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Case Report

Combined endoscopic transorbital and transnasal approach for the management of a solitary plasmacytoma of the sphenoid bone: A case report and literature review

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ABSTRACT

Background: Parasellar plasmacytomas are rare neurosurgical entities. Intrinsic characteristics of these tumors, such as adjacent bone erosion and symptoms resulting from invasion and mass effect, may lead to the possibility of a solitary extramedullary plasmacytoma (SEP) as a differential diagnosis.

Case Description: We present the case of a 39-year-old male with a 1-month history of bilateral decreased visual acuity, retroocular pulsating pain, and chromatic vision loss. A computed tomography scan of the head revealed a parasellar lesion causing chiasmatic compression, as well as clival, orbital, sphenoidal, and ethmoidal invasion. A combined transorbital and endonasal endoscopic approach was found suitable, and gross total resection was achieved. Histological analysis of the lesion established the diagnosis of a SEP. After radiotherapy, a new magnetic resonance imaging was performed, revealing a recurrence of the lesion with a high grade of invasion. The patient was treated with palliative radiotherapy, as surgical resection did not seem feasible.

Conclusion: Surgical resection and radiotherapy may achieve remission of these lesions; however, recurrence rates remain high despite any treatment modality. Patients with this condition must be followed up with a multidisciplinary team due to the high risk of multiple myeloma progression.

Keywords: Endoscopic endonasal approach, Parasellar plasmacytoma, Solitary plasmacytoma, Transorbital endoscopic approach

INTRODUCTION

According to the International Myeloma Working Group, a solitary plasmacytoma (SP) can be classified as either a solitary extramedullary plasmacytoma (SEP) or an SP of the bone (SPB), in which a single mass of plasma cells with no evidence of bone marrow malignant transformation, is located in a single area of bone with no soft-tissue infiltration.[9,11] SBP accounts for 1–5% of plasma cell tumors, whereas SEP is characterized by the presence of plasma cells infiltrating soft tissue with a prevalence of 2%.[18,22,21] SPB primarily affects vertebrae, femurs, pelvis, and ribs, which are red-marrow-containing bones, whereas sinonasal and skull base affection is exceedingly

rare.[4] Due to their rarity, parasellar plasmacytomas (PP) are often initially misdiagnosed as invasive lesions centered around the sellar or clival regions, such as pituitary adenomas or chordomas, and are believed to arise from the clival and sellar mucosa.[3,8] Given its slow progression, PP can be difficult to diagnose at early stages, and it is not until visual, oculomotor, or nasal symptoms arise that a prompt surgical intervention to ameliorate the symptoms is required. In this paper, we present a rare PP case that was surgically approached through a combined endoscopic transorbital and endonasal route.

CASE PRESENTATION

A 39-year-old male was admitted to the emergency department with a 1-month history of bilateral decreased visual acuity, intraocular pulsating pain, and loss of chromatic vision. Antibiotics and steroids were prescribed before his admission, causing symptom relief. However, recurrence after medication withdrawal prompted him to seek medical attention at our center.

A mass in the nasal region with extension toward the right eye was found. On neurological examination, total blindness with loss of photomotor responses was found in the right eye, while vision in the left eye was preserved (20/40). An initial diagnosis of optic neuritis was made, and computed tomography (CT) scan revealed a 5.6 × 4.6 × 5.1 cm heterogeneous solid sellar lesion with chiasmatic compression, as well as clival, orbital, sphenoid, and ethmoid invasion. Protrusion into the nasal cavities and right maxillary sinus was also outlined [Figure 1]. Thus, surgical management through a combined endoscopic transorbital and transnasal approach was deemed optimal for treating this lesion. An endoscopic endonasal approach (EEA) provides a panoramic view of the sellar lesion, as well as the invading portions in the nasal and maxillary cavities; however, to access the orbital portion, a transorbital approach (TOA) would be necessary. A single event with a two-staged surgery was performed with an EEA and a TOA, achieving gross total resection (GTR).

TOA

After conventional neuroanesthetic procedures, the patient was placed supine with the head in a neutral position. A superior eyelid crease incision was made, exposing the superolateral border of the orbital rim after the orbicularis muscle was incised. A subperiosteal dissection from the lateral margin of the orbit until identifying the superior orbital fissure (SOF) on the posterior wall of the orbit was performed. The recurrent meningeal artery was cauterized before reaching the SOF. High-speed drilling using a 3 mm diamond burr was performed from lateral to medial,

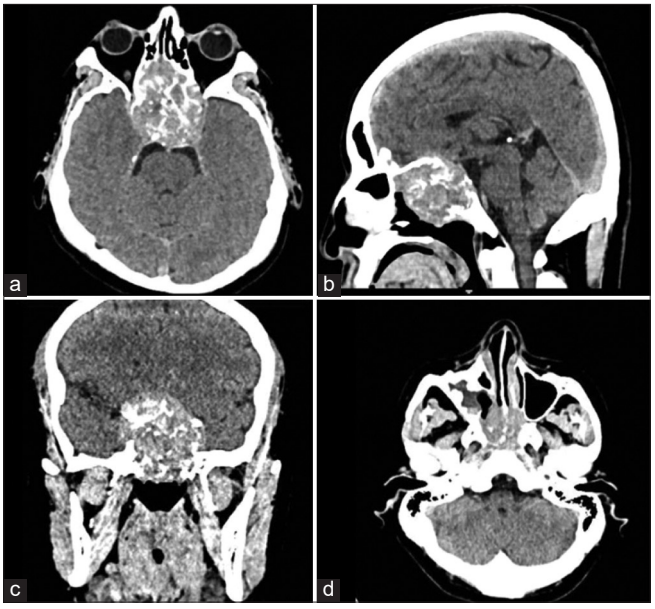


Figure 1: Preoperative computed tomography scan. (a) Axial, (b) sagittal, and (c) coronal sections in which a heterogeneous solid sellar lesion with clival invasion and suprasellar extension is observed. (d) Protrusion into the nasal cavities and right maxillary sinus are observed.

protecting the periorbita with a malleable retractor. The frontal and temporal dura were exposed, and the lesser sphenoid wing was removed using the diamond burr and Kerrison rongeurs. Partial extradural clinoidectomy was achieved, and the supraorbital and lateral component of the tumor was removed. A dural opening was not necessary due to the extradural location of the tumor.

EEA

As for the TOA, a 0° angled rigid endoscope was used. The right nostril was approached first. The lesion was immediately reached as the sphenoidal sinus and the nasal fossa were occupied by the tumor. A soft and grayish lesion was resected using biopsy rongeurs and ringed curettes. Resection of all the tumor portions located inside the sphenoid sinus was achieved. A full decompression of the right optic nerve was completed after removing the infiltrated bone from the roof and floor of the optic canal. Once the GTR of the lesion was achieved, the dura was exposed with no evidence of cerebrospinal fluid leak. Neuronavigation was used to confirm the limits of the tumor, and hemostasis was performed using gelfoam on the surgical site.

Morphological and immunohistochemical techniques revealed plasma cell dyscrasia and positive staining for lambda light chains consistent with extramedullary plasmacytoma. A PET scan with fluoro-D-glucose ruled out multiple myeloma (MM). A postoperative CT scan revealed

GTR [Figure 2]. Final examination on discharge showed no perception of smells and no visual improvement of the right eye albeit no additional ophthalmic deficits nor cranial nerve palsies (CNP) were observed. The postoperative course was uneventful, and the patient was discharged five days after surgery. At one month follow-up, the patient remained asymptomatic with minimal scarring in the periorbital region [Figure 3] and was referred for radiation protocol, which has been proven to be beneficial given the high radiosensitivity of these lesions.[12] Surgical technique is broadly described and available in video 1.

Four months later, the patient consulted due to bilateral loss of vision. A magnetic resonance imaging (MRI) was ordered showing the recurrence of the sellar lesion [Figure 4]. The tumor showed a greater degree of invasion of the right orbit, nasal cavity, sellar, and suprasellar regions, as well as extension into the right sylvian cistern. The lesion in this patient shows a very aggressive course with a high degree of local invasion. Surgical treatment for the recurrent tumor was not considered to be beneficial for the patient or curative. Thus, palliative radiotherapy was initiated.

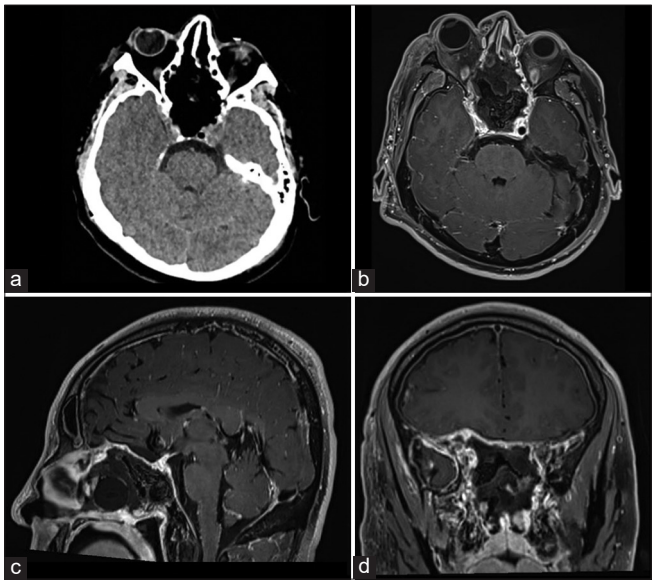


Figure 2: (a) Immediate postoperative computed tomography scan demonstrating gross total resection (GTR) of the lesion. Postoperative day four magnetic resonance imaging (b) axial, (c) sagittal, and (d) coronal sections with GTR of the lesion.

DISCUSSION

Solitary PP shows a slight male predominance, typically manifesting with mass effect and elevated intracranial pressure symptoms rather than endocrine irregularities. To the best of our knowledge, 11 cases of PP have been reported, with series including both sellar and parasellar lesions making up almost 80 cases.[1,12,17] Lee *et al*. reviewed 70 documented cases of sellar and PP, revealing that the most prevalent symptoms encompassed visual disturbances, CNP, and headaches.[17] Furthermore, DiDomenico *et al*. confirmed these findings, identifying headache and diplopia as the predominant clinical manifestations in their cohort.[8] Notably, endocrine abnormalities were relatively infrequent, with hyperprolactinemia emerging as the most prevalent hormonal aberration in these cases.[8,17] In our case, the patient presented with symptoms warranting an assessment for optic neuritis, notably devoid of any discernible CNP or endocrine irregularities. It is noteworthy that although literature exists delineating instances where pituitary neoplasms may mimic optic neuritis, our review yielded no publications documenting PP presenting with this particular symptomatology.[14,19]

Surgical approach

Plasmacytomas exhibit a propensity for infiltrating the cavernous sinus and inducing erosion of parasellar bones at a notably higher frequency in comparison to adenomas. This proclivity poses a challenge for surgeons in achieving

Figure 3: (a) Incision projection along the superior eyelid crease. (b) Exposure of the superolateral border of the orbital rim. (c) Immediate appearance of the eyelid reconstruction. (d) Residual scarring (red arrow) at follow-up shows no relevant esthetic disturbances.

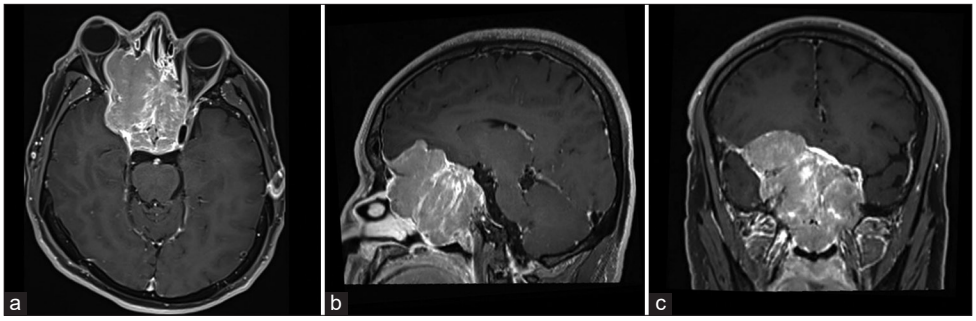


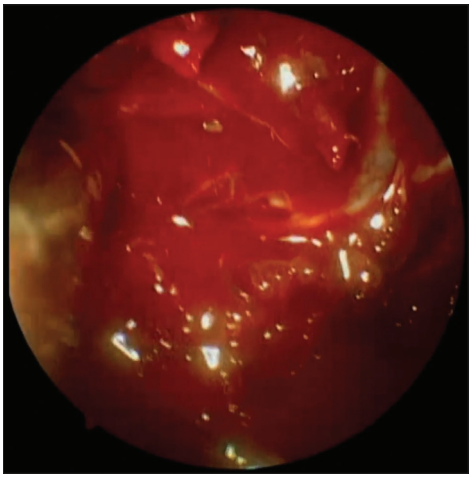
Figure 4: Magnetic resonance imaging with recurrence of the lesion in (a) axial, (b) sagittal and (c) coronal cuts with invasion in the right orbital, sellar, suprasellar and parasellar compartments.

GTR of the lesion, with an attainment rate as low as 22%. The extent of bone invasion is directly associated with the severity of CNP and diplopia while inversely impacting the incidence of visual disturbances.[8,17] In our case, despite the absence of CNP, preoperative MRI revealed chiasmatic compression accompanied by multicompartmental infiltration. Consequently, we deemed it necessary to employ a combined surgical approach with an EEA and TOA to ensure GTR, both of which have been proven to provide adequate surgical corridors with good resection and low complication rates.[7]

The EEA has remained the preferred surgical modality for lesions within the sellar compartment due to its proven effectiveness and safety profile.[5,15,20] Nevertheless, factors such as the degree of invasion into adjacent tissues and anatomical distortions can limit optimal visualization of the lesion, impeding the surgeon's ability to access the entirety of the tumor for complete resection. This necessitates the exploration of alternative approaches or combined strategies, like the EEA combined with the TOA. The TOA emerges as a viable option for access and visualization of lesions infiltrating the orbital and intracranial compartments, often referred to as cranio-orbital tumors. By removing the greater and lesser sphenoid wings while preserving the orbital rim, this approach permits exposure of the targeted region with a wider angle of attack and comparable levels of bone removal when compared to supraorbital approaches, like keyhole craniotomies. Implementing the TOA can significantly contribute to the successful planned resection of the lesion and improvement of associated ophthalmic comorbidities, including exophthalmos and optic neuropathy.[6,16] Its primary benefits include positive patient outcomes and excellent cosmetic results, coupled with minimal morbidity and quick postoperative recovery.[24]

Recurrent lesions or progression to systemic dyscrasias

Most patients with SEP can undergo remission, with <30% of cases progressing to MM or multiple tumors in a 10-year risk rate of progression, facing worse outcomes in mortality and morbidity in contrast to an SP, foregrounding the importance of determining if the disease is either localized or a systemic plasma cell dyscrasia.[2,10,23] Regional lymph node recurrence of SEP can rise to 7%.[2] Delaying radiotherapy after surgical resection can result in a high rate of recurrence.[13] Patients should be followed radiologically for the evaluation of the regional status of the lesion.



Video 1: Combined endoscopic transorbital and transnasal approach for the management of a solitary plasmacytoma of the sphenoid boneVideo is accessible from the portal.

CONCLUSION

PP remains a rare entity in the intracranial compartment and, when present, often masquerades as pituitary neoplasms. Intrinsic characteristics of the tumor may prompt the physician to consider the possibility of a SEP. Surgical treatment followed by radiotherapy remains the mainstay course of treatment in such cases.

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Ethical approval

Institutional Board approval is not required.

Declaration of patient consent

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

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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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