Case Report

Giant calvarial hyperostosis with biparasagittal en plaque meningioma

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We report a patient with an uncommon presentation in the form of massive bilateral calvarial hyperostosis with biparasagittal en plaque meningioma. The tumour was removed by bilateral fronto-parieto-occipital craniotomies. The patient was subjected to post operative radiotherapy to reduce the chances of recurrence. The management of such a case is a surgical challenge.

Key words: Calvarial hyperostosis, parasagittal meningioma en plaque, tumour invasion.

Introduction

The incidence of hyperostosis with meningioma has been reported to be about 4.5%. Since Brissaud and Lerebaullet described 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. The cause, management and prognosis of bony hyperostosis remains controversial.

Hyperostosis associated with meningioma en plaque occurs commonly either along the sphenoid ridge or the convexity. We report an unusual patient who presented with extensive bilateral calvarial hyperostosis in association with an en-plaque biparasagittal meningioma.

Case Report

A 32 year old man was admitted with history of headache and numbness of right leg for one and a half years. His left leg was weak since childhood due to poliomyelitis. He had a diffuse symmetrical swelling over the vertex of head extending from the frontal to the occipital region. There was a right upper motor neuron facial paresis and weakness of right lower limb (grade 2/5).

CT scan and MRI of the head showed a large enhancing tumour in biparasagittal region present bilaterally extending from frontal to occipital region with overlying hyperostosis. The hyperostotic bone was more than 5 cm thick, extending from the region of coronal suture to the occipital region and across the whole expanse of the skull. The hyperostotic bone was much larger and thicker than the underlying soft tissue mass. Digital subtraction angiography (DSA) demonstrated non filling of the anterior and middle part of the superior sagittal sinus and the venous system was depressed inferiorly by the tumour mass.

At surgery, a bicoronal skin flap was taken extending from one pinna to the other across the vertex. Bilateral frontoparieto occipital craniotomies were performed leaving a 3-4 cm strip of bone in midline over the region of superior sagittal sinus. The bone was extremely hard in the paramedian region and paramedian gutters were made bilaterally with the help of a drill. Laterally, the burr holes were placed in the normal temporal bone bilaterally below the superior temporal line and the 2 bone flaps were lifted off the dura, one by one. The midline bone was nibbled.

This bone was relatively soft and it was difficult to separate it from the infratentorial tumour mass. The tumour was removed by bilateral fronto-parieto-occipital craniotomy. The patient was subjected to post operative radiotherapy to reduce the chances of recurrence.

Key words: Calvarial hyperostosis, parasagittal meningioma en plaque, tumour invasion.
completely from the dura. Dura was opened bilaterally with dural flaps based on midline. The arachnoid plane was developed between the tumour and brain and tumour removal achieved with help of intratumoural decompression with an ultrasonic aspirator. In the anterior and middle part, tumour was excised along with attached dura. Posteriorly, the dura in the region of superior sagittal sinus was left intact and the tumour was removed from below the dura. A small part of tumour in the posterior occipital region in midline attached to the dura in region of superior sagittal sinus was left.

Dural closure was done with a fascia lata graft. The inner tables of the bone flaps were gradually shaved off to reduce the bulk. The midline bone was completely nibbled off and sent for histopathological examination as it appeared softer and spongy possibly because of tumour invasion. The frontoparietal bone flaps were replaced and incision closed.

The patient received about 8 units of blood in the perioperative period.

He had quadriparesis in the immediate postoperative period, but started improving in 24-48 hours. Postoperative X-ray skull [Figure 3] showed the bifrontoparieto-occipital bone flaps which had been replaced after debulking. Postoperative CT demonstrated satisfactory tumour removal. The upper limbs function returned to normal followed by right lower limb and at 3 months follow-up, he had significant improvement and was walking with support. His upper limbs had normal power and right leg had grade 3-4/5 power. To reduce the chances of recurrence and for the small residual tumour, he was subjected to postoperative radiotherapy. Post operative MRI done 1 month and again at 8 months demonstrated a small residual tumour in the posterior part in midline near the posterior superior sagittal sinus [Figure 4a-b]. He is now on 1 year follow up and remains stable with no evidence of tumour growth on MRI.

Histopathological examination showed meningothelial meningioma with bony invasion [Figure 5].

**Discussion**

The incidence of hyperostosis reported in literature is variable, Cushing and Eisenhardt\(^4\) reported this in 25% of their patients with meningioma en plaque, while others have reported an incidence of up to 49%.\(^5\) Meningiomas en plaque are more likely to provoke adjacent bony hyperostosis than the larger globular...
tumours. MEP provokes an adjacent bony hyperostosis that is often disproportionately greater than the underlying tumour. In an en plaque meningioma, the dural tumour may be less important than the hyperostosis, which may be responsible for the entire clinical syndrome. In the present case, there was extensive bilateral calvarial hyperostosis with a biparasagittal en plaque meningioma. The patient had symptoms because of both the hyperostosis as well as the underlying carpet like extent of the tumour along the superior sagittal sinus and brain compression.

It is generally accepted that in most patients the hyperostosis represents a true infiltration of the bone by the tumour: Echlin was the first to suggest a direct association between hyperostosis and tumour invasion of bone. Pieper et al. after histological examination concluded that hyperostosis, when present in association with meningiomas of the cranial base, represents a direct tumour invasion of the bone. It must also be noted that localized thickening of the bone, especially of the inner tables, is quite often not associated with invasion by the tumour.

Most of the cases of hyperostosis involve the frontal bone and the orbit. However, diffuse and widespread bone thickening, like in the present case, is rare. A similar case has been reported recently.

The wide extent of the tumour, i.e., bilateral extensive calvarial hyperostosis with invasion of the superior sagittal sinus and associated diffuse bilateral en plaque growth with compression of the underlying brain on both sides posed a formidable surgical challenge. Wherever possible, it is recommended that the entire involved bone should be resected to prevent recurrence. The bilateral frontoparieto-occipital flaps had a thick bone and the authors were reluctant to discard these as this would have left a bilateral huge cranial defect involving the frontoparietal occipital regions which would have required a massive and difficult reconstruction. Therefore it was thought prudent to shave off the inner surface of both flaps and replace them. Any way, we had left a little bit of tumour along the posterior aspect of the superior sagittal sinus. The receptor status was determined in the excised tissue. Both the estrogen and progesterone receptors were negative. The patient was given postoperative radiotherapy to reduce the chances of recurrence. Since this patient had significance infiltration of bone, demonstrable both at surgery and on histological examination, the possibility of recurrence was significant. We were quite reluctant to discard the bone flaps, which were almost like bilateral hemianiotomies, at the time of surgery or the possibility of having to discard them later, in the event of recurrence, as it would have entailed a formidable reconstructive procedure. Therefore, the decision for radiotherapy was taken which has been recommended in such situations. In the present case, the location of the tumour, involvement of posterior part of sagittal sinus, along with involvement of practically the whole calvarium precluded total excision. We believe that the desire to totally extirpate the tumour has to be tempered keeping in view that surgery should avoid causing permanent neurological deficit.

References


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