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Case Report

Solitary plasmacytoma of the calvarium: A successful management of a long-standing large lesion with a long-term follow-up

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ABSTRACT

Background: Solitary plasmacytoma of the calvarium (SPC), without evidence of multiple myeloma (MM), is extremely rare. We report a case of a long-standing large SPC that was treated successfully by surgical excision and adjuvant radiotherapy with a long follow-up period.

Case Description: A 58-year-old male patient presented with a 5-year history of painless skull swelling. On the physical examination, the mass was 6 × 6 cm in size, oval, not tender, and firm in consistency with normal skin color. A brain computed tomography scan showed a destructive skull lesion. A brain magnetic reasoning imaging (MRI) showed a large expansile lytic mass lesion arising from the skull vault in the frontal-parietal region with multiple internal septa. The patient underwent a craniectomy and excision of the lesion, followed by cranioplasty using methyl methacrylate. The final diagnosis was consistent with plasmacytoma based on the histopathological features. One month follow-up brain MRI showed no evidence of residual tumor. The skeletal survey and bone marrow biopsy did not reveal any evidence of MM. The patient was referred to medical oncology for further treatment and received radiation therapy. During nine years of clinical and radiological follow-up, there was no evidence of any metastasis or recurrence.

Conclusion: SPC is a rare disease with high rates of misdiagnosis. Careful evaluations are crucial to exclude systemic involvement. Surgical resection followed by radiotherapy may be adequate for the disease control. Close follow-up with regular lifelong examinations is important to avoid a generalized incurable disease.

Keywords: Calvarium, Case report, Multiple myeloma, Solitary plasmacytoma

INTRODUCTION

Osteolytic skull lesions are commonly observed in routine clinical work. One of these is plasma cell neoplasm, which arises from the proliferation of a single clone of B-cell lymphocytes. It produces a wide spectrum of disorders, ranging from solitary plasmacytoma (SP) to extremely malignant multiple myeloma (MM).[4,6] The appearance of an osteolytic SP of the calvarium (SPC) without signs of systemic myelomatosis is considered rare.[3] The clinical features are complex and not easily identified, leading to a high misdiagnosis rate. A comprehensive examination and analysis are needed for the correct diagnosis.[1,6] The prognosis for SPC appears to be good when

it is diagnosed on strict criteria.[1,9] We report a case of a long-standing large SPC that was treated successfully by surgical excision and adjuvant radiotherapy with a long follow-up period.

CASE DESCRIPTION

A 58-year-old male patient, who is not known to have any medical or surgical histories, presented in November 2014 to our hospital complaining of a painless skull swelling. He first noticed the skull swelling five years ago, and since then, he reported that it did not increase in size until he sustained a head trauma several months before the presentation. He claimed that the reason he had not sought medical attention for the past few years was because he was not bothered by the lesion. However, after the head trauma he sustained, the lesion got bigger, and he became worried and came to the hospital. He did not report having any neurological deficits. There was no history of weight loss, fever, or night sweating. He did not have a family history of malignancy. On the physical examination, the mass was 6 × 6 cm in size, oval, not tender, and firm in consistency with normal skin color. Other neurological examinations revealed intact functions without any subtle motor deficit. A brain computed tomography (CT) scan showed a destructive skull lesion that is causing spike-like projections that are invading the surrounding cortical bone [Figure 1]. A brain magnetic reasoning imaging (MRI) showed a large expansile lytic mass lesion arising from the skull vault in the frontal-parietal region with multiple internal septa, and a brain magnetic reasoning venography showed an occluded anterior segment of the superior sagittal sinus secondary to lesion invasion [Figure 2]. Differential diagnoses of a possible primary bone tumor or a secondary bone metastasis from another origin were raised by the team. As these findings confirmed the invasive nature of the lesion, the team decided to excise the lesion, and the patient was prepared for surgery. Under general anesthesia, the patient was positioned supine with the head fixed using Mayfield head pins. A bi-coronal skin incision was done, and then a wide craniectomy revealed the lesion. The lesion, which seemed to be originating from the intraosseous bone, was red-grayish in color, rubbery, soft, and suctionable. It was invading the cortical bone and underlying dura with adhesions, causing active bleeding in its deep parts. An excision of the lesion and the underlying dura with the aid of the navigation system was done following the ligation of the anterior one-third of the superior sagittal sinus. To ensure a complete excision, the margin of resection of bone and dura was increased beyond the lesion margins. There were no signs of invasion of the underlying neural tissue or the overlying subcutaneous tissue.

Following the lesion excision, a cranioplasty using methyl methacrylate was utilized [Figure 3]. Postoperatively, he was doing fine, and his follow-up CT brain was satisfactory, showing no hemorrhage, infarction, or residual mass. Hematoxylin and eosin stain sections of the excised lesion showed sheets of plasma cells. Immunohistochemistry showed that the cells were positive for plasma cell antigen, CD138, and Kappa; however, they were negative for Lambda and CD1a. The final diagnosis was consistent with plasmacytoma [Figure 4]. One month follow-up brain MRI showed no evidence of residual tumor [Figure 5]. Other investigations and bone marrow biopsies did not reveal evidence of MM. The skeletal survey was negative for any suspicious lytic or sclerotic lesions. The patient was referred to medical oncology for further treatment and received radiation therapy. During nine years of follow-up, brain MRIs and skeletal

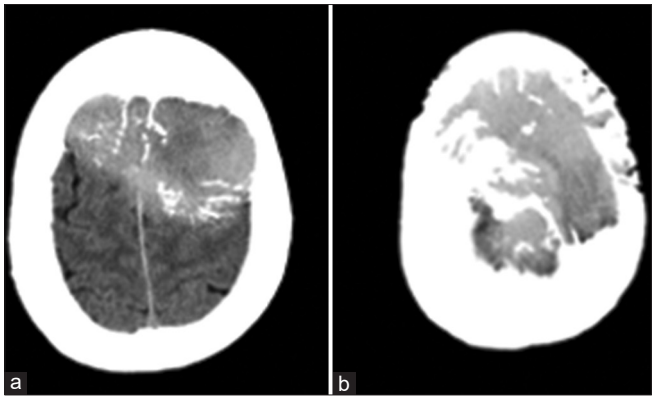

Figure 1: (a and b) Preoperative computed tomography scan.

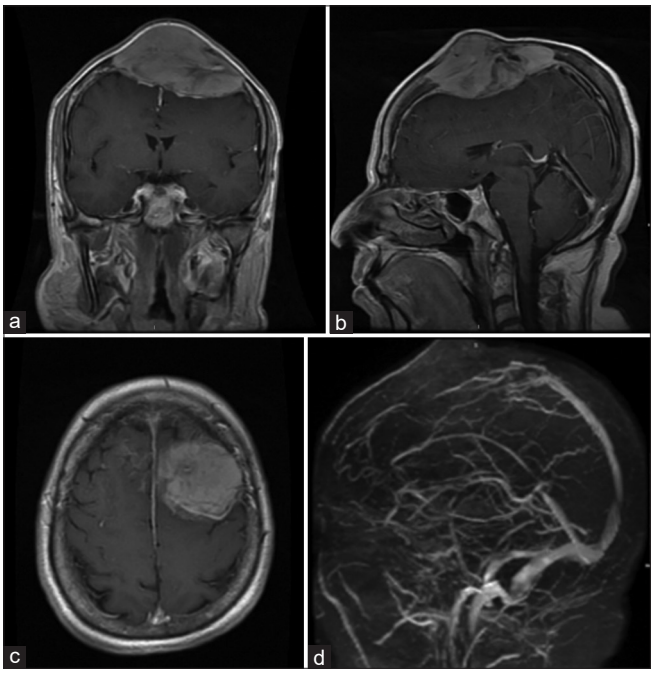

Figure 2: (a-c) Preoperative T1-weighted Magnetic Reasoning Imaging with contrast and (d) Preparative Magnetic Reasoning Venography.

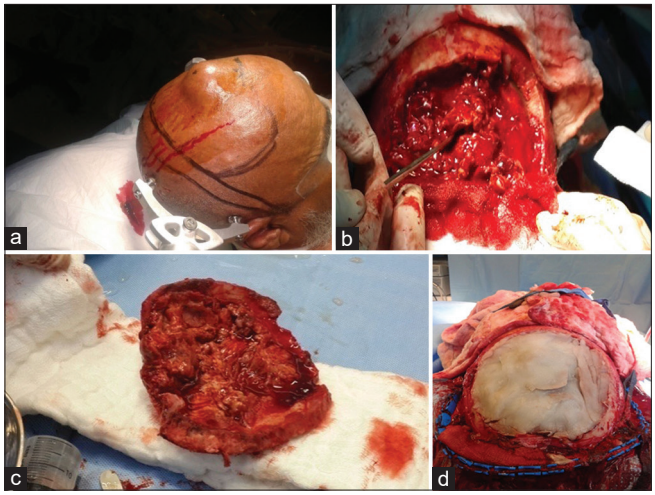

Figure 3: (a) Intraoperative figures demonstrating the positioning, (b and c) Surgical resection, and (d) Cranioplasty.

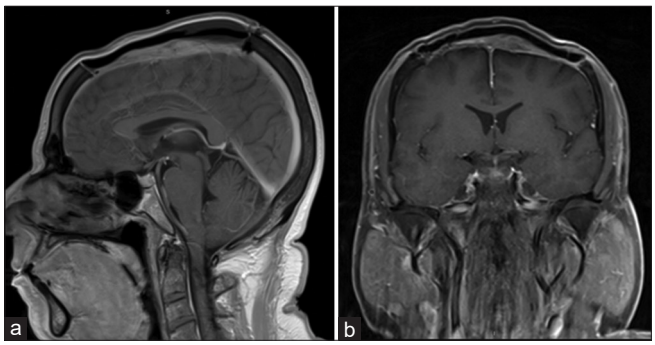

Figure 4: (a and b) 1-month postoperative T1-weighted magnetic reasoning imaging with contrast.

surveys showed no evidence of any metastasis or recurrence. The patient remained healthy without any new complaints.

DISCUSSION

SP is a rare malignancy and is characterized by malignant proliferation of monoclonal plasma cells.[4] SP has a <0.5 incident rate of all malignant plasma cell tumors.[3] It mostly occurs in the axial skeleton, such as the skull and vertebra.[1] It shows male preponderance and is extremely rare in patients <30 years of age.[3] The most common sites of SPC are the parietal bone and the skull base bones.[3,9] Solitary craniocerebral plasmacytoma is the least common form of extramedullary SPC, with an estimated prevalence of 0.7% within this category.[2] The dura is the most common site of craniocerebral plasmacytoma that does not originate from the skull, occasionally secondarily involving the calvaria and brain parenchyma. Rarely, SP can arise within brain tissue. They are typically smooth and lobulated but can cause irregular destruction of the involved bone, as seen in our case, with no infiltration of

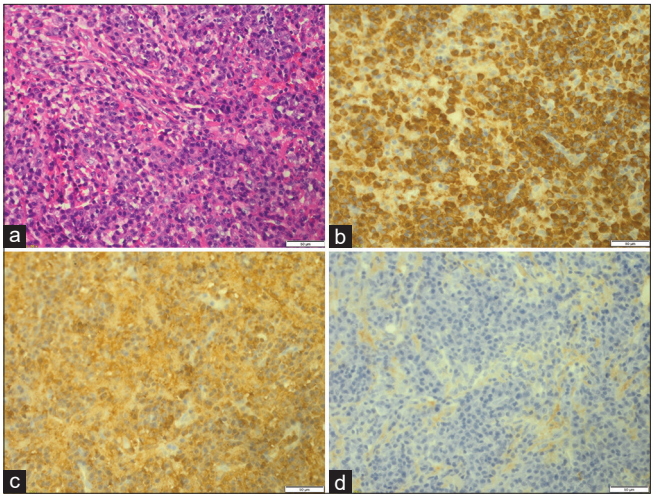

Figure 5: Pathology slides including (a) H&E, (b) PLASMA-N, (c) Kappa, (d) and Lambda.

underlying brain parenchyma.[2,5] Plasmacytoma lesions may cause cranial nerve, orbital involvement, or increased intracranial pressure symptoms. Lesions at the skull base may involve the cavernous sinus, petrous temporal bone, or sphenoid bone and, thus, are often accompanied by cranial neuropathies; abducens palsy with attendant diplopia is the most common. In addition, cranial neuropathies may be caused by direct infiltration of the nerves with myelomatous cells. Involvement of the orbit may result in exophthalmos or ophthalmoparesis. Intracranial extension of plasmacytoma may cause headaches or other symptoms of increased intracranial pressure. An intradural extension may give rise to seizures.[2,9] Plasmablastic plasmacytomas may be associated with systemic MM, and those who are not known to have MM may progress later to have MM. A thorough evaluation to exclude MM is necessary, particularly if the lesion is found at the skull base and includes bone marrow evaluation, a skeletal survey, a bone scan, and serum and urine protein electrophoresis.[8,9] Our patient's investigations were negative for MM. CT findings of plasmacytoma are characterized by well-defined lytic lesions involving the diploic and cortical bone, without a sclerotic border, typically compressing adjacent brain, and appear on CT as isodense to hyperdense. Diffuse, homogeneous enhancement after contrast administration is generally seen. Intratumoral calcifications due to bone destruction are often present, simulating calcifications seen in meningiomas.[2,7] MRI is helpful for determining the presence of any associated extraosseous component, as well as for determining the presence of flow voids because these lesions can be quite vascular. Lesions typically appear isointense to hyperintense on T1-weighted MRI and markedly hypointense on T2-weighted MRI sequences. Plasmacytomas typically demonstrate restriction on diffusion-weighted images, whereas MM will show increased diffusion.[2,8] Plasmacytoma is a highly

radiosensitive tumor. The optimal treatment of SPC is surgical resection followed by radiotherapy, although some have reported good results with surgical resection alone. At surgery, these tumors are often found to be quite vascular, and significant blood loss should be anticipated.[1-3,7,9]

Limitations

This study has some limitations as it only included one case report of SPC. However, considering how rare this condition is, we aim to contribute to the literature to help understand these lesions better. Further studies with more cases of SPC to help understand the clinical features and predict the prognosis are recommended.

CONCLUSION

SPC is a rare disease. Clinical and imaging evaluations are of paramount importance to exclude systemic involvement. Careful surgical resection, if total, may be adequate for disease control. Nevertheless, close follow-up with regular lifelong examinations is important to avoid a generalized incurable disease.

Ethical approval

The Institutional Review Board approval is not required.

Declaration of patient consent

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

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Nil.

Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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