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Imaging Characteristics of the Spheno-Orbital Meningioma: Literature Review

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Abstract | **Case Report**

Spheno-orbital \ meningioma is a subtype of ectopic meningiomas in which meningothelial cells invade the bone wall and lead to hyperostosis [1]. They originate from the meninge of the sphenoid ridge and invade the orbit after infiltrating the underlying sphenoid wings, and sometimes extend to involve the optic canal or the sphenoidal fissure. It is typically benign, with a slow evolution, asymptomatic, and often incidentally discovered during imaging evaluation, exophthalmos remains the predominant clinical sign. We report the case of a 49-year-old female patient who presents with progressive onset left unilateral exophthalmos. Examination reveals non-reducible, painless axial exophthalmos. The aim of this case is to describe imaging characteristics of spheno-orbital meningioma and to review the literature on this subject.

Keywords: Sphenoid Ridge, Meningothelial Cells, Exophthalmos, Spheno-Orbital Meningioma.

INTRODUCTION

Intracranial meningiomas account for 20% of primary brain tumors and are typically considered benign lesions. Intraosseous locations are rare, comprising only 1 to 2% of all intracranial meningiomas, their first case was described by Winkler in 1904, with the convexity and the skull base being the most frequent locations [2]. The diagnosis of sphenoorbital meningiomas is confirmed through neuroradiological investigations, which rely on the combination of brain MRI and brain CT scan.

CASE REPORT

A 49-year-old woman, with no significant medical history, presented with left-sided exophthalmos and eyelid swelling. CT scan revealed thickening and condensation of the left sphenoorbital process with areas of lysis, associated with thickening and enhancement of the meninges adjacent to the extended condensed bone and intraorbital (intraconal lateral) (Figure 1). MRI confirmed the intraosseous left sphenoorbital lesion with hypointensity on all sequences, associated with enhancement of the adjacent meninges after Gadolinium injection (Figure 2).

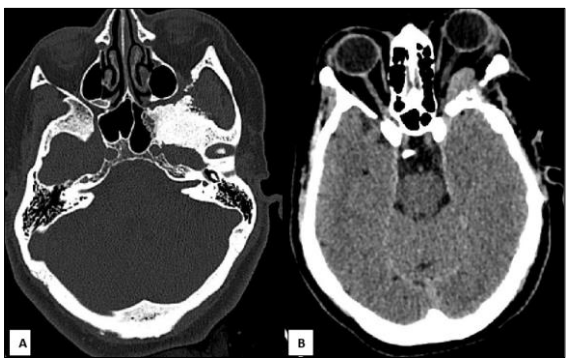

Figure 1: Axial CT scan with bone (A) and parenchymal windows (B): revealed thickening and condensation of the left sphenoorbital process with areas of lysis and intra orbital extension of tumor

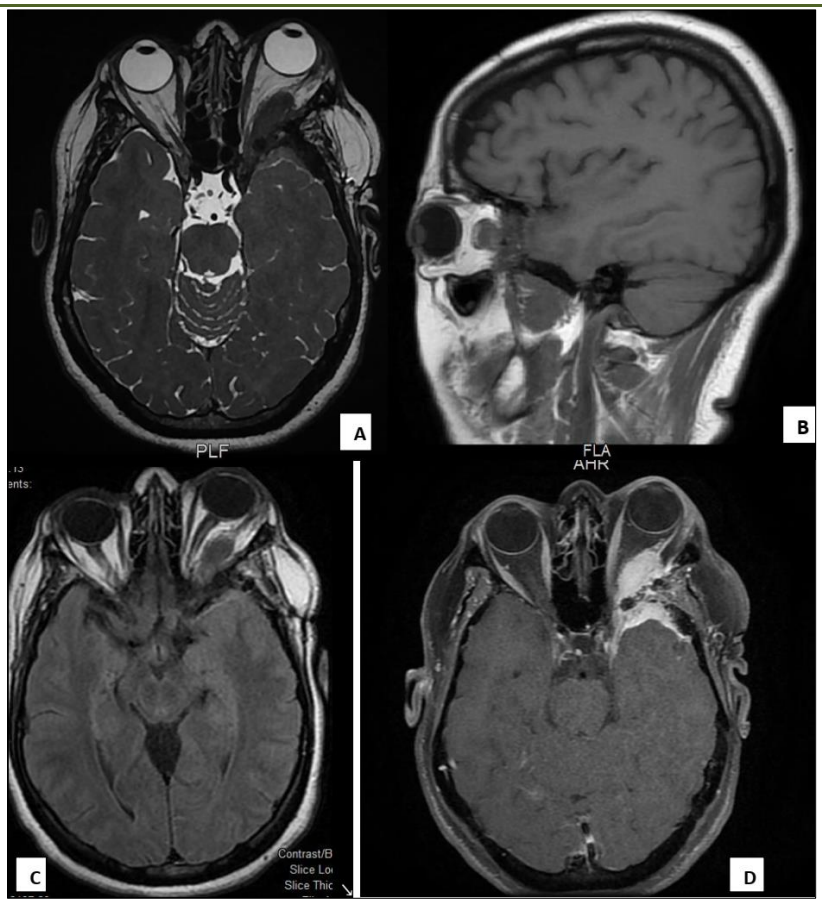

Figure 2: Axial T2 fat-suppressed sequence (A), sagittal T1-weighted MR T1(B), axial Flair (C) and Axial Gadolinium-enhanced T1 (D) MRI images ↘ showed a left sphenoorbital lesion showing hyposignal intensity on T1, T2 FAT SAT associated with enhancement of the adjacent meninges after Gadolinium injection

DISCUSSION

Sphenoorbital meningiomas have three components (osseous, meningeal, and orbital) [3, 4]. They originate from the juxtasphenoidal meninges and progressively invade the orbit by contiguity or through the optic canal, and sometimes the sphenoidal fissure. This type of meningioma is rare [1-5], with reported incidence ranging from 1% to 2% of all such tumors, The sphenoorbital and frontoparietal regions are the most common locations for intraosseous meningiomas [1-6] Meningiomas are believed to arise from arachnoid cap cells, which compose the outer layer of the arachnoid membrane, and they tend to occur more often in areas abundant with these cells. This clarifies that the majority of meningiomas adhere to the inner (meningeal) layer of the dura mater [7].

The development of this tumor within the bone would occur from ectopic meningocytes or from cells of the arachnoid capillaries that might become trapped in the cranial sutures during embryonic development, another hypothesis would also involve trapping of meningothelial cells in sutures or within cranial fracture lines secondary to trauma, however, the latter was absent in our patient. The most widely accepted theory, however, is that of tumor invasion of the bone [8, 9].

The symptoms resulting from sphenoid wing meningiomas infiltrating the bone are attributed to hyperostosis. The primary presenting symptom is typically unilateral proptosis, which is non pulsatile, non-reducible, and progresses slowly, as well as a gradual decline in visual acuity that can lead to blindness without associated neurological manifestations [1-10], which is consistent with the clinical findings in our patient.

Regarding neuroimaging evaluation, it is recommended to conduct an MR imaging study with gadolinium enhancement and fat suppression, along with a specialized oblique cut along the optic nerve. Additionally, high-resolution CT scans should be performed to precisely delineate bone involvement [8, 9].

Kim and AL [11], effectively demonstrated the key CT characteristics of en plaque meningiomas. They observed that the inward bulging of the inner aspect of the hyperostotic bone and surface irregularity of the hyperostotic bone are significant features, particularly in distinguishing other hyperostotic conditions like osteoma and fibrous dysplasia. Hyperostosis is the most common radiological image (59%), but osteolysis is also reported in 35% of cases, with a mixed image observed

in 6% of cases [1-14] .This aligns with our CT scan findings (Figure1)

Additional helpful features include intracranial changes such as brain mass effect, cerebral edema, intracranial enhancing mass, and subdural ossification. These features are also crucial for preoperative imaging assessment [12, 13].

MRI with gadolinium injection, including the T1 fat-suppressed sequence, appears to be more effective than CT in suggesting sphenoorbital meningioma. The tumor and invaded dura are most clearly visualized on contrast-enhanced MR images, also useful for assessing meningeal enhancement, particularly in tumors extending into the orbital region.

The tumor presents as a pronounced hypointensity on T1-weighted and T2-weighted sequences. Intravenous injection of gadolinium results in contrast enhancement within the intraosseous component and in the meningeal coverings adjacent to it, also allows for the detection of intracranial extension [15]. MR angiography or CT angiography offer sufficient details regarding the arterial anatomy.

Sphenoorbital en plaque meningioma is a benign tumor that, although progressing slowly, necessitates surgical intervention once diagnosed; the frontotemporosphenoidal craniectomy with resection of all invaded tissues, including the dura mater, muscle, and intraorbital tissues, along with dural and parietal reconstruction, is the treatment of choice. Adjuvant radiotherapy may be recommended in cases of incomplete resection [16, 17].

CONCLUSION

Sphenoorbital en plaque meningiomas are rare tumors that progress slowly, with predominantly ophthalmological symptoms. Their appearance on imaging is characteristic especially in MRI, with excellent topographic delineation.

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