

## Intra-Extracranial Meningioma

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**Abstract :** Intracranial meningiomas rarely present extracranial manifestations. As new and more precise radiodiagnostic tools are designed, early diagnosed make such cases are extremely rare. We

present a case with a large extracranial extension meningioma and typical radiological findings.

**Key Words :** Intra-extracranial meningioma.

### INTRODUCTION

Although, intracranial meningiomas are common, large hyperostoting meningiomas with extracranial extension are very rare (1,2,3). Calvarial swelling or deformity bringing patients for medical attention relatively early and new diagnostic tools have decreased the number of such cases (1, 4). However, if tumours reach an enormous size when the diagnosis is made late due to the patient's psychopathology leading him to deny the existence of the obvious tumour (1, 5, 6), surgical treatment is difficult and complications are frequent (5, 7, 8, 9). We report a case of giant intra-extracranial meningioma and review the literature.

### CASE REPORT

A 29 year old man was admitted to hospital with a swelling in the frontal region, headache and forgetfulness. He had noticed the swelling three years before but refused diagnostic studies and treatment and used a cap to cover the mass. There was no history of infection or ulceration of the mass and no complaint of neurological disorder, physical examination revealed a bifrontal non-tender 15x18x6 cm. mass hard and fixed with a rubber like, soft palpated centre 3 cm. in diameter. Scalp arteries were dilated and tortuous, Neurological examination was normal. Skull x-rays showed a large frontal hyperostosis

(Fig: 1). CT scan revealed a dumbbell shaped tumour consisting of intracranial and extracranial parts (Fig: 2 a,b). On internal carotid angiography, compression of the distal anterior cerebral artery and a non-vascular area at the frontal region were seen (Fig :3). External carotid angiography, on the other hand, revealed early filling, tortuosity and enlargement in the main artery of the tumour, with excess collateralisation (Fig :4).

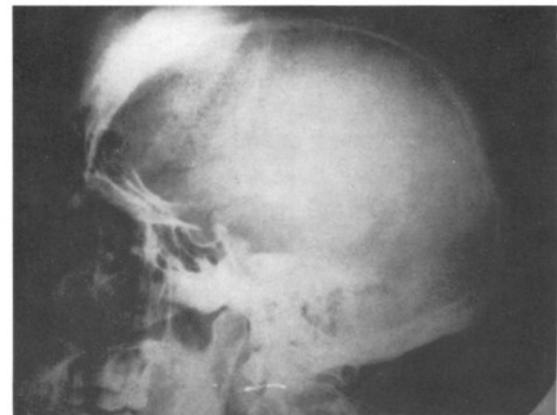


Fig. 1: Extensive hyperostosis at the frontal region on craniography.

The patient was operated through a bicoronal skin flap. The purple-gray extracranial part of the tumour was seen when the skin flap was reflected. Multiple burr holes were drilled around the

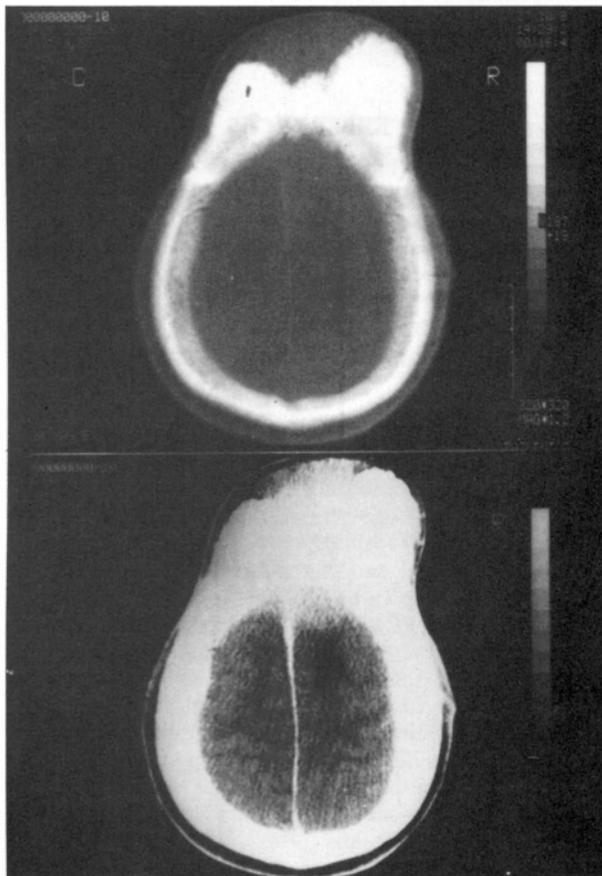


Fig. 2 a,b : CT scan showing large hyperostosis at the frontal region, intracranial and extracranial components of the tumour.

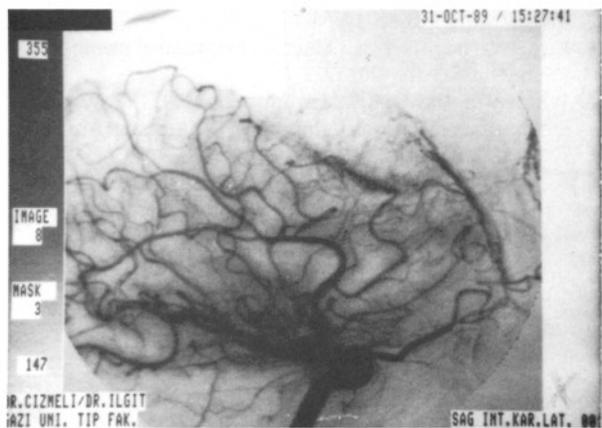


Fig. 3 : Internal carotid angiography, showing compression of the distal anterior cerebral artery.

hyperosteosing bone. After removal the bone, massive bleeding from the sagittal sinus occurred which was ligated and the intracranial part of the tumour and involved dura mater were removed. The mass including the bone weighed 600 gr. Six units

of blood were given during the operation. The postoperative course was uneventful. A cranioplasty was planned at the second operation. Histopathological examination of the tumour resulted in the diagnosis of meningioma.

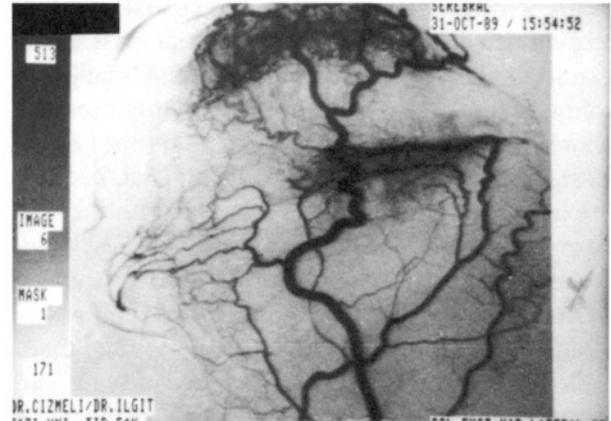


Fig. 4 : External carotid angiography, revealing early filling, tortuosity and enlargement of the main artery of the tumour and extensive collateralisation.

## DISCUSSION

Meningiomas are common intracranial tumours and make up 13-18 % of central nervous system tumours, but large hyperosteoting meningiomas and those occurring in extracranial locations are rare (10). Cushing and Eisenhardt adressed some cases of giant meningioma described in the 18th and 19th centuries as well as in prehistoric times (11). Today, as a result of modern diagnostic techniques, such cases are rarely encountered. In some hyperosteoting meningiomas, the tumour may spread extracranially. Therefore, giant meningiomas with extracranial extension are unusual.

In 1937, Davidoff removed an 835gr. tumour including hyperosteotic bone (3) and two recent cases were described by Cech et al. and Djindjian and Raulo (1,2). In the case of Cech et al. , the tumour was a parietooccipital meningioma weighing 2600 gr. (1) In the case reported by Djindjian and Raulo an 800 gr. parietal meningioma was removed in a two-stage operation after embolisation of the feeding arteries (2).

Meningiomas arise from arachnoid cap cells but the aetiology and pathogenesis of ectopic or extracranial meningiomas are controversial (13,14,15). Most authors consider that extracranial meningiomas

arise from arachnoid cell rests (14,15,16). However, transdiploic extension of the tumour and extension through a calvarial foramen are other explanations (12,13). Our case was most likely an example of transdiploic extension.

Hyperostotic meningiomas with extracranial extension usually present with a calvarial mass. If the extracranial part is large enough the mass is palpated softly. In some cases psychogenic symptoms may be prominent. Preoperative diagnosis of intra-extracranial meningiomas may be difficult, because the intracranial part of the tumour can not usually be visualised on CT. Nakagawa and Lusins advocated the use of both axial and coronal CT but were unable to demonstrate the intracranial part of the tumour in any of their 15 cases (17). In most reported cases radiological findings suggested slow-growing sarcomas. MRI may be helpful in preoperative diagnosis or if palpable extracranial soft tissue present, percutaneous needle biopsy can establish the diagnosis.

Surgical treatment of such tumours presents various problems (5,7,8,9). In the past, massive intraoperative bleeding and surface ulceration leading to infection were common cause of death. In the case of Cech et al., the patient lost 18 pints of blood during operation (1). But Djindjian preferred preoperative embolisation of the vessels to prevent massive bleeding (2). In our case, the patient tolerated the transfusion of 6 units of blood. Another bleeding source is the venous sinuses, but in most reported cases the venous sinuses adjacent to the tumour were occluded. In our case the sagittal sinus was compressed but patent and was ligated because of major bleeding. Another surgical problem is reconstruction of the scalp if a large area of skin is involved, but a myocutaneous skin flap reposition can be used (1).

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