Frontal bone epidermoid – a rare cause of proptosis

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Abstract

Frontal bone epidermoid is a rare condition which may present with proptosis, extraocular muscle imbalance, or loss of vision. Clinically and on plain radiographs an epidermoid may be confused with commoner conditions such as frontal mucocoele. However, the characteristic CT scan appearance, which shows a bone cyst with similar density to brain tissue, should lead to an accurate diagnosis and appropriate surgical management.

Frontal bone epidermoid is an extremely rare condition with less than 30 authentically documented cases reported in the literature. The presentation is generally with facial asymmetry, unilateral proptosis, extraocular muscle imbalance, and occasionally visual loss.^{1,2} The clinical and plain radiographic features of the condition show a certain similarity with frontal sinus mucocoele,^{3,4} which can lead to diagnostic confusion and inappropriate surgical treatment. Fortunately the specific CT scan appearance enables an accurate diagnosis to be made.

We report a single case of a frontal bone epidermoid which illustrates the typical clinical, plain radiographic, and CT scan features of the disease.

Case report

A 57-year-old male presented with progressive proptosis and facial asymmetry. He was otherwise well, with no history of headache, paranasal sinus disease, or trauma. His right visual acuity was 6/6, and he had an irreducible, non-pulsatile right proptosis and hypotropia, but no frank

Figure 2: CT scan with coronal projection showing destruction of the roof of the right orbit and downward displacement of the globe by the epidermoid, which can be seen to have a similar tissue density to brain.

diplopia. Further examination of his eyes, ears, nose, and throat and a general examination revealed no other abnormality.

A plain x-ray of the sinuses showed a rounded, sharply circumscribed, bony defect in the right frontal bone with marginal sclerosis (Fig 1). The CT scan coronal (Fig 2) and axial (Fig 3) projections showed destruction of the roof of the orbit, with downward displacement of the globe, and defects in the anterior and posterior surfaces of the frontal bone. The wall of the cyst was well defined by contrast media, and the contents had a similar density to that of brain tissue.

Under a general anaesthetic a curved incision extending from the medial canthus through the eyebrow to the temple was made. A large cyst (3.5 cm in diameter) occupied a cavity in the frontal bone, communicating via the bony defects noted on the CT scan with the orbit and anterior cranial fossa. The dura and the orbital periosteum were intact. The cyst was removed, and a Silastic drainage tube into the nose was left in situ for two months. Postoperatively he was treated with cefuroxime and metronidazole.

Figure 1: Plain x-ray of sinuses (occipitofrontal view) showing cavitation in the right frontal bone.

Figure 3: CT scan with axial projection showing erosion of the anterior and posterior surfaces of the frontal bone by the epidermoid.

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Histology of the cyst revealed an epithelial lining with a central mass of keratin. The proptosis has resolved completely and he has made a full recovery.

Discussion

Epidermoids result from the proliferation of epithelial elements in an inappropriate site. Primary epidermoids arise from embryonal inclusions of epithelial cell rests at sites of fusion of embryonal ectoderm. Secondary epidermoids may result from a postnatal event such as traumatic implantation of cells, cell migration, or metaplasia.⁵

Histologically an epidermoid consists of a cyst with a squamous epithelium lining containing keratin, lipid laden histiocytes, cholesterol, and a fibrous stroma.⁵ The name cholesteatoma is frequently applied to such lesions,^{3,4} though it is a misleading term which can lead to confusion with other similar pathological conditions and thus is best avoided.⁵ The presence of an epithelial cell lining enables an epidermoid to be differentiated from a cholesterol granuloma,¹

and the absence of any mesodermal elements differentiates it from a dermoid cyst.⁵

The clinical presentation of an epidermoid can easily be confused with a frontal sinus mucocele,^{3,4} as, typically, the patient experiences vague headaches, a progressive proptosis with hypotropia, and sometimes visual loss. In such cases an incorrect diagnosis can lead to an inadequate surgical approach and incomplete removal, putting the patient at risk from recurrences⁶ as long as 40 years after the original surgery⁷ and, on rare occasions, malignant transformation.⁸

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