Giant intradiploic epidermoid cysts of the skull

Report of two cases

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The authors describe two cases of giant intradiploic epidermoid cysts of the cranial vault in which there
was massive intracranial extension causing signs of neurological involvement. The very slow growth and the
benign histological nature of these tumors explain their long preoperative evolution and the mild neurological
signs in some cases. Roentgenographic and computerized tomography findings permit a correct diagnosis.
Complete removal of these cysts and their capsules can be easily accomplished, despite their large size. Total
removal of these cysts is associated with a very good long-term prognosis.

Key Words • epidermoid cyst • skull tumor • intracranial tumor •
diploic cyst • computerized tomography

E pidemiod cysts are relatively uncommon. According to Lepoir and Pertuiset,1 epidermoid
tumors represent only 1% of intracranial tumors. They can occur within the diploe,9,23 in the cranial
cavity,6,21 or in the spinal canal.14 Intradiploic epidermoid cysts are less common than the intradural vari-
eties.10 They are derived from ectodermal cell rests that remain within the cranial bones during embryonic
development.5,13,23,24,26 Müller15 was the first to describe a diploic epidermoid cyst of the skull in 1838. Bucy1
reported finding only 13 cases in the literature in 1935, and Haig9 collected 100 cases that had been reported
prior to 1956.

Diploic epidermoid cysts are benign slow-growing tumors that are small or moderate in size. They are
commonly seen as an incidental finding on radiographic examination of the skull, or may become ap-
parent as a small lump under the scalp. In rare cases they can grow to a large size and cause intracranial
hypertension and focal neurological signs. In this paper we describe two cases of giant diploic epidermoid cysts
of the skull that caused clear signs of neurological involvement.

Case Reports

Case 1

This 58-year-old man came to our attention in De-
cember, 1978. For 10 years he had complained of
headache, which occurred mainly in the right parietal
region. For 20 days before admission he had experi-
cenced progressive weakness in the left arm and leg.

Examination. Neurological examination at admission
revealed a moderate left-sided hemiparesis. The
patient was alert, with equal and reactive pupils and no
papilledema. X-ray films of the skull showed a large
lytic lesion involving the entire right parietal bone and
extending into the frontal and temporal regions (Fig. 1
upper). The lesion was intradiploic and delimited by
sclerotic borders. It contained irregular calcifications.
Computerized tomography (CT) scanning revealed a
large, well circumscribed, extracerebral hypodense area
in the right parietal region, in contact with the skull
bone and surrounded by a hyperdense rim (Fig. 1
lower). The lesion did not enhance and contained many calci-
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FIG. 1. Case 1. Upper: X-ray film of the skull showing a large lytic lesion in the right frontoparietal bone, delimited by sclerotic borders and containing irregular calcifications. Lower: Computerized tomography scans demonstrating a well circumscribed extracerebral hypodense area surrounded by a hyperdense rim and containing calcified regions.

was symptom-free. Computerized tomography scanning revealed only an internal porencephalic cavity in the right parietal region, but no recurrence of the tumor.

Case 2

This 59-year-old man was first seen by us in April, 1980. For 3 months he had complained of headache and had noticed a small painless lump under the scalp of the right temporal region. He occasionally experienced dizziness during brisk movements of the head and difficulty with mastication.

Examination. On admission, he was alert and had no neurological signs, but bilateral papilledema was noted. X-ray films of the skull showed a large sharply defined cranial defect in the right temporal region, with dense margins and erosion of both skull tables (Fig. 3 upper). Computerized tomography scanning revealed a large extracerebral hypodense area occupying the right temporal region and containing calcified nuclei (Fig. 3 lower). There was erosion of both the inner and outer tables of the bone.

Operation. On April 3, 1980, a scalp flap and the temporal muscle were reflected, and a round bone
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**Fig. 3. Case 2.** Upper: Plain x-ray film of the skull showing a large intradiploic cystic lesion in the left temporoparietal region. Lower: Computerized tomography scans showing massive intracranial extension of the tumor in the right temporal area. Calcifications and erosion of both tables of the vault can be seen.

Discussion

The important feature of these two cases is the enormous size of the tumors. Cushing was the first to report a large diploic epidermoid cyst in the parietotemporal region. In his case, the cyst measured 9 cm and markedly compressed the left cerebral hemisphere. Since that report, only a few cases of large intradiploic epidermoid cysts have been described in the literature.

Diploic epidermoid cysts commonly begin with a painless lump under the scalp, sometimes as a palpable bone defect. Headache is the most frequent symptom. In exceptional cases, large diploic epidermoid cysts can cause intracranial hypertension, convulsions, or focal neurological signs. The neurological findings in our cases were progressive hemiparesis and intracranial hypertension. A long clinical history is a common feature of large diploic epidermoid tumors, especially when the tumor grows intracranially with no extracranial extension. In these cases, the very slow growth of the tumor allows it to reach an enormous size in the cranial cavity without producing neurological deficits. The x-ray findings in our cases were characteristic of these lesions: that is, a large lytic area with erosion and expansion of both the inner and outer tables of the skull, sharply defined sclerotic borders, and calcification.

Computerized tomography scanning allows an appreciation of the size and exact location of the cyst and detects thinning or interruption of the skull tables. It also reveals the homogeneous hypodensity of the cyst contents, the presence of calcifications, and the condition of the underlying brain. Angiography, however, is nonspecific, because it reveals only an extracerebral avascular lesion.

Although these roentgenographic and CT findings are characteristic of large diploic epidermoid tumors, they can sometimes suggest the diagnosis of hydatid cyst. The typical changes of the bone tables and the presence of calcifications within the cyst are the important differential features indicating an epidermoid cyst. The difficulty in diagnosis is greatest when greenish-brown liquid is found within the cyst at operation, as in our Case 2. In such cases, only biopsy and histological examination of the wall allow a correct diagnosis.

Treatment of these large intradiploic epidermoid cysts involves complete removal, which is easily achieved despite of the large size of the tumor. As suggested by Cushing, the aim of surgery is the total removal of the capsule, which must be carefully dissected from the bone and dura mater. Sometimes, when the epithelial membrane of the cyst cannot be dissected, it is also necessary to remove the dura. Cranioplasty...
with acrylic material may be required when there is a large bone defect.

We wish to emphasize that the giant diploic epidermoid cysts in our cases were histologically benign, in spite of their size and the long clinical history. Nevertheless, rare cases of malignant degeneration into squamous cell carcinoma have been reported.9,10

We conclude that intradiploic epidermoid cysts are normally benign lesions of the skull, which can sometimes reach enormous size, causing mild neurological deficits. Total removal of the cyst and its capsule allows an excellent long-term prognosis, as shown by our two cases.

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