

A Primary Fronto-Spheno-Orbital Intraosseous Meningioma Causing Exophthalmos and Soft Tissue Invasion: Case Report

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Introduction

Meningiomas, arising from arachnoid cap cells commonly located in the arachnoid layer of the meninges, are the most common tumors of the central nervous system, and represent up to 30 per cent of all tumors “Ectopic meningiomas” refers to meningiomas with no contact with arachnoid layer.

Defined recently as “extradural meningiomas”, this subtype of meningiomas is rare (<2% of all meningiomas [1], and most arises within the calvarium).

Primary intraosseous meningioma (PIOM) is a term used to describe a subtype of extradural meningiomas that arises in bone, It represents approximately two thirds of all extradural meningiomas [2]. Roshal classified in 2004 PIOM after a study concerning 15 cases of intraosseous meningiomas and showed that the osteolytic form is the rarest. Commonly, Such PIOMs are usually mistaken for primary bone tumors [3]. Indeed, while bone invasion and hyperostosis are frequent phenomena in meningiomas; in primary intra-osseous meningiomas it remains rare.

The oldest found case of primary intraosseous meningioma on a calvarium, recently reported, was recovered in Rhodes, Greece, during excavations in the Byzantine elite cemetery, dated from 12th or 14th centuries AD. It was documented by CT scan with bone window sections.

We report a case of PIOM of the fronto spheno orbital bone that infiltrated the Dura mater and soft tissues and caused exophthalmos, and we review the literature.

Case Report

A 66 year old woman with a past medical history of diabetes mellitus and past surgical history of Extracorporeal shockwave lithotripsy, living in a rural area in Tunisia presented with a left frontal hematoma after a cranial trauma after frontal impact without loss of consciousness. At that time, CT scan was not performed. 6 months later when she had noticed growth in the lesion which seemed to be occurring at an increased rate, she consulted the doctor and her chief concern was esthetic.

Indeed, the mass was ignored until it was of a considerable size as

it was painless. A CT scan was performed (**Figure 1**) showing a lytic frontal bone lesion with evidences of intraosseous speculation involving the inner and the outer cortex of the skull. an MRI was necessary but the patient refused it .A year later, when the lesion started causing exophthalmos the patient consulted again and The MR imaging (**Figure 2**) revealed a 9 × 4 × 3.5 cm sized and strongly-enhancing lesion on the left frontal bone extended to some parts of dura mater without brain invasion but it extended to the orbit with very remarkable left exophthalmos, and to the left temporal fossa. CT scan showed a destructive skull lesion including frontal left bone ,left frontal sinus, left sphenoidal sinus and bone and left infra temporal fossa. The CT showed also multiple lesions all over the skull. The patient underwent surgery for resection of the lesion. Skin was hardly reflected and the mass was adherent to it .The lesion appeared as a hard yellowish, with few locations of bleeding, that had destroyed the calvarial bone and both frontal sinuses. It was hardly suctioned and had extension to the peripheral diploe. The infiltrated bone around the lesion was resected so that a rim of healthy bone with normal strength was reached and a sinus cranialization was performed. The dura mater was infiltrated mainly in temporal without reaching the

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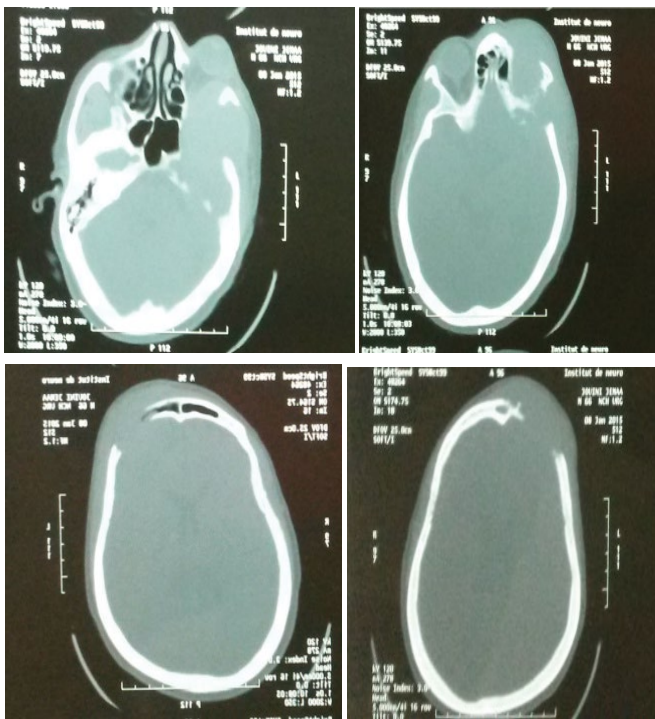


Figure 1 CT scan of the skull showing a lytic frontal bone lesion with evidences of intraosseous spiculation involving the inner and the outer cortex of the skull.

cortex excepting the apex of temporal lobe. It was compulsory to repair the dural defect after we resect the infiltrated part of it in the temporal lobe. Finally we reconstructed the calvarial defect with artificial bone material, the lesion was not easily peeled off but Most of the extradural mass and soft tissue invasion was removed simultaneously. The pathological study revealed a meningotheial meningioma of second grade.

Discussion

The proposed origin of an intraosseous meningioma may be due to a trapping of the arachnoid cells in a prior healed skull fracture, as it had been seen in our case, or to embryologic remnants of arachnoid cap cells within the developing calvaria or the optic nerve [4]. That's why most of intraosseous meningiomas are frontal. The differential diagnosis of an osteolytic lesion of the skull includes chondroma, epidermoid cyst, osteogenic sarcoma, myeloma, metastatic cancer, or fibrous dysplasia [5]. A primary intraosseous meningioma should be considered when a solitary lytic calvarial lesion is seen near the suture line or is associated with a prior skull injury.

Conclusion

PIOM is a rare tumor that raises esthetic concerns among patients as they don't cause intracranial hypertension, a history of head trauma is a key factor. Through this case, we tried to highlight the importance to think about this kind of tumors as patients may not report any disturbance unless the lesion becomes large.

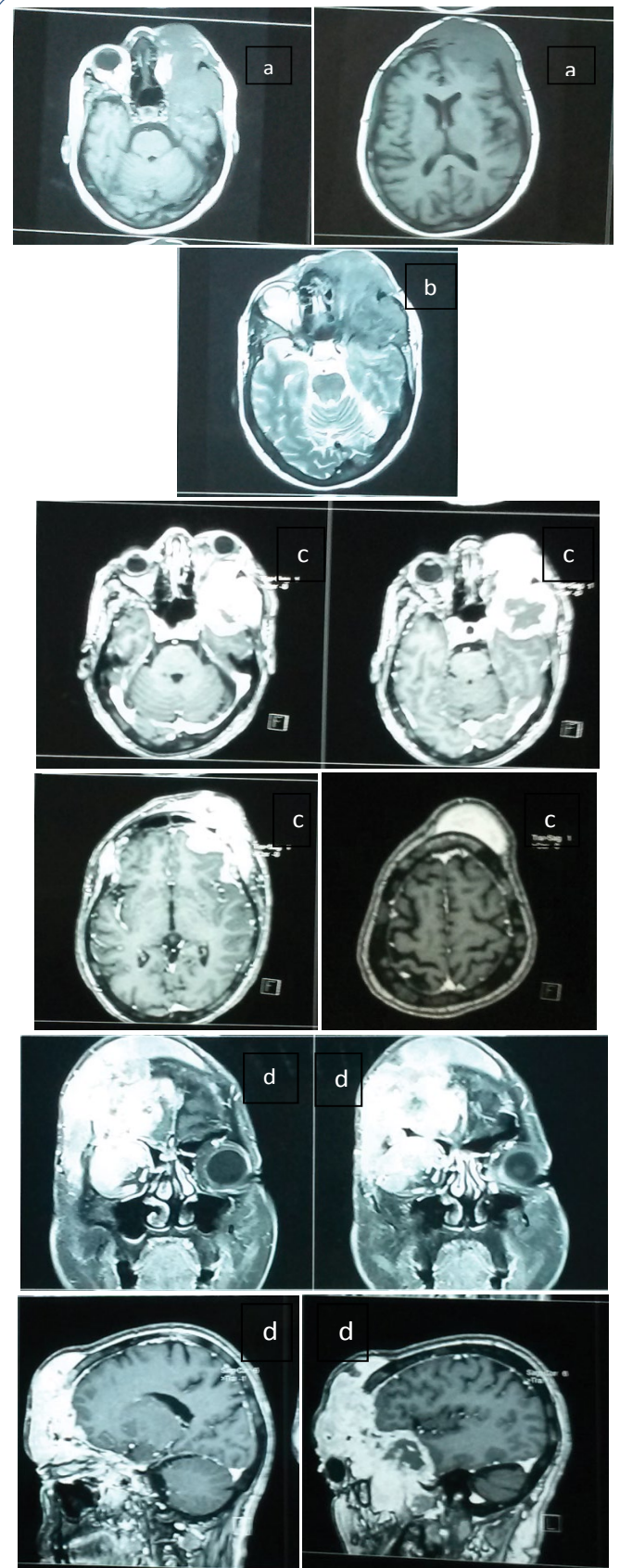


Figure 2 Cerebral MRI:T1 weighted (a), T2 weighted (b) and T1 gadolinium (c and d) 9 × 4 × 3.5 cm sized and strongly-enhancing lesion on the left frontal bone extended to some parts of Dura mater without brain invasion.

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