Epidermoid Tumor of the Skull with Intracranial Pneumatocele

Case Report

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We are reporting a case of intradiploic epidermoid tumor with an interesting and unusual mode of presentation. A brief discussion of the etiological concepts and x-ray features of this uncommon developmental tumor is included.3

Case Report

This 46-year-old man was admitted to the hospital for evaluation of a foul smelling, brown-colored discharge from the left nostril of approximately 5 months’ duration, and intermittent frontal headaches.

Examination. The nasal turbinates were swollen bilaterally, but there was no mass or abnormal discharge. Temperature, extraocular movement, pupillary reactions, and optic discs were normal. There was no proptosis.

The clear, colorless lumbar cerebrospinal fluid was under normal pressure, contained no abnormal cells, a chloride content of 25 mEq/L, and 22 mg% of protein. The electroencephalogram was normal. Plain skull x-rays demonstrated a radiolucent defect with a scalloped border within the frontal bone. The sharp margin of the lesion was sclerotic in some areas. The right frontal sinus was clouded and its mucoperiosteal margin poorly defined adjacent to the lesion. An air-fluid level in a mass about 5 cm in diameter was present posterior to the cranial defect (Fig. 1). The right carotid arteriogram demonstrated posterior displacement of the pericallosal artery and its branches by the mass, as well as displacement of these arteries across the midline (Fig. 2). These findings suggested an extra-axial mass within the right frontal area, corresponding to the intracranial pneumatocele noted on the plain skull examination. The arteriogram revealed only displacement of normal vessels and no tumor stain or abnormal vessels.

Operation. Through a transfrontal craniotomy, a large extradural cystic mass was found. The lesion had arisen within the diploic space and extended intracranially by

![Fig. 1. Left: Frontal x-ray of the skull demonstrating the destructive focus in the frontal bone, with areas of sclerosis (solid arrows), clouding, and poor definition of the frontal sinus, and an intracranial air-fluid level (open arrows). Right: Lateral x-ray demonstrates the scalloped margin of the skull defect (solid arrows) and the pneumatocele with air-fluid level (open arrow).](image-url)
destruction of the inner table of the frontal bone. The mass also extended into the frontal sinus and had partially eroded portions of the roof of the right orbit. There was no extension into the right orbit. The mass was completely excised and the skull defect closed with methacrylate. Except for transient nausea the patient experienced an uncomplicated postoperative course and was completely asymptomatic upon discharge from the hospital. Histological examination confirmed the diagnosis of benign cystic epidermoid with no evidence of malignant degeneration.

Discussion

Epidermoids are cystic masses of ectodermal origin, lined with stratified squamous epithelium and filled with keratin, cellular debris, and cholesterol crystals. Because of their cholesterol content, epidermoids are sometimes called cranial cholesteatomas. A generally accepted theory of origin, first proposed by von Remak, is that they are developmental cysts or neoplasms which arise from ectodermal cells sequestered in the early weeks of embryogenesis, during closure of the neural groove. This explains the more common midline occurrence of the lesions. Lateral tumors, within the temporal bone, paranasal sinuses, and near the orbit, are explained by displacement of ectodermal rests during development of the otic and optic vesicles. The finding of cranial epidermoids following trauma has suggested to some observers that occasional cases may be due to traumatic implantation of epithelial cells. Although rare, malignant degeneration of a benign epidermoid into epidermoid carcinoma has been reported.

The symptoms produced by cranial epidermoids are nonspecific, and almost half the patients may be entirely asymptomatic. The classic x-ray appearance is that of a well-circumscribed radiolucent defect with a sclerotic border, located within the diploic space, and occasionally producing expansion of both the inner and outer tables of the skull. Tangential examination may demonstrate destruction of the inner table which allows intracranial extension of the mass. As demonstrated by the present case, the mass may extend into the paranasal sinuses, and necrosis with partial extrusion of cyst contents into the sinus provides the potential mechanism for the development of an intracranial pneumatocele. Although external carotid arteriograms were not performed, the avascularity of the lesion noted on internal carotid angiography conforms to the consensus of previous angiographic reports.

Summary

We have reported a case of cranial epidermoid cyst which presented as a destructive focus within the frontal bone, extending into the frontal sinus and creating a large extradural intracranial pneumatocele. We have briefly discussed the development of such a lesion and the clinical features leading to its diagnosis.

Fig. 2. Left: Right carotid arteriogram demonstrates displacement of the pericallosal artery to the left by the mass. Right: The lateral examination of the carotid arteriogram demonstrates posterior displacement of the pericallosal artery and its branches by the mass.
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References


