

Giant Primary Intradiploic Occipital Bone Epidermoid Tumour: A Case Report

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Abstract : A case of giant intradiploic epidermoid cyst which eroded both tables of the skull and expanded especially to the posterior fossa is presented. Despite the large size of the lesion, the patient had only mild symptoms and left cerebellar signs.

These slow growing, giant lesions are benign and can be totally removed without any deficit.

Key Words : Epidermoid tumour, Occipital bone, Intracranial hypertension

INTRODUCTION

Epidermoid and dermoid tumours of the central nervous system are rare, benign and slow - growing lesions (13,17,18). The reported incidence is between 0.7-1.8% of all intracranial tumours (1,2,14,16,18). They develop from displaced dorsal midline ectodermal rest cells between the 3th and the 5th gestational weeks during the neural tube closure (1,2,3,9,11, 14,18). They may also develop from the implantation of skin in deeper tissues by repeated lumbar puncture, ventricular taps and inadequate suturing of scalp laceration (9,13). Epidermoid cysts grow because of the gradual accumulation of keratin and cholesterol, desquamation products of epithelial cells in time (1,9,11,18). Symptoms may appear at any age but are commonly seen in adulthood (3,6,8). These tumours can be located anywhere in the cranium.

Giant intradiploic epidermoid tumours (GIDET) are extremely rare. We were able to find only three cases in the posterior fossa in the English literature (7,11,12).

CASE REPORT

A 51-year old man was admitted to the Department of Neurosurgery, Tepecik SSK Hospital, on May 5, 1993. He had noticed a nontender subcutaneous swelling in the left occipital area accompanied by headache, blurred vision, gait disturbance, bladder incontinence and global dementia in the last 3 months. On admission, examination revealed a subcutaneous swelling (8 cm in diameter) in the left occipital region. He was alert. Minimal left cerebellar signs (dysmetria and dysdiadochokinesia) and papilloedema were detected on neurological examination.

Skull plain x-rays displayed a well-circumscribed radiolucent defect with sclerotic borders measuring 8x8 cm in the left occipital bone (Fig 1). Computerized tomography (CT) revealed a large, well-circumscribed and extraaxial hypodense tumour in the left posterior fossa. The inner skull table was eroded totally and the outer table was very thin and eroded. The lesion contained areas of calcification and showed no contrast enhancement. CT also showed the compression of the occipital

horn of the left lateral and the 4th ventricles which resulted in moderate supratentorial hydrocephalus (Fig. 2). The tumour was extending towards the foramen magnum. Angiography was planned. Unfortunately, transfemoral catheterization was not successful due to atherosclerosis.



Fig. 1 : Lateral view of the plain X-ray film of the skull, showing the circumscribed radiolucent defect with sclerotic borders.

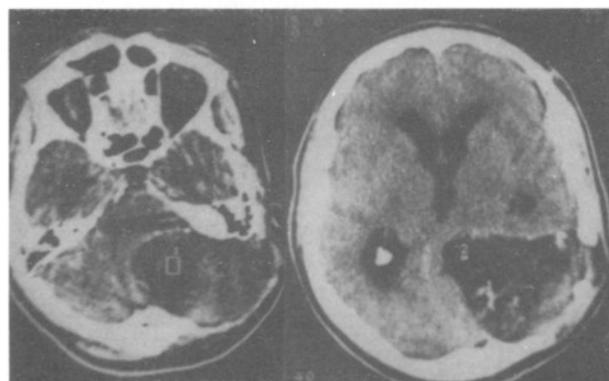


Fig. 2 : CT scans showing a large, well-circumscribed hypodense lesion, compressing the left occipital horn and the 4th ventricle. Note the calcification in the mass and mild ventricular dilatation.

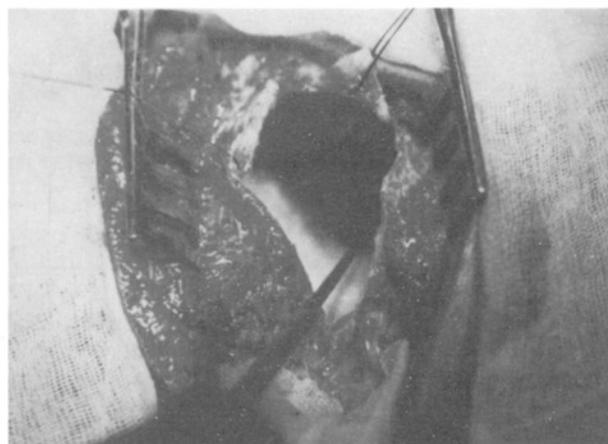


Fig. 3 : Operative photograph showing the tumor cavity after total resection. Note the eggshell-thin bone, tagged by sutures.

The patient was operated on in sitting position. Bone was found to be eggshell-thin. Semisolid cheesy material, causing compression of the cerebellar dura and elevation of the tentorium was removed totally (Fig. 3). After total removal, as the cerebellar dura did not expand and refill the epidural cavity, the space was filled with saline before the closing process.

Pathological examination confirmed epidermoid tumour. The symptoms of the patient resolved in a week. Postoperative CT scan, one week after surgery revealed total removal of the tumour and improvement of the hydrocephalus and also disclosed some air and epidural fluid in the cavity (Fig. 4) The patient was discharged without any neurological deficit.

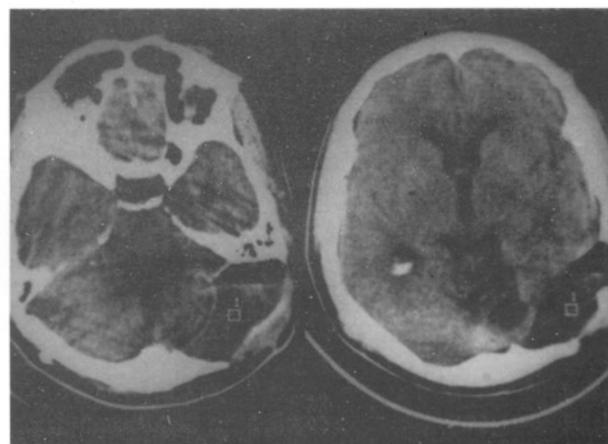


Fig. 4 : Postoperative CT scans showing total removal of the tumor and improvement of the hydrocephalus. Note air and fluid in the epidural space.

DISCUSSION

Epidermoid tumors can be located intradurally or extradurally in any part of the cranium. They are rare and approximately consist of 0.7-1.8% of all brain tumors (1,2,14,16,18). Extradural lesions comprise only 25% of the total intracranial epidermoid tumors which may occupy the scalp and the diploe (4,7,17). They are commonly seen as an incidental finding on radiographic examination of the skull (4,17). Plain x-ray films of the skull disclose the epidermoid cyst as an osteolytic lesion with well-defined sclerotic margins (3,6).

In 1838, Mller (10) was the first to describe a diploic epidermoid cyst of the skull. According to Ciagetta et al (3) a total 223 cases had been reported up to 1990. Only a few cases of GIDET have been described in the literature and three of them are located in the posterior fossa (7,11,12) (Table 1).

caused global dementia, gait disturbance and bladder incontinence which resolved after operation.

In our case, preoperative CT scan revealed the obstruction of the fourth ventricle and moderate hydrocephalus. Tumor was not recognized at the level of the confluence of the venous sinuses. Unfortunately, we were not able to demonstrate the patency of the torcular Herophili in our case. As Rengachary et al (11) pointed out, we think that obstructive hydrocephalus and venous congestion caused by the occlusion of the major venous sinuses are primarily responsible for intracranial hypertension. Since these are very slow growing tumors, collateral venous circulation can be sufficient if torcular Herophili is open.

Total removal of the tumour with its capsule is essential, because these tumors are not radiosensitive and have a high tendency to recur.

Table 1 : GIDET of Posterior Fossa

Authors	Year	Age/Sex	Side	Size	Hydrocephalus	Occlusion of torcular Herophili	Intracranial hypertension
Rengachary et al.	1978	62/M	midline	13x13 cm	+	+	+
Rubin et al.	1989	27/M	midline	3x3.5 cm	-	+	+
Grudi et al.	1990	47/M	right	5x7 cm	-	-	-
Atabay et al	1994	51/M	left	8x8 cm	+	unknown	+

M: male, +: present, -: absent.

As in our case, GIDETs usually destroy both tables of the skull. They may expand externally and cause masses in the scalp or facial region, or internal expansion may result in the compression of intracranial contents (9,15,17). Since these are extremely slow growing tumours, there may be no clinical evidence apart from a painless swelling or palpable bone defect under the scalp for a long time. Obstructive hydrocephalus and/or obstruction of the large venous sinuses, due to intracranial hypertension can cause headache which is the most common symptom (3,4, 5,11,12). However Guridi et al (7) reported a case of a GIDET in the posterior fossa without intracranial hypertension. In their case, hydrocephalus and obstruction of the torcular Herophili had not been identified despite its large size and there was no headache or other neurological symptoms. Convulsion and focal neurologic signs may be recognized. As in our case, obstructive hydrocephalus also

These is also the rare possibility of carcinomatous degeneration (1,3,4,8,11,17,18). Total removal of the tumor can be easily accomplished and is associated with good outcome, despite its large size.

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