Intracranial-extracranial meningioma mimicking an aggressive skull bone tumor

Amit Agrawal1, K.V. Murali Mohan2, Vissa Shanthi3, Umamaheshwar Reddy4

Narayana Medical College Hospital, Chinthareddypalem, Nellore, Andhra Pradesh, India
1Professor of Neurosurgery, Department of Neurosurgery
2Professor of Pathology, Department of Pathology
3Associate Professor of Pathology, Department of Pathology
4Assistant Professor of Radiology, Department of Radiology

Abstract
Intracranial meningiomas with extracranial extension are rare lesions with only few reported cases in literature. In present article we report a case of 62 year male patient presented with progressive swelling over right side frontal region of 2 year duration. In our case the tumor was in close proximity to right fronto-parietal suture and there was a possibility that suture line could have paved the way for its extracranial spread and also there was evidence of transdiploic extension on histopathological examination.

Key words: Meningioma, head injury, extracranial meningioma, intracranial-extracranial.

Introduction
Meningioma account for 24-30% of primary intracranial tumors and typically arises in proximity to the meninges (1, 2, 3). Intracranial meningiomas with extracranial extension are rare lesions with only few reported cases in literature (3-8). In present article we report a case of intracranial and extra-cranial meningioma.

Case report
A 62 year male patient presented with progressive swelling over right side frontal region of 2 year duration. Weakness of left upper and lower limbs of one month duration. There was history of headache and vomiting for last 3-4 days. Local examination revealed a non-tender, hard swelling over right fronto-parietal region, non-pulsatile, with bruit. Skin over the swelling was healthy and there were no prominent vessels. There was history of trauma at the same site about 15 year back. His general and systemic examination was unremarkable. There was mild upper motor neuron type of weakness of left facial nerve. Other cranial nerves including fundus were normal. He was conscious, alert and oriented to time, place and person. There was grade 4/5 weakness of left upper and lower limbs. Deep tendon reflexes were exaggerated on left side and were normal on right side. Planter was extensor on left side and was flexor on right side. CT scan brain plain with bone window revealed a large tumor consisting of intracranial and extracranial parts with erosion of the calvaria involving right frontal and parietal and crossing the coronal suture on right
side (Figure 1 A-F). The tumor was invading the parenchyma and there was significant peri-lesional edema. Based on the imaging findings a high grade malignant lesion was considered. The patient underwent right fronto-parietal craniotomy and total excision of the tumor including excision of the involved bone. The tumor was attached to the dural and infiltrating the bone. It could be easily separated from the underlying brain parenchyma. Histopathological examination of the tumor confirmed the diagnosis of meningioma (Figure 2 and 3). The patient recovered well and the postoperative course was uneventful.

Discussion

The reported incidence of intracranial meningiomas with extracranial extension is up to 20% of cases (9, 10).

There is strong evidence there is an elevated risk brain tumor after head trauma particularly for meningiomas in males (1, 11-16). Many explanations have been put forward to explain extracranial spread of tumor and the extracranial extension in meningiomas may occur by direct extension of an intracranial meningioma through a natural, traumatic, or iatrogenic skull defect (17, 18). This may occur through skull foramina, through the supraorbital fissure (into the orbit), through the cribriform plate (into the nasal cavities and nasopharynx), through floor of middle cranial fossa (into paranasal sinuses and pterygoid region) and extracranial extension through the suture line of the skull (4, 9, 10, 19-23). In a case report the transdiploic extension was suggested as the underlying mechanism for the extracranial spread of the meningioma (8).
Figure 2

(A) Spindle shaped tumor cells forming whorling pattern (H&E X100), (B) spindle shaped tumor cells with areas of hyalinization (H&E X100), (C) tumor cells infiltrating the skeletal muscle bundles and congested blood vessels (H&E X100) and (D) spindle shaped tumor cells with psammoma body (H&E X100)

Figure 3

Showing the bony trabeculae with interstitial tissue showing infiltrating tumor cells and psammoma bodies (H&E X100)

In our case the tumor was in close proximity to right fronto-parietal suture and there was also a possibility that suture line could have paved the way for its extracranial spread and also there was evidence of transdiploic extension on histopathological examination. Clinical symptoms are usually non-specific and can be according to the site of involvement (20). If the extracranial component is large enough and there is extensive hyperostosis the lesion can be palpable through the scalp (18). Both MRI and CT will help to suggest the extent of the lesion and this can be
confirmed by FNAC prior to surgical intervention (4, 7, 24). Whenever possible the complete excision of the tumor is the treatment of choice as this has been shown to be associated with best long-term outcome compared with subtotal excision (19, 23, 25, 26 27 28, 29). The recurrence rate ranges from 7% to 84% (30, 31) and the recurrences usually develop at the primary site of lesion and probably represent the residual disease rather than true recurrence (27, 30, 31).

Address for correspondence:
Dr. Amit Agrawal
Professor of Neurosurgery, Department of Neurosurgery, Narayana Medical College Hospital, Chinthareddypalem
Nellore-524003
Andhra Pradesh (India)
Email: dramitagrawal@gmail.com
                      dramit_in@yahoo.com
Mobile: +91-8096410032

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