Primary calvarial meningioma

K Mardi, J Sharma

Citation

K Mardi, J Sharma. Primary calvarial meningioma. The Internet Journal of Pathology. 2008 Volume 8 Number 2.

Abstract

A rare case of a diffuse calvarial meningioma in a sixty year-old female is reported. The patient presented with headache and disorientation. Imaging showed thickening of the frontoparietal calvarium on left side. In relationship to the thickened bone, there was an enhancing en-plaque meningioma. Following wide surgical resection, the histological examination revealed an intraosseous meningioma are discussed and the relevant literature is briefly reviewed.

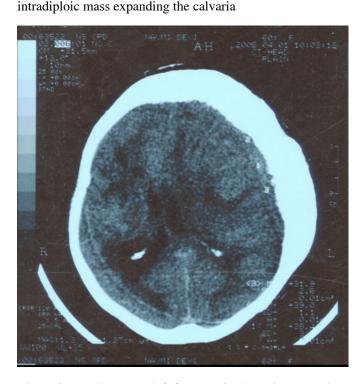
INTRODUCTION

Calvarial meningioma is a variety of 'extracranial' meningioma where the meningothelial cells primarily invade the bone and result in hyperostosis. Such meningiomas are rare when compared to the more commonly seen 'intracranial' meningiomas. We report a rare case with diffuse calvarial meningioma, which resulted in thickening of the calvarium in addition to the en-plaque meningioma.

CASE REPORT

A 60-year-old female presented with persistent left sided headaches since six months It was dull aching, centered over the left parietal region. There was no history of any trauma. The patient had no neurologic deficit. The laboratory studies were unremarkable. Radiographs of the skull revealed a well-defined area of osteolysis in the left frontoparietal region. Computerised tomography (CT) scan showed an enplaque enhancing tumour extending over left frontoparietal convexity. The calvarial bone was thickened all along the length of the tumour. The underlying dura and brain were normal. (Fig 1).

Figure 1: CT scan demonstratings a left-sided frontoparietal



The patient underwent a left frontoparietal craniotomy. The bone was thick and was extensively vascular. The involved calvarium was resected widely. The extradural meningioma was diffuse and carpet like. It was soft and greyish and only moderately vascular. This mass was removed along with the involved markedly thick dura.

Microscopic examination revealed sheets and tight whorls of meningothelial cells with round to oval vesicular nucleus,inconspicuous nucleolus and eosinophilic cytoplasm with indistinct cytoplasmic borders. The tumor cells were permeating in between the existing bony trabaculae of calvarial bone(Fig2).Occasional cells showed intranuclear pseudoinclions. There was no mitotic activity or foci of necrosis. There were large number of psammoma bodies. (Fig3).

Figure 2

Figure 2:Photomicrograph of calvarial meningioma permeating inbetween the bony trabaculae.(H&E,x20)

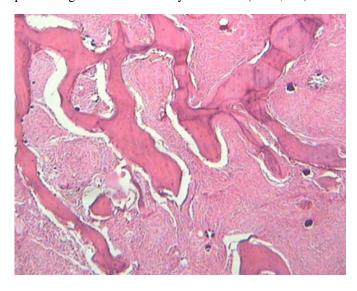
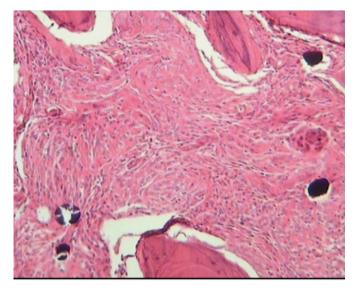


Figure 3: Higher magnification showing Psammoma bodies



DISCUSSION

Extracranial meningiomas are rare; and account for 1%-2% of all meningiomas $_{12}$. The meningiomas arising in locations outside the dural compartment have been called ectopic, extradural (epidural), calvarial, cutaneous, extracranial, extraneuraxial, or intraosseous meningiomas. To avoid the confusion in nomenclature, Lang et al $_3$ has proposed a

single term, "primary extradural meningioma" for such lesions. This term highlights the origin of these tumors as being separate from the dural coverings of any part of the brain or spinal cord and further differentiates these meningiomas from "primary intradural meningiomas," which may have secondary extracranial extensions and/or may have metastasized 3. Although some authors have emphasized that this group of meningiomas should have no connection to the dura mater or any other intracranial structures 45, other reports include tumors with intracranial growth 678

Primary extradural meningiomas are classified as purely extracalvarial (type I), purely calvarial (type II), or calvarial with extracalvarial extension (type III). According to the site of location of the tumor, Lang et al (3) further subdivided type II and type III lesions into convexity (C) or skull base (B) forms.

Many different hypotheses exist regarding the origin of primary extradural and calvarial meningioma. They are thought to arise from ectopic meningocytes or arachnoid cap cells trapped in the cranial sutures during moulding of the head at birth., Misplacement and entrapment of meningothelial cells into suture or fracture lines as a result of trauma has also been speculated as the probable cause of calvarial meningioma. 10 Involvement of multiple sutures is also reported. However, only 8% of the calvarial meningiomas are in relationship with a cranial suture. Cutaneous meningiomas could be congenital in origin where they can arise from arachnoid cell rests located in the skin as a result of defective closure of the neural tube wherein the meningeal tissue is 'pinched' off into the surface.₁₂They are also thought to arise from multipotent mesenchymal cells as a reaction to an unidentified stimulus.13 This may be relevant in our case since the meningioma was diffuse and involved the frontoparietal bone widely and extended into the overlying soft tissue. It had no special connection within the cranial sutures or the foramina of the cranial nerves.

According to the literature, 68% of the primary extradural meningiomas involved the calvaria₃. Frontoparietal and orbital regions are the most common locations for intraosseous meningiomas. They occur with approximately the same frequency in each sex ₃. Though primary extradural meningiomas occur predominantly during later decades of life, they also have a second peak incidence in younger patients (especially during the second decade). Patients with calvarial intraosseous meningiomas typically present with

slow-growing scalp swelling that may or may not be painful. They do not show any neurologic symptoms or signs, unless the lesion extends through the inner table and compresses intracranial structures. These lesions may be asymptomatic and detected incidentally 13. However Calvarial meningiomas are known to be associated with intracranial hypertension. The marked dural thickening overlying and adjacent to the tumour as well as the hyperostotic bone is attributed to cause intracranial hypertension. Dural sinus occlusion can also be an important cause of the raised intracranial tension.

The incidence of hyperostosis with meningioma has been reported to be about 4.5%.₁₄ Since Brissaud and Lerebaullet₁₅described this association in 1903, several theories have been postulated regarding its cause. The bony thickening is associated with the presence, in the medullary spaces, of clumps of tumour cells.₁₆ The cause, management and prognosis of bony hyperostosis remains controversial

Biologically, calvarial meningiomas have been observed to be benign and slow-growing. On the other hand, calvarial meningiomas are more prone to develop malignant changes (11%) compared with intracranial meningiomas (2%) ($_{3,13}$). Meningiomas presenting with scalp swelling, osteolytic skull lesions and extracranial soft tissue masses are more aggressive in nature.₁₇

Surgical resection is the treatment of choice. Although radiotherapy is advocated, it is usually not recommended unless there is evidence of rapid progression of the disease. In cases of diffuse involvement of the calvaria, a wide surgical resection is advisable whenever possible followed by a cranial reconstruction. Radiation therapy is advocated for rapidly progressive disease.

CORRESPONDENCE TO

Dr.Kavita Mardi ,MD,DNB Assistant Professor 12-A, Type V Quarters GAD Colony Kasumpti, Shimla, H.P E. mail:

kavitamardi@yahoo.co.in

References

- 1. Muzumdar DP, Vengsarkar US, Bhatjiwale MG, Goel A. Diffuse calvarial meningioma: a case report. J Postgrad Med 2001;47:116–8.
- 2. Whicker JH, Devine KD, McCarty CS. Diagnostic and therapeutic problems in extracranial meningiomas. Am J Surg 1973;123:452–7.
- 3. Lang FF, Macdonald OK, Fuller GN, DeMonte F. Primary extradural meningiomas: a report on nine cases and review of literature from the era of computerized tomography scanning. J Neurosurg 2000;93:940–50.
- 4. Hoye SJ, Hoar CS, Murray JE. Extracranial meningioma presenting as a tumor of the neck. Am J Surg 1960;100:486–9.
- 5. Crawford TS, Kleinschmidt-Demasters BK, Lillehei KO. Primary intraosseous meningioma: case report. J Neurosurg 1995;83:912–5.
- 6. Cech DA, Leavens ME, Larson DL. Giant intracranial and extracranial meningioma: case report and review of the literature. Neurosurgery 1982;11:694–7.
- 7. Geoffray A, Lee YY, Jing BS, et al. Extracranial meningiomas of the head and neck. AJNR Am J Neuroradiol 1984;5:599–604.
- 8. Oka K, Hirakawa K, Yoshida S, et al. Primary calvarial meningiomas. Surg Neurol 1987;32:304–10.
- 9. Azar-Kia B, Sarwar M, Marc JA, Schechter MM. Intraosseous meningioma. Neuroradiology 1974;6:246–53.
- 10. Turner OA, Laird AT. Meningioma with traumatic etiology. J Neurosurg 1966;24:96–8.11. Lee HY, Prager J, Hahn Y, Ramsey RG. Intraosseous
- 11. Lee HY, Prager J, Hahn Y, Ramsey RG. Intraosseous meningioma. CT and MR appearance. J Comput Assist Tomogr 1992;16:1000-1001.
- 12. Lopez DA, Silvers DN, Helwig EB. Cutaneous meningiomas: a clinicopathologic study. Cancer 1974;34:728–74.
- 13. Shuangshoti S, Netsky MG, Fitz-Hugh GS. Parapharyngeal meningioma with special reference to cell of origin. Ann Otol Rhinol Laryngol1971; 80;464-73.
- 14. Cushing H, Eisenhardt L. Meningiomas. Their classification, regional behaviour, life history and surgical end results. Charles C. Thomas: Springfield, IL; 1938
- 15. Brissaud, Lerebaoullet P. Deux cas d'hemi craniose. Rev Neurolo 1903;11:537-40
- 16. Bruner JM, Tien RD, Enterline DS. Tumours of the meninges and related tissues in Bigner DD, Mclendon RE, Bruner JM, editors. Russel and Rubinstein's pathology of tumours of the nervous system. Arnold: London; 1998. p. 79.
- 17. Muthukumar N. Primary calvarial meningiomas. Br J Neurosurg 1997;11:388- 92.

Author Information

Kavita Mardi, MD, DNB

Assistant Professor, Dept of Pathology, Indira Gandhi Medical College

Jaishree Sharma, MD

Professor and Head of Dept, Dept of Pathology, Indira Gandhi Medical College