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Case Report

Atypical intradiploic meningioma: A case report and review of the literature

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ABSTRACT

Background: Intradiploic meningiomas with osteolytic bony changes are rarely reported in the literature. Intradiploic meningiomas are usually slow-growing benign lesions but atypical histopathology predicts aggressive behavior. Atypical intradiploic meningiomas (WHO Grade II) have some controversies in the management which are highlighted in this article.

Case Description: A 40-year-old male, with a history of trauma to the head 12 years back, presented with a hard, slow-growing painless swelling exactly at the site of trauma. On imaging, lesion was intradiploic one with osteolytic margins and homogeneously enhancing on contrast magnetic resonance imaging. Biopsy was that of atypical meningioma (WHO Grade II).

Conclusion: Atypical meningiomas with osteolytic changes are rarely reported in the literature. Because of potential aggressive behavior, they need a regular follow-up with radiological imaging.

Keywords: Atypical, Intradiploic, Meningioma, Primary extradural meningioma

INTRODUCTION

Intradiploic meningiomas or intraosseous meningiomas are a subtype of primary extradural meningiomas (PEMs) that have been rarely reported in the literature, accounting for 1–2% of all meningiomas.[1] They are often mistaken for primary bony tumors. The majority of the intraosseous meningiomas are of osteoblastic subtypes. In general, they are slow-growing, histologically benign lesions; however, few cases of atypical and malignant histological subtypes with aggressive behavior have been reported in the literature. In this report, we report an unusual case of atypical intraosseous meningioma with osteolytic changes, without dural invasion, abutting the superior sagittal sinus that underwent gross total surgical excision and mesh cranioplasty in the same sitting.

CASE REPORT

A 40-year-old male patient presented with a history of large bony swelling over the right parietal region. He had a history of trauma to the head 12 years back, following which he started noticing

a gradually progressive, painless swelling over the right parietal eminence. On examination, the swelling was bony hard, immobile, and globular in shape with an approximate size of 8.0 × 9.0 cm [Figure 1a]. Imaging showed a large extradural, intradiploic lesion over the right high parietal region near the superior sagittal sinus crossing the midline. Differential diagnosis was giant cell tumor, osteogenic sarcoma, eosinophilic granuloma, aneurysmal bone cyst, and metastatic lesion. With a provisional diagnosis of an intradiploic meningioma, the patient was planned for surgical excision of the tumor. The patient was placed in prone position and U-shaped skin flap was raised across the midline (to adequately expose the superior sagittal sinus). The patient then underwent wide right-sided parietal craniectomy that included removal of all the diseased bone. There was bleeding from superior sagittal sinus, which was controlled by keeping around 1" broad gelfoam and gauze pieces over the sinus and accompanied by gentle pressure for 5–10 min. Bleeding was further reduced by elevating head end of the table. Intraoperatively, there was a larger extradural, intradiploic lesion that had eroded the inner and outer table of the skull with osteolytic margins. Dura was not involved and it was just pushed inside by the lesion. Gross total excision of tumor was done, while preserving superior sagittal sinus. Intraoperative impression was that of a meningioma. The bony defect was reconstructed with titanium mesh (mesh cranioplasty), as shown in [Figures 1a-e]. The postoperative period was uneventful. Histopathological examination showed proliferation of the meningothelial cells in the form of whorls, storiform patterns, and sheets with a prominence of nucleoli. Hyalinization of stroma was present without necrosis. Bone infiltration was seen at places by tumor cells with accompanying multinucleated giant cells and osteoclasts [Figure 2a]. All these features are suggestive of atypical meningioma WHO Grade II, infiltrating into the skull bone. The patient was discharged home after 5 days of hospital stay, since then he was on regular follow-up. Repeat imaging after 1 year showed no evidence of recurrence or residual disease.

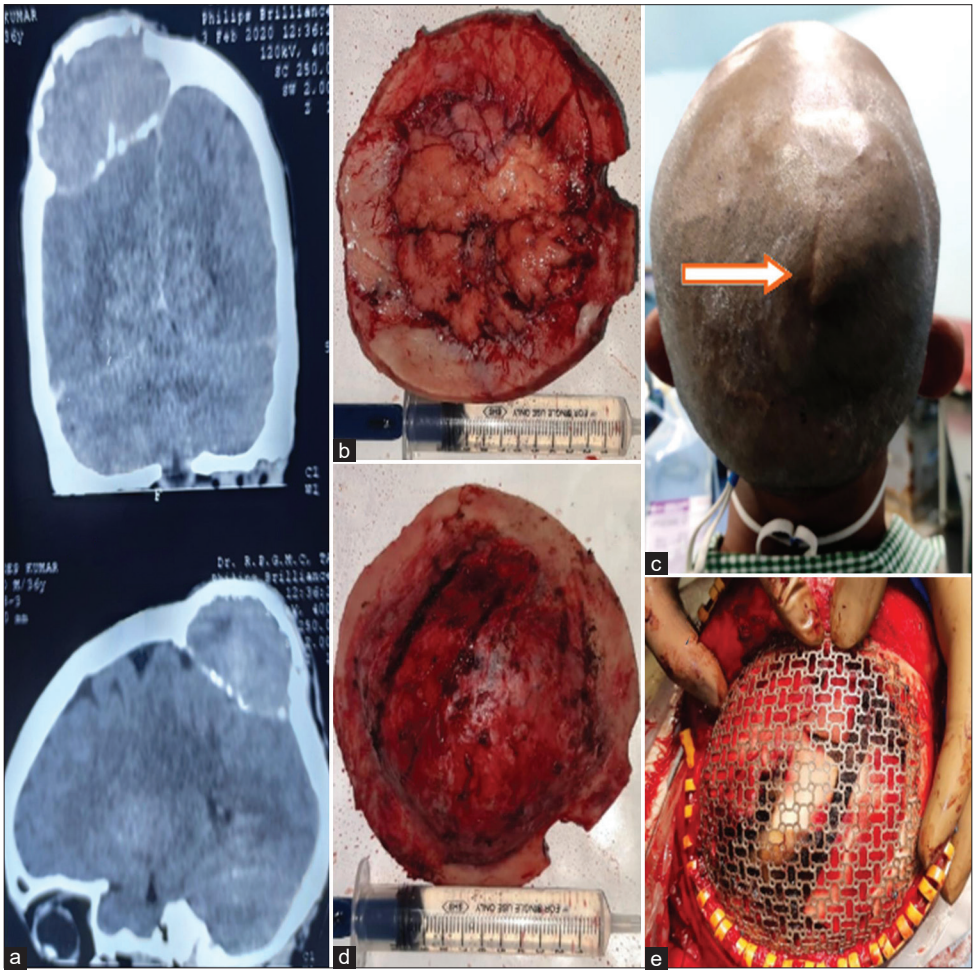

Figure 1: Noncontrast computerized tomography head showing lesion within the outer and inner table of the skull with osteolytic margins (a). Meningioma eroded both the tables (b and d). Scalp showing evidence of scar mark (Arrow) induced by trauma over the lesion (c). The defect created is reconstructed by mesh (e).

DISCUSSION

Meningiomas are the most common primary brain tumors.[14] Usually, meningiomas arise from dura (arachnoid cap cells)[17] but at times, meningioma can arise extracranially and is called as PEM. PEMs can be classified further into three types by Lang *et al.* purely extracalvarial (type I), purely calvarial (type II), or calvarial with extracalvarial extension (type III). According to the site of the location of the tumor, Lang *et al.* further subdivided type II and type III lesions into convexity (C) or skull base (B) forms.[10] Our case falls in type III (C) classification as per Lang's classification and as per literature, most of these tumors are IIIC [Table 1].[4],[5],[8],[12], [13],[15],[18]

Origin of intradiploic meningiomas

The hypothesis regarding their origin is still controversial. Entrapment of arachnoid cap cells or meningocytes along the cranial suture's lines during molding of the head at birth has been proposed; however, it was found that only 8.3% of the calvarial meningiomas were related to the cranial suture line.[10] Shuangshoti[16] postulated the role of multipotential mesenchymal cells in the development of these meningiomas that may explain the tumor location far from the common locations such as head and neck. Cushing and Eisenhardt[7] proposed that during head trauma, few meningocytes cells may get entrapped along the traumatic fracture lines and possibly develop into meningiomas later in the life. However, only around 0.2–4% of patients with PEMs are reported to have a history of head trauma in the past.[10] In our case, there was a history of head trauma present 12 years back and he developed intradiploic meningioma exactly on the same site a few years later [Figures 1c and 2b]. Although, the most of case reports/series have not shown a very strong association with trauma in the literature, it should be considered still one of the important factors in the development of intradiploic meningiomas.

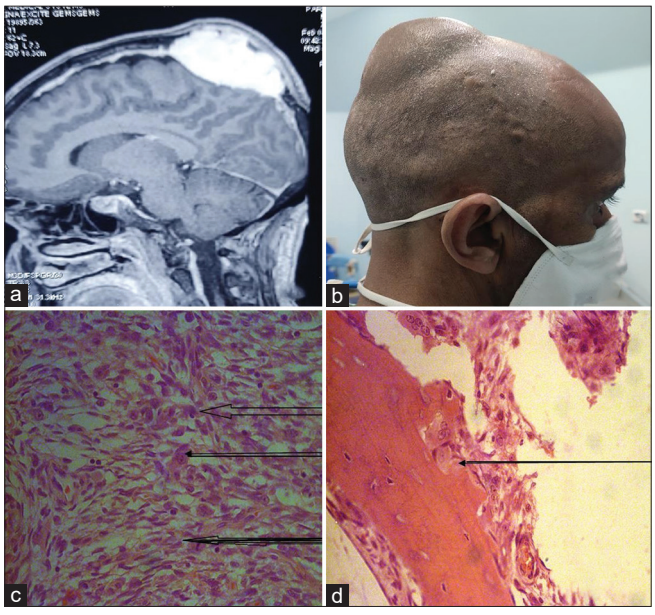

Figure 2: Magnetic resonance imaging brain showing intradiploic meningioma (a). Patient having external swelling of tumor (b). Histopathology showing whorl of meningothelial cells (upper large arrow), prominent nucleoli (middle arrow), atypical mitosis (lower dark arrow) suggestive of atypical meningioma (c). Infiltration of meningothelial cells into the bony tissue with H and E 200× (d).

Clinical presentations

They usually present as a hard, painless, and slow-growing swelling over the scalp. Mild headache and dizziness, along with neurological deficits, may also be present depending on the size, type, and location of the tumor.

On imaging

Although, these tumors delineate well on contrast magnetic resonance imaging (MRI), noncontrast computerized tomography head with bone window is often required to rule out sclerotic margins/osteolytic margins and further characterization of the lesion. These tumors are usually hypointense on T1-weighted MRI imaging and hyperintense on T2-weighted MRI imaging and show vivid and homogeneous enhancement after gadolinium administration [Figure 2a]. Intradiploic meningiomas can have hyperostosis and osteoblastic as well as osteolytic pictures on imaging.[11] It is noteworthy that osteolytic meningiomas associated with a soft-tissue component have more tendency of being atypical or malignant and must be considered malignant until proven otherwise.[2]

Differential diagnosis

Intradiploic meningiomas are generally misdiagnosed as bony tumors of skull and fibrous dysplasia's. Other differential diagnosis for osteolytic intraosseous meningiomas includes metastatic cancer, plasmacytoma, giant cell tumor, osteogenic sarcoma, eosinophilic granuloma, and aneurysmal bone cyst.

Histopathologically, the most of the intraosseous meningiomas are WHO Grade 1 lesion with psammomatous and meningothelial subtypes being the most frequently seen.[6,11] Atypical intraosseous meningiomas are very rarely, reported in the literature. To the best of our knowledge, only 11 such cases have been reported in the English literature till date [Table 1] including ours, which had whorls of meningothelial cells, prominent nucleoli, atypical mitosis and infiltration of meningothelial cells into bony tissue as well [Figure 2c and d].

Treatment of choice

The treatment modality of choice is *en bloc* wide resection. When there is suspicion of having an atypical/malignant

Table 1: Reports of primary intraosseous osteolytic atypical meningiomas.

S. No.	Author/year [References]	Age/ sex	Location	Type (Lang's classification)	Treatment	Dural involvement	Recurrence	Follow-up period (months)
1	Kwon *et al*., 2019[9]	80/M	Petrous	III B (Skull base)	GTR	No invasion	No	12
2	Parington *et al*., 1995[13]	84/F	Frontotemporal	IIIC	GTR with RT	Whole dura	Twice at 6 and 9 months	24
3	Bassiouni *et al*., 2006[3]	62/F	Frontal	IIC	GTR	Whole dura	No	NM
4	Kim *et al*., 2012[8]	68/M	Parietal	III C	GTR	Outer dural layer	No	12
5	Yun *et al*., 2014[18]	64/F	Frontal	III C	GTR	Whole dura	No	12
6	Bohara *et al*., 2016[4]	38/M	Parietal	III C	Wide resection with RT	Adherent to dura only	No	6
7	Lang *et al*., 2000[10]	59/M	sphenoid wing		GTR	Whole dura	No	24
8	Cheng *et al*., 2012[5]	68/F	Frontal	III C	GTR	Surgery with RT	Dural and brain invasion present	3
9	Sakakibara *et al*., 2018[15]	80/F	Parietal	III C	GTR	No dural invasion	No	8
10	Liu *et al*., 2015[11]	NM	NM	NM	NM	Wide resection only	NM	NM
11	Nakae *et al*., 2017[12]	30/M	Parietal	III C	GTR	Outer dural layer invasion	No	NM
12.	Present case, 2021	40/M	Parietal	III C	GTR	No dural invasion	None	12

GTR: Gross total resection, RT: Radiotherapy, NM: Not mentioned, M: Male, F: Female

component because of extracalvarial extension, one should do wide *en bloc* resection of the lesion with 1 cm resection of normal bone margin,[18] if possible. Literature wise, most of such tumors underwent GTR with or without radiotherapy.[3],[4],[5],[8],[9],[10],[12],[13],[18] If the surgical resection is wide, cranial reconstruction must be done. In case of incomplete resection, the residual tumor should be considered for radiation therapy, regardless of pathological grade.[18] However, for atypical as well as malignant PEMs, despite surgically complete resection, there is still controversy existing regarding considering radiotherapy in the head.[10,18] Whereas, chemotherapy should be reserved for those who are unresectable, growing WHO I and all WHO II and III PEMs.[18] No single chemotherapeutic agent can be recommended for these meningioma. Few cytotoxic chemotherapeutics agents have demonstrated some efficacy including hydroxyurea, irinotecan, and temozolomide.[2]

CONCLUSION

Atypical intradiploic meningiomas with osteolytic changes are rare entities among all meningiomas. One should go for a wide *en bloc* resection with at least 1 cm apparently healthy bony margin and should strongly consider adjuvant radiotherapy/chemotherapy for unresectable, partially resectable, residual, or recurrent lesions.

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Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Agrawal V, Ludwig N, Agrawal A, Bulsara KR. Intraosseous intracranial meningioma. Am J Neuroradiol 2007;28:314-5.
2. Austin M, Betsy H, James O, David R, McLendon R, Cory A.

Treatment recommendations for primary extradural meningiomas. Cancer 2011;117:24-38.
3. Bassiouni H, Asgari S, Hubschen U, Konig HJ. Dural involvement in primary extradural meningiomas of the cranial vault. J Neurosurg 2006;105:51-9.
4. Bohara S, Agarwal S, Khurana N, Pandey PN. Primary intraosseous atypical inflammatory meningioma presenting as a lytic skull lesion: Case report with review of literature. Indian J Pathol Microbiol 2016;59:386-8.
5. Cheng HT, Cheng CH, Chio CC, Chang ST. Atypical intraossoeous osteolytic meningioma mimicking calvarial metastasis. Form J Surg 2012;45:69-72.
6. Cirak B, Guven MB, Ugras S, Kutluhan A, Unal O. Fronto-orbitonasal intradiploic meningioma in a child. Pediatr Neurosurg 2000;32:48-51.
7. Cushing H, Eisenhardt L. Meningiomas: Their Classification, Regional Behavior, Life History and Surgical End Results. Springfield, Ill: Charles C Thomas; 1938.
8. Kim H, Jung TY, Kim IY, Lee JK. Two cases of primary osteolytic intraosseous meningioma of the skull metastasizing to whole skull and the spine. J Korean Neurosurg Soc 2012;51:151-4.
9. Kwon SM, Ko Y, Bang SS. Primary intraosseous osteolytic meningioma: A case report and review of the literature. BMC Neurol 2019;19:176.
10. Lang FF, Macdonald OK, Fuller GN, DeMonte F. Primary extradural meningiomas: A report on nine cases and review of the literature from the era of computerized tomography scanning. J Neurosurg 2000;93:940-50.
11. Liu Y, Wang H, Shao H, Wang C. Primary extradural meningiomas in head: A report of 19 cases and review of literature. Int J Clin Exp Pathol 2015;8:5624-32.
12. Nakae S, Uchida H, Takayanagi T, Hasegawa M. Atypical intraosseous meningioma with growth hormone deficiency and hyperparathyroidism after craniospinal irradiation. Childs Nerv Syst 2017;33:2077-8.
13. Partington MD, Scheithauer BW, Piepgras DG. Carcinoembryonic antigen production associated with an osteolytic meningioma. J Neurosurg 1995;82:489-92.
14. Rogers L, Barani I, Chamberlain M, Kaley TJ, McDermott M, Raizer J, *et al*. Meningiomas: Knowledge base, treatment outcomes, and uncertainties. A RANO review. J Neurosurg 2015;122:4-23.
15. Sakakibara Y, Taguchi Y, Nakamura H, Onodera H, Wakui D. A case of primary extradural meningioma located at the high convexity: A case report and review of the literature. No Shinkei Geka 2018;46:523-8.
16. Shuangshotis S. Primary Meningioma Outside the Central Nervous System: Meningioma. New York: Raven Press; 1991. p. 107-28.
17. Thompson LD, Bouffard JP, Sandberg GD, Mena H. Primary ear and temporal bone meningiomas: A clinicopathologic study of 36 cases with a review of the literature. Mod Pathol 2003;16:236-45.
18. Yun JH, Lee SK. Primary osteolytic intraosseous atypical meningioma with soft tissue and dural invasion: Report of a case and review of literatures. J Korean Neurosurg Soc 2014;56:509-12.

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