



A CASE OF EN PLAQUE MENINGIOMA – OUT OF CLASSIFICATION

Neurosurgery

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ABSTRACT

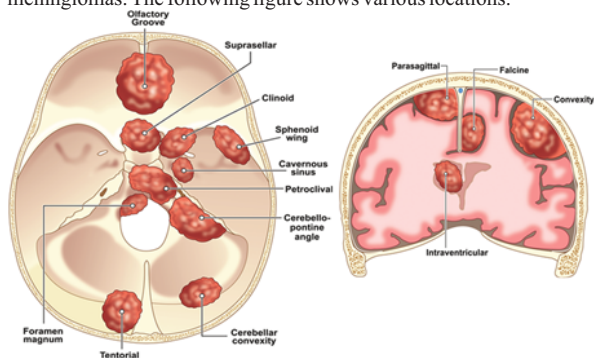
Meningiomas are the most common benign intracranial tumors, representing 36.4% of CNS tumors. Meningiomas arise from meningotheial cells of arachnoid layer. They are usually single, can be multiple and sometimes form a diffuse sheet of tumor(meningioma en plaque). **CASE DESCRIPTION:** Presenting a case of a 34 year old female who presented with proptosis, total visual loss and headache. MRI finding were suggestive of en plaque meningioma. Histopathology came out to be atypical meningioma. **CONCLUSION:** Diagnosis and surgical excision of meningioma is usually rewarding for both the patient and neurosurgeon. We describe the importance of diagnosing this rare variety of meningioma and its management challenges.

KEYWORDS

Meningioma, Atypical, En Plaque

INTRODUCTION

Meningiomas are the most common benign intracranial tumors, representing 36.4% of CNS tumors.¹ Meningiomas arise from meningotheial cells of arachnoid layer. They are usually slow growing, circumscribed (non-infiltrating), benign lesions. About 10% are histologically malignant and/or rapidly growing. They can be multiple in 8% of cases,² commonly in neurofibromatosis. Occasionally meningiomas form a diffuse sheet of tumor (meningioma en plaque). Meningioma en plaque represents a morphological subgroup defined by a carpet or sheet-like lesion that infiltrates the dura and sometimes invades the bone. The term was given by Cushing and Eisenhardt to differentiate them from the most common form called meningioma en masse. Incidence of meningiomas peaks in 5th decade of life, female to male ratio being 1.8:1. Meningiomas have been described based on location as convexity; falcine; parasagittal; different sites in anterior, middle and posterior skull base; tentorial and infratentorial including cerebellar meningiomas. The following figure shows various locations.



The clinical presentation depends upon the location of tumor, most remain asymptomatic throughout the patient's life.³ En plaque meningiomas involving the sphenoid ridge usually present with proptosis of eyeball, blurring of vision, restriction of extra-ocular movement and headache.

Contrast enhanced MRI is required for the diagnosis of meningioma and planning further management.

CASE DESCRIPTION

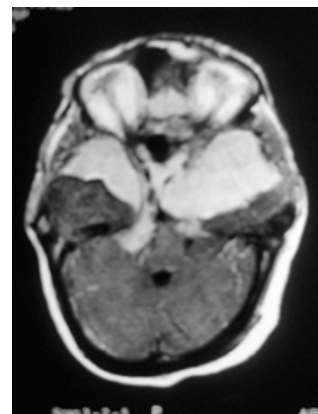
A 34 year old female patient presented with proptosis, total visual loss and headache. On examination she her extra-ocular movements were restricted.

CT done from outside was of poor quality. We did MRI brain with contrast which showed intensely enhancing diffuse skull base lesion involving the middle skull base on both sides. The lesion involved the greater and lesser wing of sphenoid, sphenoid ridge, sellar and parasellar region, also spreading to the anterior skull base involving

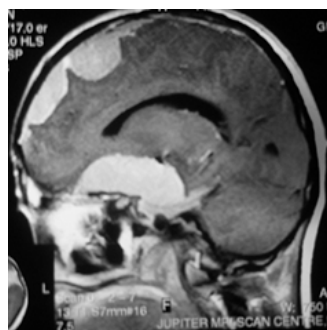
the tuberculum sellae and planum sphenoidale regions. The lesion also spread to frontal convexities bilaterally, on the left side occupying the parasagittal and falcine regions. It also involved the posterior and superior orbital regions on both sides.



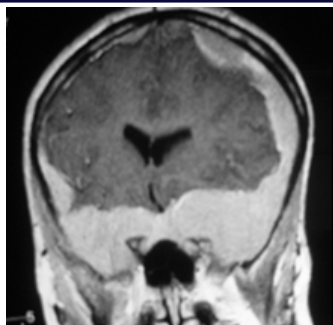
Coronal CT Brain



AXIAL MRI CONTRAST



SAGGITAL MRI CONTRAST



SAGGITAL MRI CONTRAST



CORONAL MRI CONTRAST

MANAGEMENT

This type of case is encountered once in many years and it required proper planning and counselling of relatives as the tumor involved extensive regions in skull base, convexity and parasagittal regions and thus unlike other meningiomas where gross total resection is the rule, didn't apply to this case. We did a left frontotemporal craniotomy and as we started resection of tumor, it was highly vascular and we couldn't do decompression as planned. We closed after appropriate hemostasis. Patient did well in immediate post-operative period but her lungs showed pneumonitic changes. She developed CSF leak on the 2nd post-operative day. She did well with antibiotics for few days but on 4th post-operative day her condition worsened and she developed septicemia. She succumbed to her complications on 8th post-operative day. Her biopsy report came as atypical meningioma.

DISCUSSION

En plaque meningiomas are not frequently encountered in daily practice. Meningioma en plaque is almost exclusively found in females, although some male cases have been reported.⁴ Toledo et al⁵ reported that meningioma en plaque represents about 2% of operated meningiomas. Pompili et al⁴ reported that hyperostosing meningiomas of the sphenoid ridge represent 9% of all the cases of intracranial meningiomas treated surgically.

For reasons that are unclear, meningiomas en plaque are more likely to provoke adjacent bony hyperostosis than the larger globular tumors^{4,6,7}. It is this bony hyperostosis that frequently produces the clinical signs and symptoms by pressing against adjacent structures⁶. The duration of symptoms is usually long because of the minimal discomfort produced. Proptosis is the most common presentation in patients with meningioma en plaque⁴. It is unilateral, non-pulsating, and irreducible; causing forward displacement of the eyeball. Other symptoms and signs produced by this tumor include decreased visual acuity or blindness, headache and extraocular movements disturbances⁴.

The sphenoid ridge is the most common site for meningiomas en plaque, but other locations in the convexity may be affected as well^{6,7}. Differential diagnosis of this lesion includes fibrous dysplasia, osteoma, and osteoblastic metastasis⁷.

Most authors agree with Cushing's⁶ conclusion that the infiltration of the bone by meningioma cells stimulates osteoblastic activity resulting in the hyperostosis⁴. Kim et al⁷ postulated that the new bone growth probably resulted from periosteal stimulation by tumor invasion.

In 1952, Castellano et al⁸ concluded that patients with meningioma en plaque should not be operated and surgical measures should be considered only as a last resort because of the high surgical mortality rate. Nowadays, a fronto-temporal approach with lateral cranial base resection is used to remove these tumors⁹. Removal of the orbitozygomatic bone can be added to increase exposure.

Complete removal of meningiomas en plaque is often difficult because of their extensive involvement of the sphenoid ridge and cavernous sinus. The best results are obtained with tumors located at the middle and external third of the sphenoid ridge because they do not involve the cavernous sinus or the periorbital⁴.

In this case it was not possible to go for a curative approach, to resect such a diffuse tumor involving the skull base on both sides would have been detrimental for the patient. In such cases, biopsy followed by adjuvant therapies in the form of radiation or chemotherapy or newer drugs based on meningioma receptor studies such as estrogen/progesterone receptor may form the management strategy.

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None

CONFLICT OF INTEREST

None

REFERENCES

1. Achey RL, Gittleman H, Schroer J, et al. Non-malignant and malignant meningioma incidence and survival in the elderly from 2005-2015 using the Central Brain Tumor Registry of the United States. *Neuro Oncol*. 2018. DOI: 10.1093/neuonc/noy162
2. Sheehy JP, Crockard HA. Multiple Meningiomas: A Long-Term Review. *J Neurosurg*. 1983; 59:1-5
3. Vernooji MW, Ikram A, Tanghe HL, et al. Incidental findings on brain MRI in the general population. *N Engl J Med*. 2007;357:1821-1828
4. Pompili A, Derome PJ, Visot A, Guiot G. Hyperostosing meningiomas of the sphenoid ridge—clinical features, surgical therapy, and long-term observations: review of 49 cases. *Surg Neurol* 1982;17:411-6
5. Toledo E, Dujovny M, Israeli JB. Cranial vault hyperostosis overlying meningioma "en plaque." *Isr J Med Sci* 1973;9:62-5
6. Cushing H. The cranial hyperostoses produced by meningeal endotheliomas. *Arch Neurol Psychiatry* 1922;8:139-54
7. Kim KS, Rogers LF, Goldblatt D. CT features of hyperostosing meningioma en plaque. *AJR* 1987;149:1017-23
8. Castellano F, Guidetti B, Olivecrona H. Pterional meningiomas "en plaque." *J Neurosurg* 1952;9:188-96
9. Gaillard S, Lejeune JP, Pellerin P, Pertuzon B, Dhellemmes P, Christiaens JL. Long-term results of the surgical treatment of sphenoid-orbital osteomeningioma. *Neurochirurgie* 1995;41:391-7