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A Case of Sphenoid Plasmacytoma

ABSTRACT

Extramedullary plasmacytoma is a rare pathology of the upper aerodigestive tract, of which sphenoid plasmacytoma is a rare variant. It usually presents like a sellar mass, with headache being the most common symptom, and severity of other symptoms proportional to the extent of invasion of surrounding structures. The importance of identifying red flag symptoms associated with headache cannot be exaggerated, in this regard. Bereft of specific clinical or imaging findings, it can prove to be a diagnostic dilemma. Standard protocols of treatment of multiple myeloma are followed, due to the rarity of the condition and the lack of site specific data thereof. Subject to resectability, the outcome with multimodality therapy is generally good. We describe our own experience in dealing with this relatively scarcely described condition with respect to diagnosis, surgery, additional evaluation, adjuvant therapy, and prognostication.

Key words: Extramedullary plasmacytoma, Myeloma, Red flags of headache, Sphenoid plasmacytoma

INTRODUCTION

Plasmacytoma is a discrete mass of neoplastic monoclonal plasma cells. It most commonly involves marrow of flat bones, that is, pelvis and vertebrae. Less commonly (<10%) it produces solitary involvement of sites away from flat bone marrow, which in turn may be of two types: (1) Solitary bone plasmacytoma (5%) and (2) extramedullary plasmacytoma (EMP) (3%). Sphenoid plasmacytoma represents a very small subset of EMP. There are around 65 cases of sphenoid plasmacytomas described internationally. A PubMed search revealed only 25 cases of solitary sphenoidal plasmacytoma reported worldwide. The disease is more common in men and may present with nothing but an innocuous headache. Identifying red flag symptoms associated with headache can prove invaluable in such situations.[1] Clinical and radiological signs are nonspecific, and histopathology provides the only definitive diagnosis.

CASE REPORT

A 56-year-old gentleman, complaining of mild intermittent headache for 2–3 months, with a normal neurology, underwent computed tomography (CT) brain, which led to the incidental detection of an osteolytic sphenoidal mass. Further contrast-enhanced CT and contrast-enhanced magnetic resonance imaging brain showed a homogenously enhancing solid osteolytic, extradural, sphenoidal mass, without neurovascular involvement [Figure 1]. The hormone profile was within normal range. The diagnosis being uncertain, a decision was made to perform transnasal endoscopic excision [Figure 2]. Frozen section suggested – (1) invasive pituitary adenoma and (2) plasmacytoma (myeloma). Gross total resection was carried out. Postoperatively, the patient's headache was completely relieved.

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Final histopathology with immunohistochemistry confirmed a plasmacytoma/myeloma [Figure 3]. Serum electrophoresis revealed M-protein with λ-chain restriction [Figure 4]; whole-body positron emission tomography (PET)-CT (^{18}FDG) confirmed complete excision and no systemic spread. With a diagnosis of EMP, the patient was referred to a medical oncologist. At 12 months post-operative, he has no recurrence of disease.

DISCUSSION

Etiopathogenesis: Plasmacytoma lies along the lower end of the spectrum of plasma cell dyscrasias: [2]

Monoclonal gammopathy of unknown significance → Plasmacytoma → Smoldering Multiple myeloma → Multiple myeloma → Plasma cell leukemia

EMP constitutes 3% of all plasmacytomas. About 85% of EMP are localized to the upper aerodigestive tract, most commonly, the paranasal air sinuses, where it forms a submucosal mass.

The criteria to differentiate solitary plasmacytoma from myeloma are[2] as follows:

1. Biopsy-proven solitary disease
2. Absence of marrow involvement
3. Absence of systemic manifestations of disease (CRAB), namely, hypercalcemia (C), renal insufficiency (R), anemia (A), osteolytic lesions (B)
4. Absence of skeletal involvement

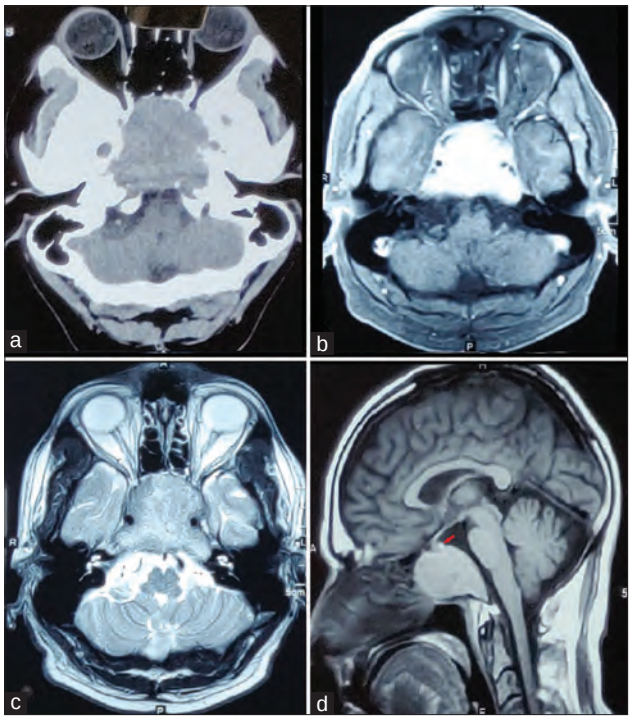

Figure 1: a: Computed tomography brain plain: Osteolytic homogenous sphenoidal mass, mildly hyperdense, without hemorrhage, necrosis, or calcification. b: Magnetic resonance imaging T1 axial with contrast: Solid homogenously enhancing sphenoidal mass. Smooth margins. No dural invasion. c: Magnetic resonance imaging T2 axial: Isodense solid sphenoidal mass, with a "waist" due to the indentation by ICAs. Extradural. d: Magnetic resonance imaging T1 sagittal: Isodense solid intrasphenoidal mass, with pituitary seen clearly separate (arrow)

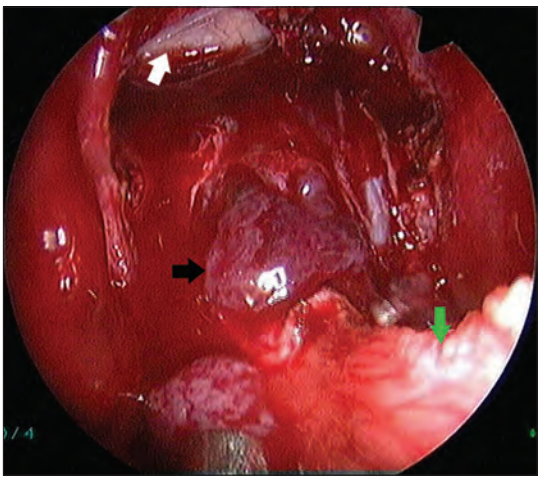

Figure 2: Intraoperative endoscopic image showing highly vascular purplish solid tumor (black arrow) in the sphenoid sinus. Optic chiasmal recess (white arrow) and nasal septum (green arrow) can be seen

Sphenoid plasmacytoma represents a very small rare subset of EMP. The origin is from mucosa associated lymphoid tissue of the paranasal sinuses, with the purported etiology being chronic low-grade infections. The role of Epstein–Barr virus has been elucidated.[3] Chromosomal aberrations include deletions in 13q,[4] 1p, and 14q or additions in 19p, 9q, and 1q. IL-6 is a proven driver of plasma cell proliferation.

The disease commonly affects persons above 40 years of age. It is approximately 3 times more common in men.

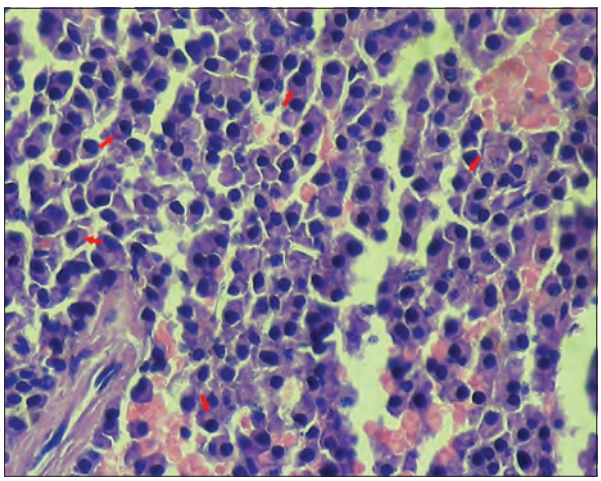

Figure 3: Histopathology showing a monomorphic mass of small round to polygonal cells, with perinuclear cytoplasmic eosinophilic mass, that is, perinuclear "hof" (red arrow)

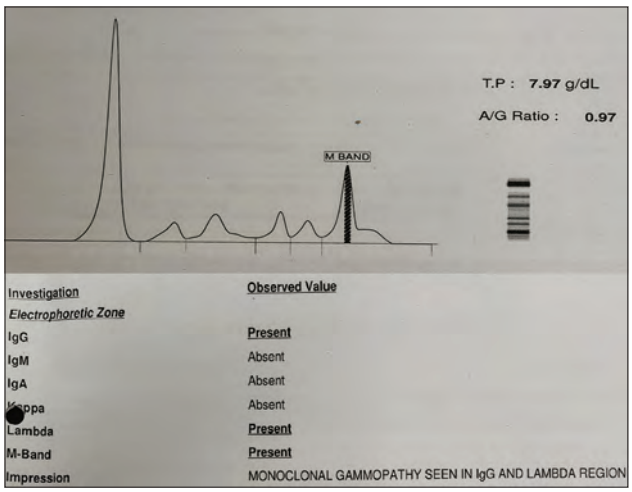

Figure 4: Serum protein electrophoresis showing IgG "M-band" with λ light chain restriction

Symptomatology

The most common symptom in sphenoid plasmacytoma is headache. This may or may not be associated with any combination of palsies of cranial nerves II, III, IV, V1, V2, and VI; depending on the extent of parasellar involvement. The presence of plain innocuous headache may lower clinical suspicion of any pathology, and the gravity of the symptom may tend to be underestimated. It is well to remember certain red flags in headache, which necessitate further investigation/imaging: [1]

- Abnormal neurological examination (other than typical aura), dizziness, incoordination, signs of systemic illness, and papilledema
- New headache in older patients, or risk factors for HIV infection or cancer
- Headache increasing in frequency and severity, or with Valsalva maneuver
- Worst headache ever
- Sudden onset of headache
- Headache subsequent to head trauma

Other symptoms may include nasal discharge, epistaxis, nasal obstruction, sore throat, hoarseness, dysphonia, dysphagia, and dyspnea.[5] There is a high likelihood of regional lymphadenopathy.

Imaging

There is no definite pathophysiologic appearance on imaging. The tumor is solid, homogenously enhancing, with osteolysis. It may infiltrate into adjacent neurovascular structures. Regional lymphadenopathy may be observed. Lesions consistently show hypermetabolic activity on ^{18}FDG-PET.

Pathology

The tumor is composed of a monomorphic sheet of plasma cells, which are small, oval cells with large nuclei, and a perinuclear "Hof" composed of Golgi apparatus.[6] Final diagnosis is made using immunohistochemistry, which stains positive for CD138 (syndecan-1) – a plasma cell-specific marker.[7] The tumor usually shows light chain restriction on IHC.[8]

About 20–25% of patients have a positive M band on serum protein electrophoresis. Positive M band 2 years post-therapy suggests high propensity for recurrence and progression to multiple myeloma (MM).[8] A newer indicator is a serum free light chain ratio, that is, ratio of serum concentrations of κ to λ light chains. When abnormal (normal range – 0.26–1.65) at diagnosis, this is a predictor of progression.[8]

Therapy

The tumors are highly radiosensitive, hence, radiotherapy forms an essential component of all therapy. Surgery may have limited role, due to location and extension/invasion. However, when resectable, surgery produces results equal to radiotherapy. Radical surgery is avoided, due to the unacceptable mutilation produced, and the radiosensitive nature of disease. The best control rates are with a combined approach.[9] The recommended dose of radiation is 40Gy for lesions less than 5 cm in size and 50 Gy when more than 5 cm; at 2 Gy per fraction.[9] Draining nodes may be included only if clinically involved.

Adjuvant chemotherapy, although controversial, may be offered to patients with lesion > 5 cm and/or with a high-grade histology.[9] Regimen is the same as for MM. Thalidomide has a role in cases of recurrent plasmacytoma.[9]

Treatment results

In a review of 721 cases by Alexiou *et al*., 65% of patients were free of recurrence and progression, 22% experienced recurrence, and 15% progressed into MM.[5] Recurrence rates appear to be independent of treatment modality.[5]

Long-term follow-up

The NCCN Guidelines for Clinical Practice in Oncology (Myeloma Section) recommend follow-up surveillance every 3–6 months: [9]

- CBC – Hb, TLC, and DLC
- Biochemistry – creatinine, albumin, and calcium
- Serum protein electrophoresis, 24 h urine total protein, and urine protein electrophoresis
- Serum-free light chain assay.

Prognosis

Prognosis of EMP appears to be generally good with 5-year survival rate of about 82% and 10-year disease-specific survival rate >50%.

There are three patterns of treatment failure – recurrence, dissemination (to other extramedullary sites), and progression (to MM).[10]

The rate of progression to MM ranges from 11% to 30% at 10 years.[10] Patients with EMP that progressed to MM had a 5-year survival rate of 100%.

Our Case

The patient in the reported case presented with the sole symptom of headache, and was incidentally discovered to have a mass, by a local practitioner. The diagnosis was unclear on pre-operative imaging. Frozen section biopsy report was inconclusive as well. Hence the patient underwent complete surgical excision, although it is not the recommended therapy for head neck plasmacytomas.

CONCLUSION AND LEARNING POINTS

- Plasmacytomas are a less aggressive subgroup of plasma cell dyscrasias. Sphenoid plasmacytoma is a rare variant of this rare disease.

- Headache may be the only symptom of sphenoid plasmacytoma at presentation.
- Red flag symptoms of Headache must be carefully evaluated1. New onset of headache in an older individual may portend a sinister diagnosis.
- Myeloma and plasmacytoma are essentially extradural diseases, and may thus undergo significant local advancement before producing any neurodeficit.
- Surgical treatment is not the first choice in solitary plasmacytomas of the head-neck region, however, if executed well, it may serve as an excellent modality for disease extirpation.
- Survival rates with multimodality treatment exceed 80% at 5 years.

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