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Case Report

Solitary bone plasmacytoma as posterior fossa cranial neoplasia, presentation of two clinical cases

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ABSTRACT

Background: Solitary bone plasmacytoma is a plasmatic cell dyscrasia; its presentation in the posterior fossa is very rare.

Case Description: We present two cases, a 59-year-old male and a 50-year-old female, both with heterogeneous clinical presentation. One had symptoms compatible with endocranial hypertension, and the other presented with a hemispheric cerebellar syndrome and ipsilateral trigeminal neuralgia. They were both related to an intraosseous tumor of the occipital region near the torcula with large extension to the posterior fossa. The diagnosis of a plasma cell neoplasm arising from the diploe of the squamous portion of the occipital bone was confirmed with immunohistochemistry.

Conclusion: The treatment for a cranial tumor that is suspected to be a solitary bone plasmacytoma requires a multidisciplinary team to diagnose, plan a total resection, and after surgery continue with the follow-up of the patient. Solitary bone plasmacytoma should be considered as a differential diagnosis for a tumor that produces cancellous bone widening without sclerotic borders.

Keywords: Intraosseous cranial tumor, Plasmatic cell tumor, Posterior cranial fossa, Solitary bone plasmacytoma

INTRODUCTION

Plasmacytoma is a rare plasma cell neoplasm that represents <10% of monoclonal gammopathies.[4] There are two known clinical presentations: (a) solitary bone plasmacytoma (SBP), which affects of frequency: spine, ribs, skull, and pelvis and (b) soft tissue ("extraosseous" or "extramedullary") (EMP), mostly found in the upper respiratory tract. To diagnose plasma cell dyscrasias, the International Myeloma Working Group[9] criteria are used; these are summarized in [Table 1].

In the skull, the most affected regions are skull base, sphenoclival, petrous apex, nasopharynx, and orbital roof. It is less frequent in the calvaria (0.7% of the total)[2] and exceptionally rare in the posterior skull base (five reported cases in revised literature).[3] We describe here two cases of SBP located in the bone structures of the posterior fossa.

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Table 1: Diagnostic criteria of the International Myeloma Working Group (adapted from reference[4,9]).

Solitary bone plasmacytoma	Extramedullary plasmacytoma	Plasma cell myeloma
Absence of M protein in serum and/or urine	Absence of M protein in serum and/or urine	Presence of M protein in serum and/or urine
Localized destruction area due to clonal plasma cells	Extramedullary tumor made up of clonal plasma cells.	Clonal plasma cells from bone marrow or plasmacytoma
Bone marrow studies are incompatible with multiple myeloma	Normal bone marrow	Related organ or tissue deterioration (end-organ damage, including bone lesions)
Normal bone imaging studies (in addition to spine and pelvis MRI)	Normal bone imaging studies	
No related organ or tissue deterioration (no end-organ damage other than the solitary bone lesion)	No related organ or tissue deterioration (end-organ damage, including bone lesions)	

CASE PRESENTATION

Case 1

Case 1: Fifty-nine-year-old male, without relevant prior medical history, began in April 2018 with a frontal oppressive headache of moderate intensity; he was initially diagnosed with hypertension and received treatment with calcium channel blockers. He was initially diagnosed with hypertension and received treatment with calcium channel blockers. In August 2018, he started noticing unsteady gait, disorientation, and intermittent urinary incontinence. He sought medical attention at his regional hospital in December 2018, where an unenhanced CT brain scan was diagnostic for obstructive hydrocephalus secondary to a posterior fossa tumor. A right precoronal ventriculoperitoneal high-pressure shunt was placed, apparently without complications. He was sent for consultation to our unit in February 2019. At his initial physical examination, a right hemispheric cerebellar syndrome was documented. In the initial MRI, a posterior fossa tumor was observed, of approximately 61 × 60 × 52 mm, with irregular borders, isointense in T1- and T2-weighted images, with homogenous enhancement on contrast-enhanced images; with compression and displacement of the fourth ventricle and the cerebellar left hemisphere obliteration of the right transverse sinus was also documented [Figure 1]. All laboratory results were within normal parameters. A tentorial meningioma was considered as the presumptive diagnosis.

In June 2019, surgery was performed. A combined supra- and infra-tentorial approach with a bilateral parieto-occipital craniectomy, ligation of the right transverse sinus, total resection of the tumor, duraplasty, and cranioplasty with polymethylmethacrylate was performed. A solid, extra-axial tumor of a rubbery consistency, with invasion to parietal bone, occipital squama in its three layers, right transverse sinus, torcular Herophili, tentorium, and adjacent dura, was found.

At the pathology laboratory, bone fragments with an approximate diameter of 8 cm were processed. A tumor which expanded the diploe up to several centimeters, with an external smooth surface and an internal fractious surface was observed. When cut, a separation created by the tumor between the internal and the external periosteum was made evident [Figure 2]. Several grayish-brown soft-tissue samples with hemorrhagic foci, approximately 6.5 × 5.5 × 2.7 cm, were also examined. The histopathological analysis showed sheets of plasma cells with variation in their nuclei size and shape (mature plasma cells, plasmablasts, and multinucleated plasma cells) which infiltrated the cortical and trabecular bone [Figure 3]. With immunohistochemistry (IHC), a plasma cell differentiation was proven: CD138-positive diffuse expression 3+/3+; CD79-positive diffuse expression 3+/3+; CD19-positive focal expression 3+/3+ (5%); CD56, CD117, and cyclin D1 negative; kappa-positive focal expression 3+/3+ (5%); and lambda-negative and high proliferative rate evaluated with Ki-67 (10%).

The patient showed a satisfactory postoperative evolution and was discharged a week after. Afterward, treatment was continued with 28 fractionated craniospinal radiotherapy sessions in December 2019. At his 6-month follow-up consultation, he was found to be disease free [Figure 4].

Case 2

A 50-year-old female with prior medical history of smoking and controlled hypertension, presented to clinic with an 18-month history of a moderate intensity, occipital, and oppressive headache, it partially improved with NSAIDs. The headache was accompanied with gait lateropulsion, right peripheral facial palsy, and right hemifacial electric shock-like pain, as well as paresthesia. On initial head MRI, there was a posterior fossa tumor, which extended from the right petroclival fissure to the occipital squama, of approximately 88 × 51 × 40 mm, with irregular borders, isointense in T1-weighted images and hypointense in T2-weighted images,

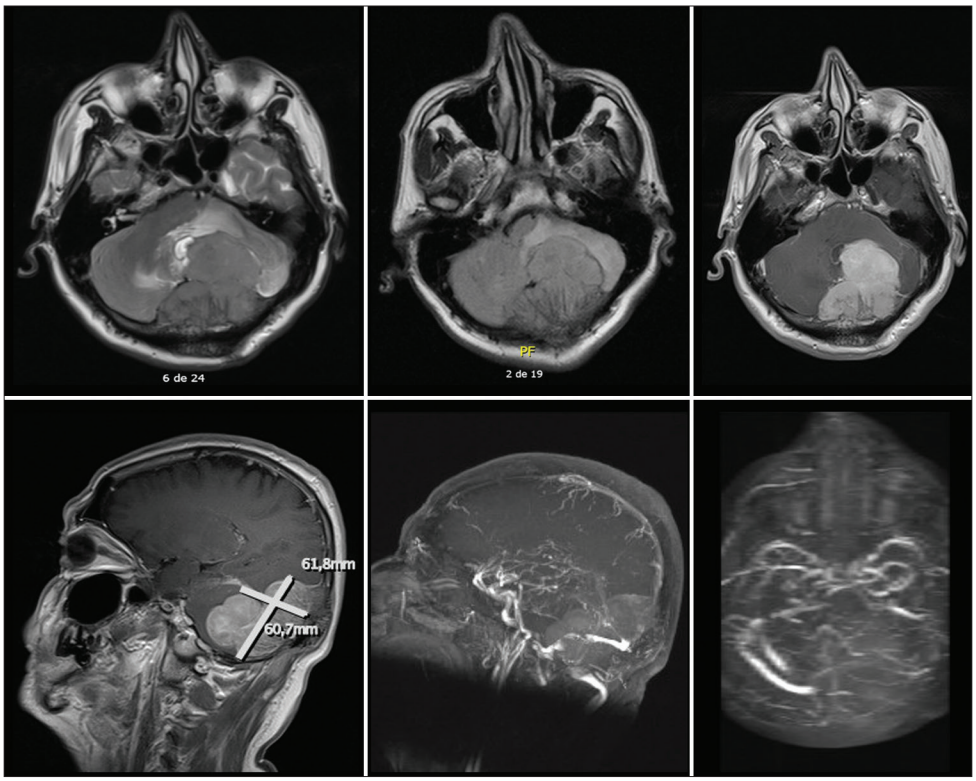

Figure 1: T1-weighted contrast-enhanced brain MRI, where skull invasion to the posterior fossa, the squamous portion of the occipital bone and both parietal bones can be appreciated. The fourth ventricle is displaced to the right and partially obliterated; there is also significant compression of the cerebellar left hemisphere, as well as obliteration of the right transverse sinus on the magnetic resonance angiography.

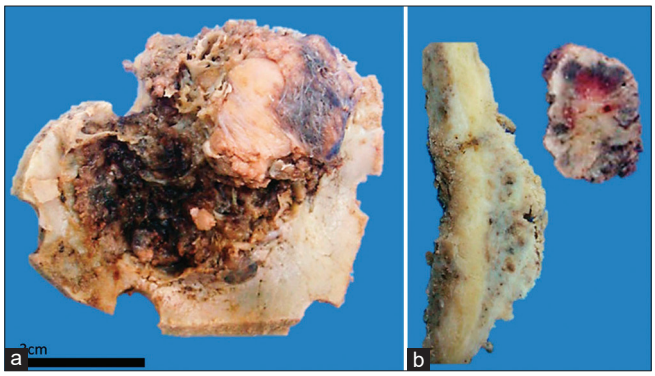

Figure 2: Macroscopic image of Case 1 where osseous involvement can be appreciated. (a) Internal side of a fragment of occipital bone. A lesion with irregular borders and recent hemorrhage can be observed. It is infiltrative and fixed to the inner table. (b) The sample has been cut perpendicular to the lesion. One can recognize the infiltration and deformation to the external table, there is also a fragment of tumor which is not fixed to the bone with hemorrhaging areas.

with homogenous enhancement on contrast-enhanced images, it caused descending compression of the cerebellar right hemisphere and encased the exiting roots of cranial nerves V, VII-VIII. [Figure 5]. All laboratory results were within normal parameters.

In January 2018, a suboccipital craniectomy and subtotal resection of the tumor were performed. A soft, extradural, neovascularized tumor, which eroded the adjacent occipital squama, was found. The patient had no postsurgical complications.

At the pathology laboratory, irregular, soft, fractious, brownish-white tissue fragments were processed; they measured 6 × 5 × 1.5 cm approximately. No bone fragments were processed. The histopathological analysis showed a neoplasm which was made up of plasma cells; most were mature, with minimal variation in nuclear morphology. IHC: CD138-positive diffuse expression 3+/3+; CD79-positive focal expression 2+/3+; CD19-positive focal expression 2+/3+ (<5%); CD56-positive diffuse expression 2+/3+; CD117 and cyclin D1 negative; kappa negative; lambda-positive focal expression 3+/3+ (5%); and proliferative rate (Ki-67) 5% [Figure 6]. She received fractioned radiotherapy, but during her 6-month follow-up consultation, she showed relapse.

DISCUSSION

SBP presenting as a posterior fossa tumor is extremely rare,[3] there is no statistics as to its prevalence worldwide, which is why we decided to report these cases. The most

common reported location is axial skeleton, predominantly thoracic and lumbar vertebrae.[7] Plasmacytoma is a differential diagnosis for tumors in this location,[3] they have been reported like falcotentorial tumors, both supra and infratentorial.[1]

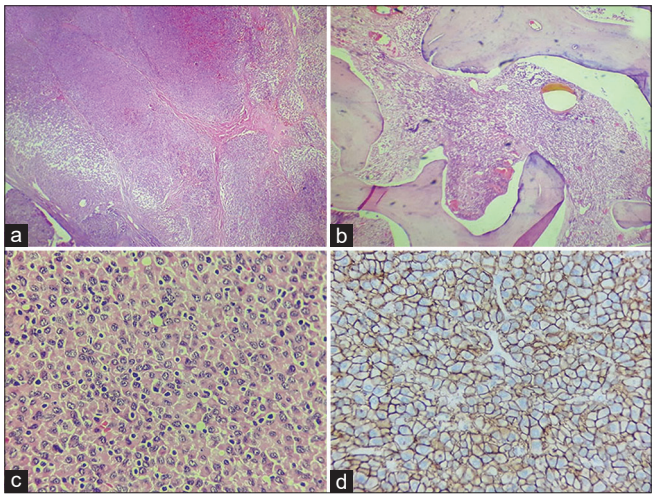

Figure 3: Photomicrograph of Case 1. (a) Panoramic image which shows sheets of neoplastic cells separated by fibrous septa (HE; ×50), (b) they infiltrate the medullary cavity of trabecular bone (HE; ×100). (c) Close-up which shows plasmacytoid cells and atypical plasma cells associated to small, reactive lymphocytes (HE, ×200). (d) IHC for CD138 which is positive in the cytoplasmic membrane (immunoperoxidase, ×250).

Approximately 5% of patients with multiple myeloma (MM) are initially diagnosed with SBP.[8] In the presented clinical cases, MM was ruled out during follow-up due to absence of clinical criteria. In most series reported, the average age for presentation is 55 years, with a 1.9:1 male-to-female ratio.[2,8]

The clinical-radiological differential diagnosis for a tumor in the posterior fossa includes meningiomas, sarcomas, primary bone tumors, and metastatic carcinoma,[2,7] all which were considered in the presented cases.

Due to its common location in the skull base, these tumors frequently affect the cranial nerves. Other reported clinical presentations are headache and intracranial mass effect.[6,8] In the presented cases, both had lesions with intracranial mass effect. One presenting with obstructive hydrocephalus and cerebellar syndrome; the second case presenting with cranial nerve palsy.

Regarding radiological presentation, plasmacytomas present in an unenhanced CT scan as osteolytic lesions without a sclerotic periphery. SBP presents on MRI as an enhancing lesion, iso- or hyperintense on T1-weighted images, and iso- or hypointense in T2-weighted images. These characteristics were observed in our patients' CTs and MRIs. On an unenhanced CT scan, the lesions' sharp edges, lack of sclerosis, and minimal periosteal reaction strongly correlate with a plasmacytoma. This suggests that a paramount study to identify bone remodeling is unenhanced CT scan [Figure 7]. Sometimes, a "mini-brain" appearance

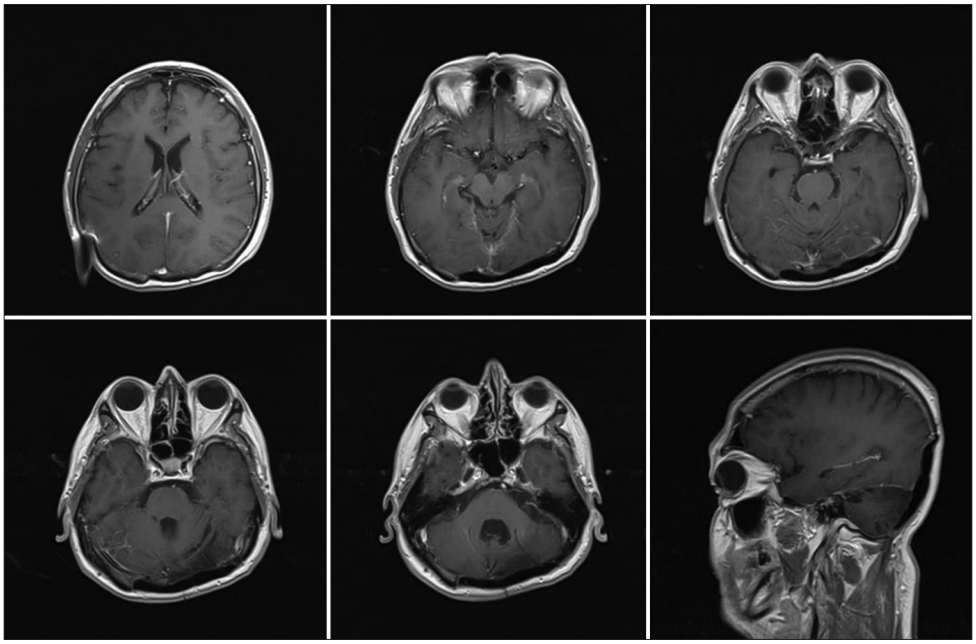

Figure 4: Six-month postoperative T1-weighted contrast-enhanced brain MRI of Case 1. Local disease control with radical resection of the tumor is demonstrated.

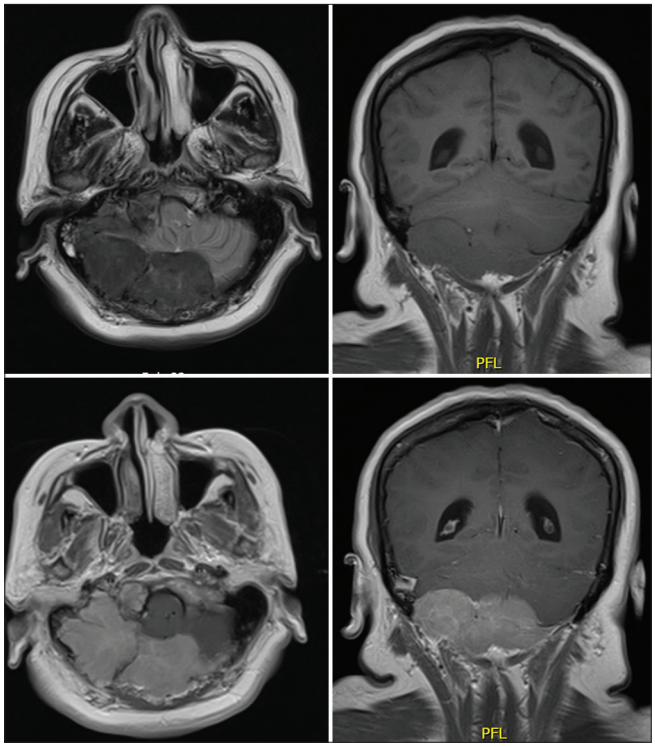

Figure 5: T1-T2-weighted and T1-weighted contrast-enhanced brain MRI of Case 2. There is widening of the cancellous bone of the occipital squama and the petrous bone.

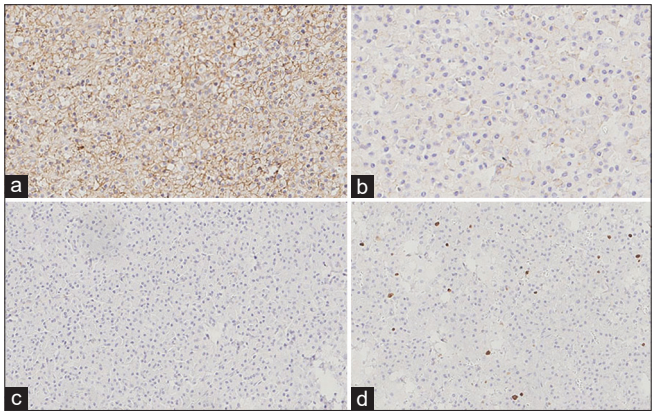

Figure 6: Photomicrographs of IHQ for Case 2. (a) CD56 which is positive in the cytoplasmic membrane. (b) CD117 negative. (c) Cyclin D1 negative. (d) Ki-67 (MIB1) which shows a proliferation index of 5% (a-d: immunoperoxidase).

can be seen, as described for the 1st time by Major *et al.* in his spinal plasmacytoma series;[2,7] this was not observed in the presented cases. The tumor characteristics, specifically in posterior fossa, in radiological studies, as reported in the literature, are presented in [Table 2].

In the histopathological study, plasmacytoma is a malignant proliferation of sheets of plasma cells with few scattered

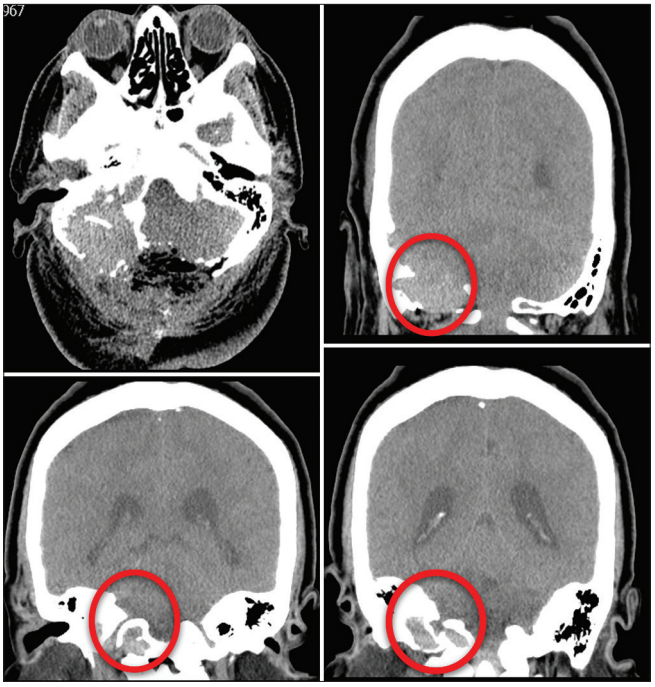

Figure 7: Six-month postoperative CT scan of Case 2 in axial and coronal views. Bilateral progressive widening of the cancellous bone of the squamous portion of the occipital bone and of the left condyle can be observed. There is also invasion to the adjacent petrous bone. It has been referred to as an osteolytic lesion without a sclerotic border. There is residual unresectable tumor due to blood loss during surgery.

cells of different lineage. Tumors can be made up of small mature plasma cells with small round nuclei, their chromatin grouped peripherally; but they can also have bigger cells with a plasmablastic morphology, with big, atypical nuclei, vesicular chromatin patters, and prominent central nucleoli. Cytological atypia is variable. The cytoplasm is moderately abundant in most cases, eccentric, and may contain inclusions of condensed immunoglobulins.[2] The IHC for diagnosis and prognosis for abnormal plasmatic cells is as follows: low CD19 and CD27 expression, weak CD38 and CD45 expression, higher positive expression of CD28, CD33, and CD56, and a variable expression in a small proportion of cells of CD20 and CD117.[5] The presented cases exemplify the heterogeneity in the expression of these markers. It is worthy to emphasize that CD117 and cyclin D1, poor prognosis markers for more aggressive disease (regarding MM), were negative. There are no reported specific prognostic IHC markers for SBP.

After an SBP diagnosis, patients need to be evaluated to rule out MM with serum electrophoresis, Bence-Jones protein in urine, PET scan, and bone marrow biopsy;[7] these tests were done during follow-up in our cases, ruling out MM.

After a bibliographical review of surgical resection of plasmacytomas, we found in an article from Na'ara *et al.*

Table 2: Features of the different case reports of SBP from posterior fossa in the literature.

Authors	Age	Sex	Clinical presentation	Localization	Features in TC	Features in MRI		Diagnosis of MM	Surgical treatment	Postsurgical treatment
						T1/T2 weighted	T1 weighted contrast enhanced			
Someren (1971)	47	F	Headache Ataxia	Posterior fossa (from the tentorium to the foramen magnum)	N/S	N/S	N/S	No	TR	N/S
Weisberg (1985)	60	M	Headache Ataxia	Right posterior fossa	Hyperdense without osseous erosion	N/S	Homogeneous enhancement	Yes	TR	N/S
Patel (2010)	42	F	Headache	Right posterior fossa	Hyperdense with osseous erosion	(↑)/(↓)	Homogeneous enhancement	Yes	PR	Steroids Chemotherapy Radiotherapy
Daghighi (2012)	37	M	Headache	Right posterior fossa	Hyperdense with osseous erosion	(-)/(-)	Homogeneous enhancement	No	TR	Steroids Radiotherapy
Frierich (2014)	45	M	Headache Vomiting	Left posterior fossa	Hyperdense without osseous erosion	(↑)/(↓)	Not acquired	Yes	PR	Radiotherapy

TR: Total resection, PR: Partial resection, N/S: Nonspecified, ↑: Hyperintense, ↓: Hypointense

that a R0 resection (microscopically margin-negative tumor bed) was achieved in 45% of the patients, R1 resection (microscopic margins positive for tumor) in 18% and R2 resection (gross residual tumor) in 37%.[8] It is worthy to mention that these terms are used in oncologic surgery for EMP and are thus not commonly used in neurosurgery. However, this concept is straightforward and can be used for SBP. Surgery is the preferred treatment for small accessible tumors to avoid the possible complications of radiotherapy,[8] and the objective of achieving a curative resection applies to this type of tumors.

There is evidence that if a R0 resection is achieved, radiotherapy can be withheld until there is tumor recurrence. Radiotherapy generally requires a 40–50 Gy dose administered over 20–25 fractions over a 4-week period. This is adjusted depending on the tumor's size. Favorable response to radiotherapy is usually over 80%.[2,9] A R0 resection seems decisive in the presented cases. Even though both patients received postoperative radiotherapy, relapse was documented during follow-up of the patient with subtotal resection. This can be due to multiple factors, some intrinsic to the tumor, but one cannot deny the evidence that suggests that prognosis improves with more extensive resection. Since the origin of the tumor is osseous, a complete resection can be achieved without sacrificing cerebral tissue.

A progression to MM from SBP has been established, thus chemotherapy should be initiated in patients in which this progression is documented.[2,9] During follow-up consultations of the presented patients, there was no progression to MM. Prognosis for SBP and EMP is better compared to MM.[7]

CONCLUSION

Treatment for a cranial tumor that is suspected to be SBP requires a multidisciplinary team to diagnose, plan a total resection and after surgery continues with the follow-up of the patient. SBP should be considered as a differential diagnosis for a tumor that produces osseous changes, which can be particularly observed in a unenhanced CT brain scan [Figure 7]. We consider that in patients with giant lesions, with indolent, progressive clinical course in posterior fossa, it is important to differentiate between SBP, meningioma, and chordoma to accurately define a radical surgical plan, if feasible.

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.

REFERENCES

1. Alappatt JP, Anto D, Ajayakumar A. Falcotentorial plasmacytoma: A case report. Surg Neurol 2004;62:178-9.
2. Bouali S, Taallah M, Bahri K, Zehani A, Abderrahmen K, Kallel J. Solitary plasmacytoma of the skull vault: A rare case report. Neurochirurgie 2020;66:66-9.
3. Daghighi MH, Poureisa M, Shimia M, Mazaheri-Khamene R, Daghighi S. Extramedullary plasmacytoma presenting as a solitary mass in the intracranial posterior fossa. Iran J Radiol 2012;9:223-6.
4. International Myeloma Working Group. Criteria for the classification of monoclonal gammopathies, multiple myeloma and related disorders: A report of the international myeloma working group. Br J Haematol 2003;121:749-57.
5. Kumar S, Kimlinger T, Morice W. Immunophenotyping in multiple myeloma and related plasma cell disorders. Best Pract Res Clin Haematol 2010;23:433-51.
6. Ma XJ, Li D, Wang L, Hao SY, Zhang LW, Zhang JT, *et al*. Clinical features, radiological profiles, and surgical outcomes of primary intracranial solitary plasmacytomas: A report of 17 cases and a pooled analysis of individual patient data. J Neurooncol 2019;142:263-72.
7. Munjal S, Srivastava A, Jain S, Mehta V. Skull vault plasmacytoma mimicking parasagittal meningioma: Mini-brain appearance. Asian J Neurosurg 2019;14:231-3.
8. Na'ara S, Amit M, Gil Z, Billan S. Plasmacytoma of the skull base: A meta-analysis. J Neurol Surg B Skull Base 2016;77:61-5.
9. Webb M, Barrett C, Barrett S, van Rensburg JJ, Louw V. Cranial plasmacytoma: A case series and review of the literature. Indian J Hematol Blood Transfus 2013;29:43-7.

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