Case Report

Skull Vault Plasmacytoma Mimicking Parasagittal Meningioma: "Mini-Brain Appearance"

Abstract

Plasmacytomas occur as lesions in soft tissue or bone. Skull vault plasmacytomas are rare lesions comprising 0.7% of all plasmacytomas. Workup for myeloma must be done in such cases to rule out multiple myeloma. Here, we report a case of a 63-year-old female who presented to us with a large skull vault swelling which appeared to mimic a parasagittal meningioma on imaging. Histopathological imaging revealed it to be a plasmacytoma. A retrospective review of the radiology revealed the characteristic "mini-brain appearance" in our case. Literature on the subject is also reviewed.

Keywords: *Mini brain, plasmacytoma, skull, vault*

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Introduction

Plasma cell neoplasms can be classified as multiple myeloma, solitary plasmacytomas of bone, and extramedullary plasmacytomas.[1] Plasmacytomas can occur in soft-tissue lesions in soft tissue or bone.[1] They are most commonly found in the axial skeleton (thoracic and lumbar spine).[1] Skull vault plasmacytomas are rare lesions comprising 0.7% of all plasmacytomas.[2,3] They can mimic meningiomas, osteogenic sarcomas, or metastatic carcinomas on radiologic imaging.

Case Report

A 63-year-old-female presented to us with a swelling over the vertex of the skull for the past 1 year which was insidious in onset, painless, of the size of a lemon when it was first noticed and gradually progressed to achieve the present size of about a cricket ball. The swelling was nontender on palpation and the overlying skin was normal. On examination, she was conscious oriented and had no neurological deficits. The progressively increasing scalp swelling made her seek medical attention. Noncontrast computed tomography (CT) head done showed a large extra-axial swelling of size 10.3 cm × 8 cm × 7 cm over midline (left > right) frontoparietal parasagittal region causing compression of underlying brain and erosion of skull [Figure 1]. Magnetic resonance imaging (MRI) brain [Figure 2] revealed a large heterogeneously enhancing lesion over midline (left > right) frontoparietal parasagittal region. On magnetic resonance venogram [Figure 3], the middle part of the superior sagittal sinus was not visualized. Rest of the veins was normal. A provisional diagnosis of parasagittal meningioma was made. On digital subtraction angiography, tumor was fed by feeders from a bilateral superficial temporal artery, middle meningeal artery, and occipital artery. During embolization, dangerous anastomosis between anterior trunk of the middle meningeal artery and ophthalmic was carefully ruled out before all feeders were superselectively embolized using polyvinyl alcohol particles to achieve tumor devascularization. After embolization, the patient was planned for surgery after 5 days. The patient's head was fixed in three pins and positioned in a semi-sitting position. There was a large, gray white, soft-to-firm, moderately vascular, extradural tumor destroying the calvarium. No dural or sinus invasion was seen. The intraoperative impression was that of a tumor of bony origin. Histopathology [Figure 4] revealed a tumor with sheets of plasmacytoid cells with large areas of necrosis and microabscesses. The cells were round to oval with moderate amounts of cytoplasm and eccentric nuclei. On immunohistochemistry, most cells were positive for CD138 and lambda light chain suggestive of a plasmacytoma. Serum light

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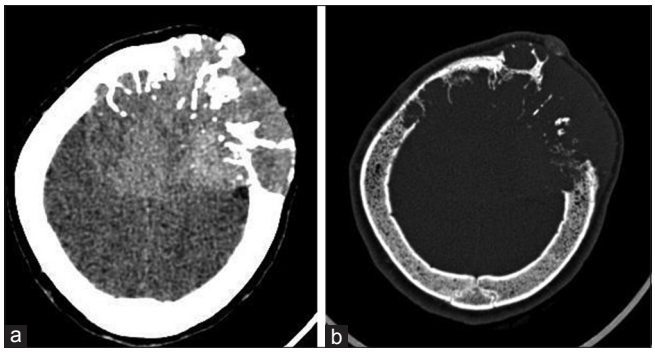

Figure 1: (a) Noncontrast computed tomography head showing large hyperdense extra-axial lesion over midline frontoparietal region. (b) Noncontrast computed tomography head (bone window) showing erosion of the skull

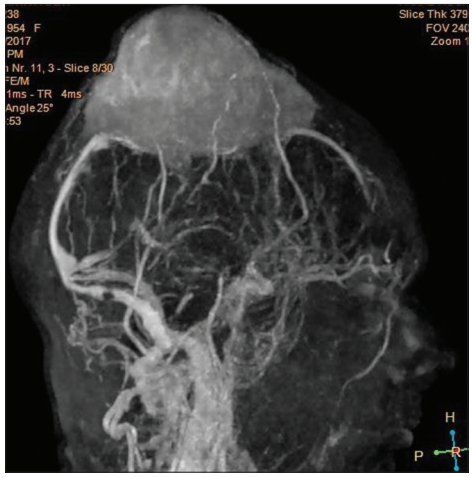

Figure 3: On magnetic resonance venogram, middle part of superior sagittal sinus was not visualized

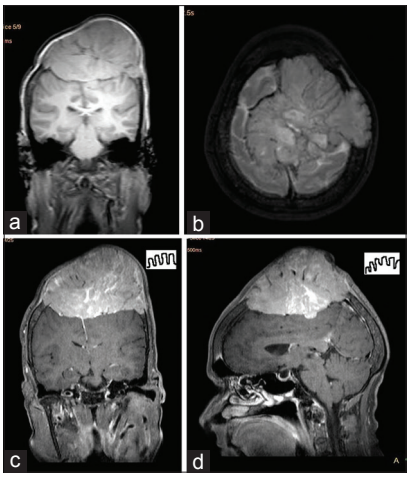

Figure 2: (a) Magnetic resonance imaging showing an isointense tumor on T1-weighted sequence. (b) Magnetic resonance imaging axial view showing a hyperintense tumor on T2-weighted sequence. (c) Contrast-enhanced magnetic resonance imaging (coronal) showing heterogeneously enhancing tumor (with appearance suggestive of sulci and gyri: mini-brain appearance; see inset for illustration). (d) Contrast-enhanced magnetic resonance imaging (sagittal) showing heterogeneously enhancing tumor (with appearance suggestive of sulci and gyri: mini-brain appearance; see inset for illustration)

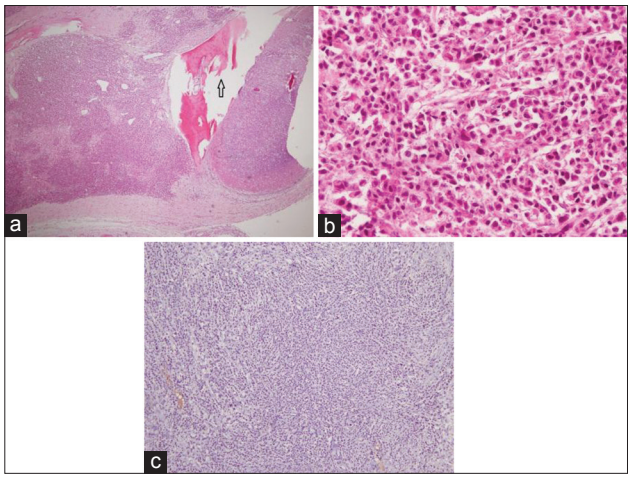

Figure 4: (a) Low-power microscopy (H and E, ×20) showing a cellular tumor lying in sheets. Tumor is seen breaching the bone (arrow). (b) High-power microscopy (H and E, ×40) showing plasmacytoid cells with atypical and binucleate forms. (c) Immunohistochemistry showing diffuse positivity for lambda light chain and absence of kappa chain (light chain restriction)

chain assay showed grossly elevated lambda light chain levels. On serum electrophoresis, M spike was not seen. Serum calcium levels and renal function were normal. Positron emission tomography (PET) scan revealed uptake in C3, C4, and C5 vertebra and left iliac bone establishing a final diagnosis of light chain myeloma. The patient was then referred to hematology-oncology department for bone marrow evaluation and adjuvant chemotherapy.

Discussion

Myeloma is a hematological malignancy that originates from plasma cells of bone marrow.[1,2] Plasma cell neoplasms can be classified into multiple myeloma, solitary plasmacytomas of bone, and extramedullary plasmacytomas.[1,2] While multiple myeloma is a systemic disease, solitary plasmacytoma and extramedullary plasmacytomas are focal forms of plasma cell neoplasms. Plasmacytomas occur as mass lesions in soft tissue or bone. Skull vault plasmacytomas are very rare, comprising 0.7% of all plasmacytomas, most common locations being thoracic and lumbar spine.[2,3] As per our information, this is the first case report of a plasmacytoma in a parasagittal location which closely mimicked the appearance of a parasagittal meningioma.

In such cases, a differential diagnosis of meningioma, osteosarcoma, metastatic carcinoma, and plasmacytoma must be considered. We considered meningioma as the most likely possibility because of its common occurrence in the parasagittal location. For meningioma, it must be remembered that they rarely cause osteolysis albeit for intraosseous forms.[4] Osteosarcomas occur mostly in long bones and rarely involve the skull when they cause the destruction of outer table and present as extracranial masses.[2,4] Osteolytic metastatic carcinoma is usually multiple and has punched out margins.[4]

Table 1: Reported cases of skull vault plasmacytoma with "mini-brain appearance"

Serial number	Author	Year	Age/sex	Location	Lesion size (mm)
1	Nagatomo *et al*.[5]	1994	56/male	Temporo-parietal	60 × 40
2	Matsuda *et al*.[6]	1996	55/female	Fronto-temporal	70 × 50
3	Okamoto *et al*.[7]	1997	72/female	Occipital	NA
4	Okamoto *et al*.[7]	1997	64/male	Occipital	60 × 90
5	Tanaka *et al*.[3]	1998	55/male	Frontal	80 × 80
6	Zigouris *et al*.[8]	2009	78/female	Temporo-parietal	98 × 80
7	Bakar and Tekkok[4]	2010	49/male	Frontal	90 × 85
8	Simoni *et al*.[9]	2013	48/male	Frontal	60 × 35
9	Present case	2017	63/female	Fronto-parietal	103 × 80

NA - Not available

Plasmacytomas may be identified as osteolytic lesions on X-ray.[2,4,5] On a CT scan, they are seen as osteolytic lesions without sclerotic rim and homogeneous contrast enhancement. They show variable intensity on T1-weighted images but enhance strongly on administration of gadolinium.[4,5] The sharp borders of the lesion, lack of sclerosis, and minimal periosteal reaction are strongly suggestive of a plasmacytoma. In our case, lesion was hyperintense, osteolytic without sclerosis on the CT scan. It was hyperintense on T1 and was heterogeneously contrast enhancing on MRI. On retrospective review, lesion was seen to have "mini-brain appearance." "Mini-brain appearance" was first described by Major *et al*. in their series of spinal plasmacytomas.[1] As in our case, "mini-brain appearance" can be retrospectively seen in cases of calvarial plasmacytomas reported in the literature [Table 1]. We feel that finding this characteristic appearance can aid in forming a radiological diagnosis of a calvarial plasmacytomas even in locations where this lesion is extremely rare.

For embolization of tumors fed by the middle meningeal artery, overt and covert "dangerous anastomosis between anterior trunk of the middle meningeal artery and ophthalmic artery" must be ruled out during angiography and superselective distal embolization of tumor feeders must be done to avoid catastrophic complications.

Chemotherapy is the treatment of choice for multiple myeloma.[2] Skull plasmacytomas are treated by excision followed by radiotherapy.[6,7] After a diagnosis of skull plasmacytoma, patients must be evaluated for myeloma using a battery of tests such as serum light chain assay, serum electrophoresis, urine for Bence–Jones protein, PET scan, and bone marrow examination.[9] The monoclonal nature of tumor cells must also be confirmed to rule out the possibility of plasma cell granuloma. The prognosis of solitary plasmacytomas and extramedullary plasmacytomas is better as compared to that of multiple myeloma.[7,8] These patients must be followed up closely to detect progression to multiple myeloma.

Conclusion

Skull vault plasmacytomas are rare lesions. Their appearance can often mimic other skull vault lesions such as meningioma, osteosarcoma, and metastatic carcinoma. Findings of "mini-brain appearance" can help in the formation of diagnosis in such cases.

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.

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Conflicts of interest

There are no conflicts of interest.

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