

A Rare Case of Compartmentalized Presentation of Extradural Meningioma

Abstract

Meningioma is a common benign intracranial neoplasm. The incidence of its extracranial extension is rare. Of the meningioma types, meningothelial variety is the most common histological type. Authors report a case of compartmentalized presentation of extradural meningioma in a 35-year-old male who presented to us with a frontal region swelling (exostosis). The patient was evaluated with computed tomography scan of the brain. At surgery, he was found to have compartmentalized presentation of the scalp, calvarium, and dura which we would like to report in this case.

Keywords: *Compartmentalized meningioma, extradural meningioma, rare presentations of meningioma*

**Sarang Gotecha,
Prashant Punia,
Ashish Chugh**

*Department of Neurosurgery,
Dr. D. Y. Patil Medical
College and Hospital, Pune,
Maharashtra, India*

Introduction

Meningioma is a common benign intracranial neoplasm that arises typically in proximity to the meninges.^[1] Cap cells of arachnoid layer are ‘thought’ to be the layer of origin histologically.^[2] Meningiomas are commonly found along whole of neural axis and can be found commonly at overlying surface of the brain or skull base, as well as uncommonly at intraventricular, intraspinal, intraparenchymal, or intraosseous location. Extra-neuraxial meningiomas are rare and may be found as an extracranial tumor in the sinonasal tract, temporal bone, and scalp. This is a report of a compartmentalized, extracranial “carpet” meningioma involving the extracranial soft tissue and calvarium, wherein the patient presented to us with a frontal region bony swelling (exostosis).

Case Report

A 35-year-old male presented to our institute with a right frontoparietal scalp swelling which was painless and gradually progressive for the past 4 years. The patient did not have any neurological symptoms such as headache, fever, and vomiting. He had no cranial nerve involvement and behavioral or urinary symptoms. There was no history of trauma before the development of swelling. On clinical examination, there

was a large immobile, bony swelling over the right frontoparietal region which was extending across the midline. There was evidence of dilated tortuous vessels in the scalp over and around the swelling. The swelling was nontender, and the overlying skin appeared normal in appearance but tense and unpinchable.

Radiology

A preoperative computed tomography (CT) scan was done which showed an expansile osteoblastic/sclerotic lesion with lobulated outline measuring approximately 92 mm × 82 mm × 28 mm involving right frontoparietal bone with obliteration of diploic space and diffuse enhancing overlying extracranial soft tissue component. Differential diagnoses included calcified bony cavernous hemangioma, intraosseous meningioma, fibrous dysplasia, and Paget’s disease.

CT angiography showed a right frontoparietal osseous lesion nourished by the right superficial temporal artery. The mass did not receive supply from the internal carotid artery. The superior sagittal sinus was found to be in close contact with the mass lesion, which was predicted to be an intraoperative challenge.

Keeping the CT differentials in mind, the patient was taken up for surgery. First, occlusive ligatures were taken around the superficial temporal and supraorbital

Submitted: 14-Jan-2021

Revised: 09-May-2021

Accepted: 15-May-2021

Published: 28-Oct-2021

Address for correspondence:

*Dr. Prashant Punia,
Department of Neurosurgery,
Dr. D. Y. Patil Medical
College and Hospital, Pimpri,
Pune, Maharashtra, India.
E-mail: getdprashant@gmail.
com*

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

For reprints contact: WKHLRPMedknow_reprints@wolterskluwer.com

How to cite this article: Gotecha S, Punia P, Chugh A. A rare case of compartmentalized presentation of extradural meningioma. *Clin Cancer Investig J* 2021;10:254-6.

Access this article online

Website: www.cci-j-online.org

DOI: 10.4103/cci-j.cci-j_6_21

Quick Response Code:



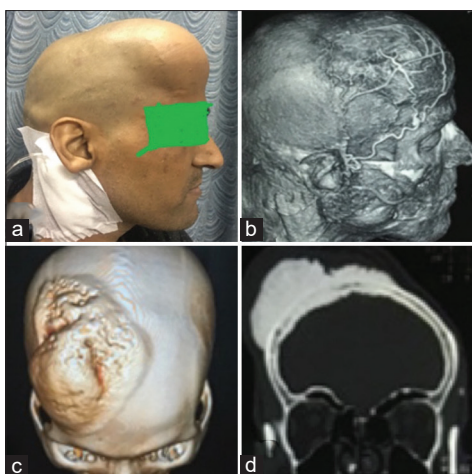


Figure 1: (a) Clinical photograph showing swelling in patient's right frontal region. (b) Computed tomography brain soft tissue reconstruction image showing hypervascular tumour with blood supply from superficial temporal artery. (c) Computed tomography brain reconstructed image showing bony swelling and sclerotic bone around the swelling. (d) Computed tomography brain, bony window showing bony swelling and sclerotic bone around the swelling

arteries. A bicoronal skin incision was taken over the tumor and flap was reflected. Intraoperatively, there was evidence of firm to hard vascular soft tissue lesion over the bone, which was adherent but separable from the underlying irregularly surfaced bone. The bone was sclerotic; however, in the bone, there was no delineation between normal and abnormal bony tissue. An intraoperative frozen section was suggestive of meningioma. This was followed up with a frontal craniotomy. Underlying dura was found to be thickened but separable from the overlying bone. The dura which was bluish grey, although thickened, was found to be friable on excision, and part of dura inside the craniotomy margins was excised and sent for biopsy.

Histopathology of the excised extracranial soft tissue component, bone, and dura revealed it to be meningotheial meningioma (WHO Grade 1). Thus, a diagnosis of extracranial meningotheial meningioma was made. Postoperative period was uneventful, and follow-up scans showed complete resolution of the lesion with no recurrence of tumor. Immunohistochemistry report for the tumors showed tumor cells positive for vimentin and focally positive for epithelial membrane antigen (EMA). Cytokeratin was negative. MIB-1 labeling index was 2%–3%.

The patient was closely followed up at intervals of 6 months, 1 year, and 2 years and has been asymptomatic since [Figures 1-2].

Discussion

Meningiomas are benign intracranial neoplasms with neural crest origin (arachnoid cap cells). They commonly affect middle-aged female population and account for 24%–30% of primary intracranial neoplasms.^[2]

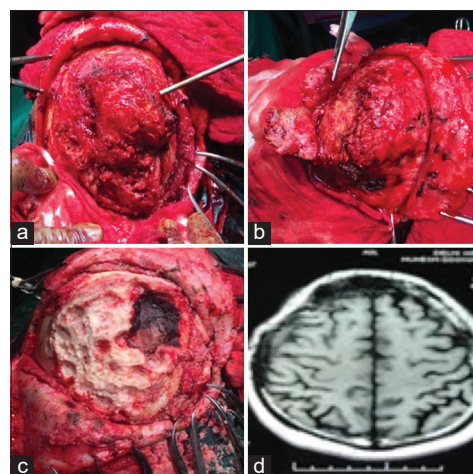


Figure 2: (a) Intraoperative photograph showing tumour and surrounding sclerotic bone. (b) Intraoperative photograph showing soft tissue overlying the bony tumour. (c) Intraoperative photograph showing excision of tumour with dura and flattening of sclerotic bone around the tumour. (d) Postoperative magnetic resonance brain showing no evidence of residual tumour

Meningiomas rarely show extracranial extension and those which arise outside the skull are generally benign lesions in spite of the seemingly aggressive behavior. Shuangshoti^[3] have reported a prevalence of extracranial extension with primary intracranial meningiomas to be 9%–20%. Common locations are orbit, outer skull table, and scalp. Extracranial spread occurs in many ways. Cap cells of arachnoid layer are ‘thought’ to be the layer of origin histologically

The different mechanisms postulated for the spread are:

1. The presence of arachnoidal cells in the sheaths of nerves or vessels where they emerge through the skull foramina
2. Detachment and entrapment of displaced pacchionian bodies during embryologic development in an extracranial location
3. Displacement of arachnoid location by a traumatic event
4. Origin from undifferentiated or multipotential mesenchymal cells.^[4]

The clinical presentation depends on the size and location of tumor, and the same may present as intracranial hypertension which can be attributed to the marked dural thickening overlying the tumor and/or dural venous sinus occlusion. Although likely, the same was not seen in our case where the only presenting complaint of the patient was a visible scalp swelling. Mazumdar *et al.*^[5] reported a similar case wherein no symptoms of intracranial hypertension were seen in spite of extensive and diffuse frontoparietal hyperostosis. They further asserted that such features were distinctive and not recorded previously in the literature which makes the case even rarer.

The mainstay of treatment is complete surgical excision of lesion along with involved soft tissue and dura. Although

radiotherapy is recommended, it is usually not advocated unless there is a rapid progression of the disease. In case of diffuse involvement of calvaria, a wide surgical resection is advised whenever possible followed by a cranial reconstruction. If subtotal resection is performed due to the involvement of structures such as orbit, paranasal sinuses, or dura as seen in our case, the lesion can be followed up at regular intervals with appropriate imaging studies.^[5]

Histologically, meningothelial variety shows hyaline inclusions on electron microscope which are periodic acid–Schiff-positive proteinaceous structures. High MIB-1 levels are suggestive of rapid growth and aggressive nature of tumor and predict possibility of recurrence. Another test which describes the aggressive nature of the tumor is by array-comparative genomic hybridization wherein complex cytogenetic profile and progression-associated chromosomes are studied, which gives an idea of the potential targets for drug delivery and therapeutic intervention in the future. Other markers such as EMA, vimentin, PR, and mitosis-specific antibody against phosphohistone-H3 aid in diagnosis and prognostication of meningiomas.^[6] MIB labeling index was low in our case, and thus, excellent long-term prognosis can be expected with limited chances of recurrence.

Conclusion

Meningioma is a benign intracranial neoplasm with rare extracranial invasion. Extracranial variants of this pathology are a rarity. Complete excision of tumor is the treatment of choice which shows good prognosis; however, subtotal excision with flattening of the cranial vault is an acceptable treatment modality in cases of diffuse “carpet” lesions. This modality has to be supplemented strongly with regular follow-up and serial radiology. The rarity of this case and the atypical compartmentalized presentation

prompted us to report the same, underlining the importance of awareness of this entity while dealing with calvarial lesions, as being forewarned is being forearmed.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Bassiouni H, Asgari S, Hübschen U, König HJ, Stolke D. Dural involvement in primary extradural meningiomas of the cranial vault. *J Neurosurg* 2006;105:51-9.
2. Drummond KJ, Zhu JJ, Black PM. Meningiomas: Updating basic science, management, and outcome. *Neurologist* 2004;10:113-30.
3. Shuangshoti S, Panyathanya R. Ectopic meningiomas. *Arch Otolaryngol* 1979;98:102-5.
4. Panjvani SI, Gandhi MB, Sarvaiya AN, Chaudhari BR, Gupta GS. An extracranial invasive meningioma mimicking malignant bone tumor - “carpet meningioma”. *J Clin Diagn Res* 2013;7:1159-62.
5. Mazumdar DP, Vengsarkar US, Bhatjiwale MG, Goel A. Diffuse calvarial meningioma: A case report. *Postgrad Med* 2001;47:116.
6. Rushing EJ, Bouffard JP, McCall S, Olsen C, Mena H, Sandberg GD, *et al.* Primary extracranial meningiomas: An analysis of 146 cases. *Head Neck Pathol* 2009;3:116-30.