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TEST YOURSELF: ANSWER

A 36-year-old man with sudden severe headache

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Diagnosis

Giant intradiploic epidermoid cyst. A large arachnoid cyst of the middle fossa of the brain is visible inferior to the intradiploic epidermoid cyst.

Discussion

Epidermoid cyst (EC) is a rare, benign, slow-growing lesion of the skull. The origin of ECs of the skull is debated. According to the congenital theory, ECs arise from ectodermic inclusions during neural tube closure (weeks 3–5 of embryogenesis). Epithelial cells become trapped in the bone by the laterally migrating otic or optic capsule [1–4].

Acquired ECs of the skull are believed to develop from inclusions of the epithelium after trauma [1, 2, 5].

Epidermoid cysts of the skull are detected incidentally by imaging for other reasons [1].

Large ECs of the skull may become symptomatic, causing compression of the brain and neurological effects, including headache, seizures, optic nerve or venous sinus compression, and intracranial hypertension [1, 5, 6]. In our case, the cause of the patient's headache was nonspecific; it could have been attributed to direct meningeal irritation by the lesion or, less likely, to chronic changes in intracranial pressure, related to the mass.

Late complications of ECs include a fistulous tract that opens to the skin, suprainfections, and meningeal syndrome subsequent to cyst rupture [4, 7]. Rarely, intradiploic ECs undergo malignant transformation into squamous cell carcinoma [8].

On computed tomography (CT), ECs of the skull usually appear as well-defined, heterogeneous masses with densities ranging from −20 to +20 Hounsfield units (HUs), interspersed with higher-density areas that correspond to protein or cholesterol deposits (Fig. 1; see Test Yourself: Question) [9].

Epidermoid cysts typically have low signal intensity on T1-weighted MRI and high signal intensity on T2-weighted MRI. The T1-weighted MRI signal may increase focally, corresponding to cholesterol deposits (Fig. 2; see Test Yourself: Question) [1, 10].

Most ECs fail to show enhancement after injection of contrast media (Fig. 2) [10].

In our case, a large arachnoid cyst of the middle fossa was visible inferior to the EC.

The EC and CSF of the arachnoid cyst could not be differentiated based on the density by CT (the average density was approximately 12 HU for both lesions; Fig. 1), whereas EC was easily detected by MRI (Fig. 2). ECs are heterogeneous and hyperintense relative to cerebrospinal fluid (CSF) on fluid attenuation inversion recovery (FLAIR) images (Fig. 2) [11]. Further, unlike arachnoid

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cysts, ECs do not restrict on diffusion-weighted imaging (DWI) and have similar apparent diffusion coefficients (ADCs) to brain parenchyma (Fig. 2) [12].

The differential diagnosis of EC of the skull includes other solitary lytic lesions, such as bone metastases, dermoid cysts, eosinophilic granulomas, meningioma, and solitary plasmacytomas. It is particularly common to misdiagnose EC as a dermoid cyst, the definitive diagnosis of which is primarily histological (Fig. 3). Nevertheless, ECs are typically off the midline, whereas dermoid tumors lie closer to the median sagittal plane [6].

Eosinophilic granulomas develop in infancy [13]. Local pain is the chief symptom, and the bone lesion is accompanied by a palpable tender mass that enhances after administration of contrast medium. At times, a button sequestrum is present within the osteolytic lesion, representing residual bone; the sequestrum can be seen better on CT [14].

Solitary plasmacytoma, meningioma, and bone metastases, unlike EC, have solid density on CT and enhance with administration of contrast media [15].

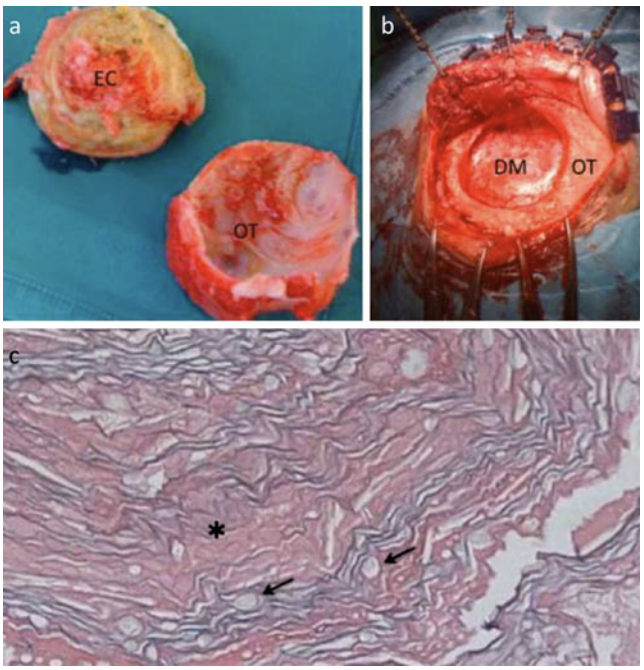

Fig. 3 **a** Gross anatomy, **b** surgical images, and **c** histological specimen (hematoxylin and eosin stain; magnification ×100) of the epidermoid cyst. The surgical images show a thinned outer table (*OT*), the epidermoid cyst (*EC*) with no macroscopic appearance of the inner table, and an intact dura mater (*DM*), demonstrating the intradiploic origin of the mass. These findings formally demonstrate the bony origin of the mass being the superficial layer of the dura mater, the skull's inner periosteum. According to the histology, there is amorphous material that contains keratin debris (*asterisk*) and cholesterol deposits (*arrows*), typical of an epidermoid cyst

Total removal of ECs is associated with a good long-term prognosis. If they are not removed completely, ECs can recidivate, recurring in 8.3% to 25.0% of surgical series cases [8].

In conclusion, intradiploic ECs of the skull are uncommon, but should be included in the differential diagnosis of solitary lytic lesions of the skull. In rare cases, a giant cyst can become symptomatic. MRI, including FLAIR and DWI sequences, is crucial for surgical planning, to detect complications and visualize surgical residuals.

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