

A Case of Primary Calvarial Meningioma, A Relatively Rare Histological Subtype

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Extracranial meningiomas are rare, the reported incidence being 1-2% of all meningiomas.¹⁵ There are inconsistencies in nomenclature and inclusion criteria of meningiomas arising in locations outside the dural compartment.⁶ They have been referred to as ectopic, extradural, calvarial, cutaneous, extraneuraxial or intraosseous meningiomas. To avoid this confusion, Lang et al has proposed a single term 'primary extradural meningioma' (PEM) for such lesions.⁶

The dura and the dural sinuses are displaced away from the inner table of the skull in these cases. Bone remodelling and calvarial thickening is frequent with these tumours. Consequently, they are classified as purely extradural (Type I), purely calvarial (Type II) or calvarial with extradural extension (Type III). According to the site of location of the tumour they are further subdivided into convexity (C) or skull base (B) forms.

We present a lady with scalp swelling and headache. CT scan of brain showed a right parietal bony lesion without any dural or parenchymal invasion. Tumour was removed. Bony gap was repaired by acrylic bone cement. Post operative recovery was uneventful. Biopsy report of tumor was primary calvarial meningioma. This is a very rare type of CNS tumor. Only very few cases were reported before.

Key words: Calvarial meningioma, Cranioplasty, Meningiomas

Calvarial thickening at the site of origin of meningioma is common. The meningotheial cells invade into the calvarium and expand the bone. However, diffuse and widespread bone thickening is rare. Most of the similar cases reported were in the region of the frontal bone and involved the orbit and presented with the symptom of proptosis. En-plaque involvement of the dura is also associated with gliomatous, carcinomatous, sarcomatous and melanomatous invasion of the meninges.

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.² Misplacement and entrapment of meningotheial cells into suture or fracture lines as a result of trauma has also been speculated as the probable cause of calvarial meningioma.¹⁴ Involvement of multiple sutures

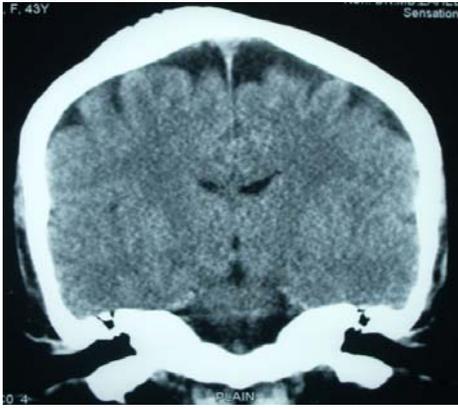


Figure 1. Preoperative CT scan of brain showing calvarial meningioma.(arrow)

is also reported.⁷ 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.⁸ They are also thought to arise from multi potent mesenchymal cells as a reaction to an unidentified stimulus.¹²

Calvarial meningiomas are known to be associated with intracranial hypertension.⁷ The marked dural thickening overlying and adjacent to the tumor 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. However, despite the extensive and diffuse frontoparietal hyperostosis, there was no evidence of raised intracranial tension in our case.

Biologically, calvarial meningiomas have been observed to be benign and slow-growing. Calvarial meningiomas are more prone to develop malignant changes (11%) when compared to intracranial meningiomas (2%).¹³ Meningiomas presenting with scalp swelling, osteolytic skull lesions and extracranial soft tissue masses are more aggressive in nature.¹⁰

Computerized tomography with bone windows is helpful and MR imaging is useful in the evaluation of the extent of extradural and calvarial involvement. Angiography is non-specific and of little value. The differential diagnosis includes plasmacytoma, chondroma, chondrosarcoma, haemangioma, myeloma, eosinophilic granuloma, aneurysmal bone cyst, metastatic cancer or fibrous dysplasia.

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.⁴



Figure 2. Intraoperative picture showing cranioplasty with bone cement

Case report

We present a 43 year old diabetic, hypertensive, and non asthmatic right handed menopausal lady who presented to us with the complaints of swelling in her right side of head for about 3 years. This swelling gradually increased in size and with in 1 and half months it became pain full. She had no history of trauma, vomiting. She had never experienced any convulsion and loss of consciousness. CT scan of brain revealed patchy irregular hyper density with mild focal expansion and speculated appearance of bone is noted in parasagittal aspect of right parietal bone (**Figure 1**).

On examination her vitals were normal. She was conscious and oriented. There was no cranial nerve deficit. Motor and sensory system was intact. No cerebellar deficit was noted. All other systemic examination reveals no abnormalities. Examination of swelling-(scalp) there was round swelling over right parietal area of the scalp. There was no punctum, no discoloration and hair loss of the skin over the swelled area. The swelling measured 5 cm × 5 cm. It was tender, bony hard non pulsatile and swelling was fixed with overlying bone.

Patient was scheduled for surgery under general anesthesia. Plan was craniectomy and cranioplasty. U shaped right fronto-parietal skin incision was marked. Skin flap was reflected keeping the base towards base. Bone over the lesion was punched out, eroded and moderately vascular. There was no dural attachment of tumour. Tumour bone (5 cm × 5 cm) was removed with the help of craniotome. A strip of tumor (1 cm × 2 cm) over the superior sagittal sinus area was drilled off. Artificial bone flap of craniectomy size was made of bone cement (acrylic bone cement) and was fixed to the craniectomy site with no. 1 vicryl. Burr hole gaps were filled with bone dust (**Figure 2 & 3**).

Microscopic examination of the specimen showed bony tissue. It revealed loose fibrovascular tissue in the bone marrow space. Some of these spaces showed proliferation of oval and spindle cells forming whorls and focal syncytial pattern. Intranuclear pseudoinclusion was identified. There



Figure 3. Post operative 3D CT scan showing good closure of bony defect

was not much of atypia or mitosis was seen. The histologic appearance was consistent with meningioma, WHO grade-I (Figure 4).

Her post operative recovery was uneventful. She was discharged on her 8th postoperative day after sutures were removed. Her headache subsided after surgery. At the time of 2 months follow up she was doing well and there was no complain of pain. Follow up CT scan of brain showed well alignment of cranioplasty flap without any bony gap or new tumour.

Discussions

We are presenting a case of primary calvarial meningioma that underwent craniectomy and cranioplasty. Only few cases have been reported about primary calvarial meningioma so far.

Muzumdar et al described a sixty three year old female complained of painful and progressive proptosis and chemosis of the left eye for six months.⁹ CT scan showed an en-plaque enhancing tumor extending over both frontoparietal convexity and into the superolateral aspect of the left orbit displacing the eyeball inferiorly. He underwent a left frontoparietal craniotomy with wide resection of the involved calvarium. The extradural meningioma was also removed along with the involved markedly thickened dura.

Kavita et al described a 60 year old female presented with persistent left sided headache for six months without any neurological deficit.⁵ CT scan showed an enhancing tumor extending over left frontoparietal convexity with thickened calvarial bone all along the length of the tumor. The location of our case was right parietal parasagittal region. Like the tumor of Kavita our tumor didn't invade dura.

Khawaja et al described a 50-year-old male patient presented with headache and gradually expanding scalp mass over a few months.³ Imaging showed localized skull expansion at the vertex, and osteolysis of the inner and outer plates of the skull with brain tissue herniation through

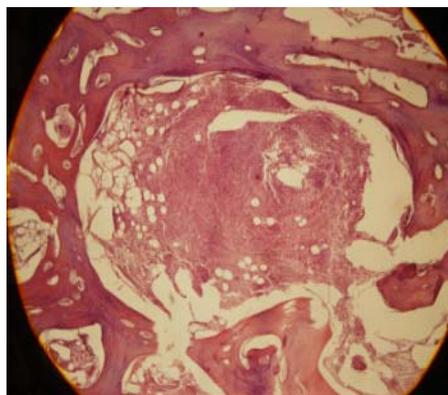


Figure 4. Histological picture showing diploic meningioma

a defect in the inner plate. The patient underwent wide surgical excision, dural repair, and mesh cranioplasty, following which his headaches ceased. Histological examination showed WHO grade I meningioma. Our patient also presented with gradually increasing painful swelling in right parietal region without any neurological deficit. Our case also reported as WHO grade- I meningioma.

Nil et al described a 44-year-old man presented with gradually increasing painless hard scalp swelling of the right frontoparietal region for 8 years¹¹. The patient had no neurologic deficit. CT revealed a right-sided frontoparietal intradiploic contrast enhancing mass expanding the calvaria with prominent bone destruction extending through the skull defect both intra- and extracranially. The tumor and the surrounding bone were removed, followed by cranial reconstruction. There was no intradural extension of the lesion. Histologically, the tumor was diagnosed as chordoid meningioma. In our case presentation, radiological findings of the lesion were same and also did not require dural excision.

Abdolreza et al described a 62-year old male farmer presented with a soft fluctuating enlarging mass in the left fronto-parietal region over last 8 months¹. The patient had undergone scalp radio therapy for treatment of ringworm about 45 years ago. Skull x-rays and CT showed a lytic lesion of the skull in the left fronto-parietal region. MRI of head showed an extradural intradiploic enhancing mass lesion without intracranial extension. The patient underwent surgery for resection of the mass lesion. It was easily 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. The dura was intact and the lesion was easily peeled off. Pathologic examination of the lesion revealed fibrillary meningioma with areas of syncithial differentiation. However in our case the lesion was intradiploic hard one without dural invasion.

After removal of the tumour, bony gap was successfully repaired by acrylic bone cement. Follow up CT scan of brain showed well aligned replacement of bone graft by bone cement. Other criteria of the primary calvarial

meningioma showed a fair similarity with other published cases. We present the rare case for reminding the classification of meningiomas where non-dural attached primary tumours presents a very small group. Cranioplasty by bone cement also showed a good result for primary calvarial meningioma.

References

1. Abdolreza Sheikhrezaie, Ali Tayebi Meybodi, Mohammad Hashemi et al. Primary intraosseous osteolytic meningioma of the skull: a case report. **Cases Journal** 2:7413, 2009
2. Azar-Kia B, Sarwar M, Marc JA, et al. Intraosseous meningioma. **Neuroradiology** 6:246-253, 1974
3. Darweesh Al-khawajaa, Rajmohan Muralib. Primary calvarial meningioma. **Journal of Clinical Neuroscience** 14:1235-1239, 2007
4. Goel A, Mehta A, Gupta S. Unusual mode of spread and presentation of meningioma: A case report. **Neurol India** 47:311-313, 1999
5. K. Mardi, J. Sharma. Primary calvarial meningioma. **The Internet Journal of Pathology** 8:2, 2009
6. Lang FF, Macdonald OK, Fuller GN, et al. Primary extradural meningiomas: a report on nine cases and review of literature from the era of computerized tomography scanning. **J Neurosurg** 93:940-950, 2000
7. Lee HY, Prager J, Hahn Y, et al. RG. Intraosseous meningioma. CT and MR appearance. **J Comput Assist Tomogr** 16:1000-1001, 1992
8. Lopez DA, Silvers DN, Helwig EB. Cutaneous meningiomas- a clinicopathologic study. **Cancer** 34:728-744, 1974
9. Muzumdar DP, Vengsarkar US, Bhatjiwale MG et al. Diffuse Calvarial Meningioma: A Case Report. **Journal of Postgraduate Medicine** 47:116-118, 2001
10. Muthukumar N. Primary calvarial meningiomas. **J Neurosurg** 11:388- 392, 1997
11. Nil Tokgoz, Yusuf A. Oner, Memduh Kaymaz et al. Primary Intraosseous Meningioma: CT and MRI Appearance. **AJNR Am J Neuroradiol** 26:2053–2056, 2005
12. Shuangshoti S, Netsky MG, Fitz-Hugh GS. Parapharyngeal meningioma with special reference to cell of origin. **Ann Otol Rhinol Laryngol** 80:464-73, 1971
13. Shuangshoti S. Primary meningiomas outside the central nervous system. In: Al-Mefty O, editor. **Meningiomas**. New York: Raven press: 107-128, 1991
14. Turner OA, Laird AT. Meningioma with traumatic etiology. **J Neurosurg** 24:96-98, 1996
15. Whicker JH, Devine KD, McCarty CS. Diagnostic and therapeutic problems in **extracranial meningiomas**. **Am J Surg** 123:452-457, 1973