerebral hypodense area surrounded by a hyperdense rim. B: CT with bone window reveals erosion of both tables of the vault.

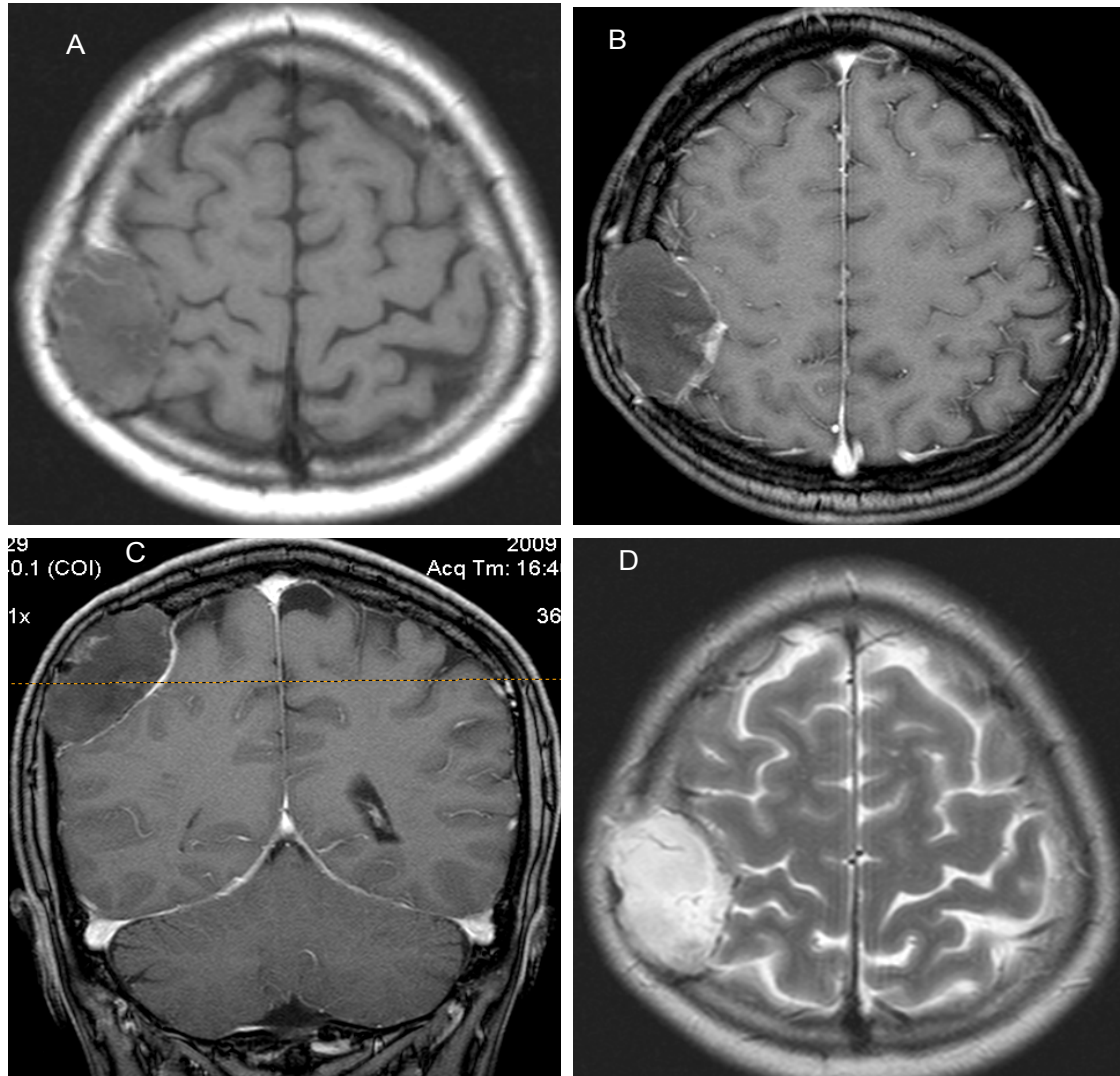


Figure 2 Preoperative plain MRI images. **A:** Axial T1-weighted section revealing well demarcated tumor.

B: Tumor did not show enhancement after Gd administration. **C:** Coronal section.

D: T2-weighted axial section shows no perifocal edema.

Discussion

Intracranial epidermoid cysts are congenital lesions with benign histological features and very slow growth, via progressive accumulation of normally dividing cells (epidermal cells) (11). This tumor arises from ectodermal cellular remnants, during gestation (weeks 3-5), as a result of incomplete cleavage of the neural ectoderm from the cutaneous ectoderm (4). The most common presentation is within

the third or fourth decade of life with a painless, long-standing lesion, subcutaneous scalp swelling covered with normal skin (10). Men are affected more often than women (2). Epidermoid cysts have a thin capsule of stratified squamous epithelium filled by keratin, cellular debris and cholesterol, and do not contain hair or other dermal elements which differentiate them from the dermoid tumor (2,7). The epidermoids are having a slow growing rate,

approximating that of normal skin, a condition which allows the intracranial structures, by using its reserve spaces, to adapt and tolerate the presence of mass lesion until all the reserve spaces are used, situation in which further compensation is no longer possible and the lesion becomes symptomatic (according to the localization and the compressed structures) (12, 8).

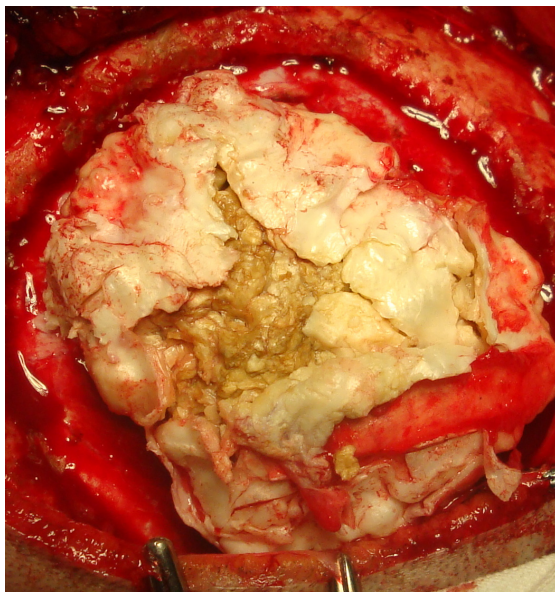


Figure 3 Intraoperative photograph showing this large tumor, which is located between the inner and outer table of the bone.

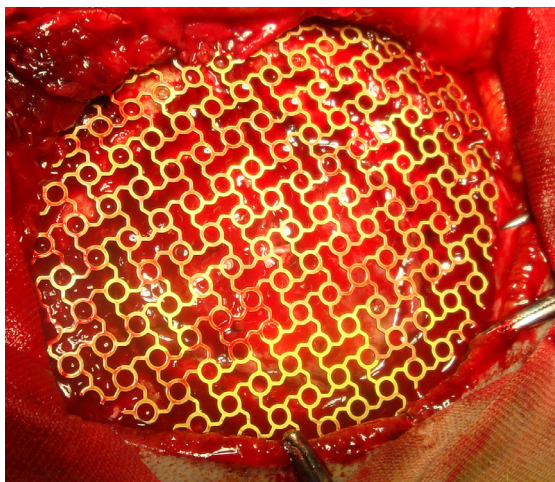


Figure 4 Cranioplasty with titanium mesh.

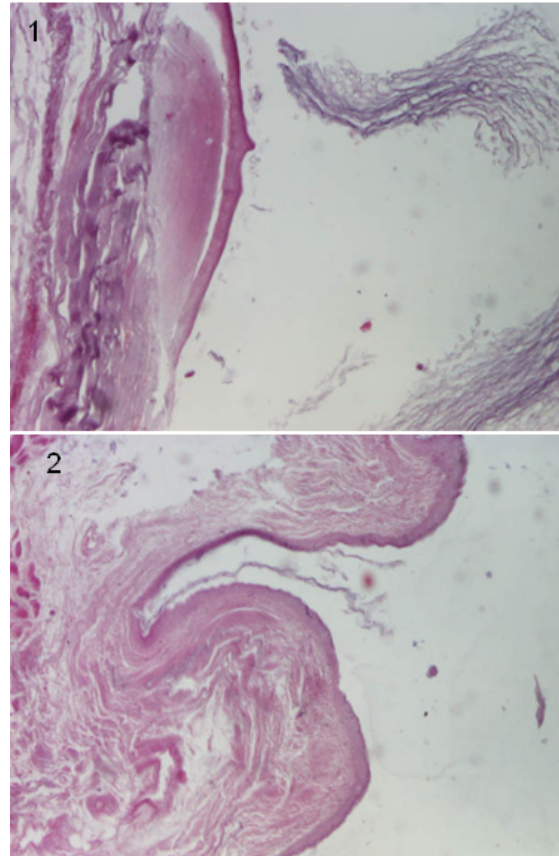


Figure 5 - 1: Photomicrograph of resected intradiploic epidermoid cyst demonstrates multiple concentric laminated layers of keratin within the lumen. The cyst is lined by a squamous epithelium. In this area there is normal osseous lamella adjacent to squamous epithelium. (Hematoxylin-Eosin, original magnification x 40).

Figure 5 - 2: Another low-power photomicrograph of resected intradiploic epidermoid cyst shows normal striated muscle and conjunctive tissue below the epidermoid cyst wall as the bone was distructed and the cyst penetrate beyond it. (Hematoxylin-Eosin, original magnification x 40).

A possible complication of intradiploic epidermoid tumors is the spontaneous cyst rupture with secondary discharge of the cystic content (keratin, cellular debris and cholesterol) into the subarachnoid space, producing aseptic meningitis which frequently leads to chronic granulomatous arachnoiditis (5).

Findings of CT are acknowledged to give correct diagnosis for intradiploic epidermoid cyst preoperatively. On CT, simultaneous erosion of inner and outer table of the skull is reported to occur in up to 72% of cases with intradiploic epidermoids (10). They are soft masses, which may be hypointense or isointense to the brain, and the dural infiltration is reported in 10% of the cases (3). Other lesions of the scalp and skull to be considered in differential diagnosis include: calcified cephalhematomas, occult meningocele and eosinophilic granuloma, osteolytic intradiploic metastasis (8, 4). The goal of surgery in the treatment of epidermoid tumors is total surgical removal of the tumor together with its capsule, which is the only living and growing part of the tumor.

Conclusions

We recommend the prophylactic removal of these tumors through surgery and primary repair when required. Adequate exposure of the epidermoid cyst is needed to allow complete excision of the tumor and its capsule, with the goal of preventing recurrences, inflammation and the possibility of malignant transformation.

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