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Beyond the Bone: Extramedullary Plasmacytoma as a Rare Otorhinolaryngological Surprise in Multiple Myeloma

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ABSTRACT

Extramedullary plasmacytoma (EMP) is an uncommon manifestation of Multiple Myeloma that may involve the nasal cavity and paranasal sinuses. We report a case of a known Multiple Myeloma patient on chemotherapy presenting with a nasal mass. EMP in the sinonasal region is clinically significant due to its varied presentation and resemblance to common nasal pathologies. Radiological findings were non-specific, highlighting the diagnostic challenge in such cases. Biopsy confirmed Plasmacytoma. EMP may indicate disease progression or relapse of Multiple Myeloma. This case underscores the importance of including EMP in the differential diagnosis of vascular or atypical nasal masses to ensure timely diagnosis, appropriate management, and improved prognostic assessment, particularly from an ENT perspective.

Keywords: Plasma Cell Neoplasms; Multiple Myeloma; Nasal Neoplasms; Epistaxis; Chemotherapy; Biopsy

*See End Note for complete author details

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INTRODUCTION

Multiple myeloma (MM) is a cancer of Plasma cells that arises in the bone marrow. Although it represents only a small fraction of all cancers, it makes up a notable proportion of hematological malignancies.[1] Plasmacytoma refers to a localized tumor formed by clonal plasma cells. Unlike Multiple Myeloma, it shows little or no bone marrow involvement and typically presents with symptoms limited to the site of the lesion. Two types of which include Extramedullary plasmacytoma (EMP), which occurs outside the bones, and Solitary bone plasmacytoma (SBP), which develops within skeletal tissue.[2] Extramedullary plasmacytomas (EMP), though very rare, affect the head and neck region, particularly the nasopharynx, oropharynx, and paranasal sinuses.[3]

CASE REPORT

A 55-year-old man who was diagnosed with Multiple Myeloma two years ago and is on maintenance chemotherapy with intravenous Bortezomib presented with a right nasal mass and recurrent epistaxis for three months. He also complains of increased right-sided nasal obstruction and reduced smell over the last two weeks, along with right-sided facial pain and reduced facial sensation. He is a known diabetic and hypothyroid patient with a history of Spinal Tuberculosis, for which he underwent Antitubercular Therapy. Additionally, there is a family history of bone cancer in his father. He has been an alcoholic and tobacco chewer for 30 years.

On anterior rhinoscopy, there is a reddish fleshy mass filling the right nasal cavity, and deviation of nasal septum to right. There is right maxillary sinus tenderness and reduced right-sided facial sensation. There is also mild right eye mild proptosis with normal vision and extraocular movements. There is also a 1cm smooth bulge in the hard palate on the right side close to the midline at the level of the right upper second molars **(Figure 1).**

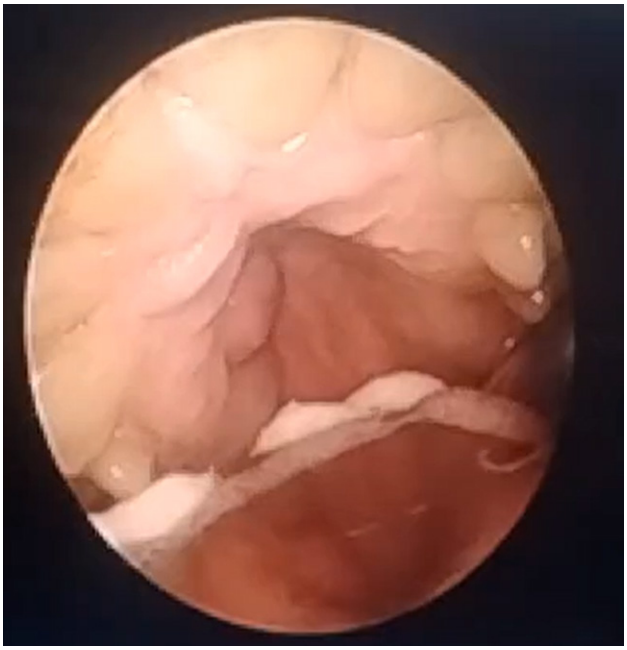

Figure 1: Palatal bulge

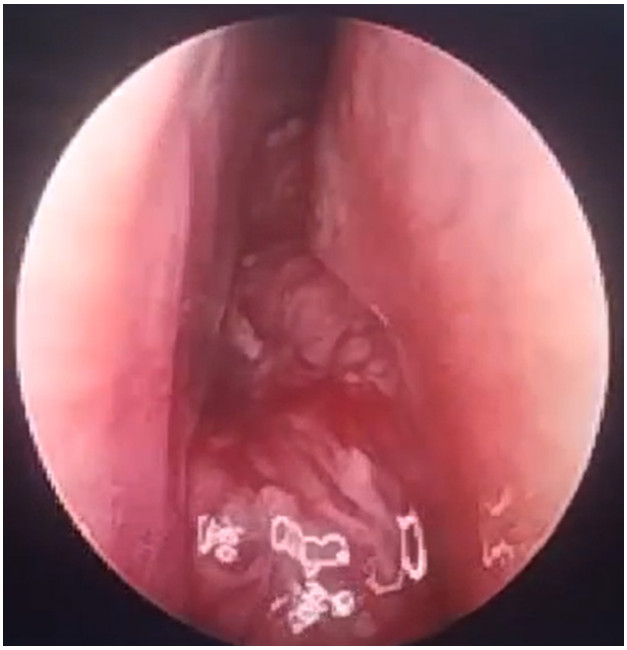

Figure 2: Vascular nasal mass

Diagnostic nasal endoscopy showed a smooth, reddish, fleshy mass filling the right nasal cavity **(Figure 2).**

Contrast-enhanced computed tomography of paranasal sinuses shows obliteration of the right maxillary sinus, osteomeatal unit, and adjacent middle meatus with minimal hyperdense contents within the sinus cavity. Diffuse sclerosis and rarefaction of the sinus walls were noted. Post-contrast imaging shows diffuse enhancement of maxillary sinus contents, likely severe chronic sinusitis/ chronic invasive fungal sinusitis/ malignancy **(Figure 3).**

A working diagnosis of Invasive Fungal sinusitis/Malignancy was made, and endoscopic excision of the nasal mass was planned under general anaesthesia. Intraoperatively, a firm, smooth, vascular mass was seen arising from the right maxillary sinus, filling the nasal cavity and partially obscuring the view of the middle and inferior turbinate. Biopsy taken, and sample was sent for impression smear, fungal smear, CBNAAT, and histopathological examination.

Impression smear indicative of a dense plasma cell-based chronic inflammatory lesion. No signs of fungal hyphae. Tissue culture was sterile, and CBNAAT was negative. The biopsy showed a neoplasm composed of cells arranged in sheets. These are small- to medium-sized cells with eosinophilic cytoplasm, a high N:C ratio, and a round, vesicular to hyperchromatic nucleus.

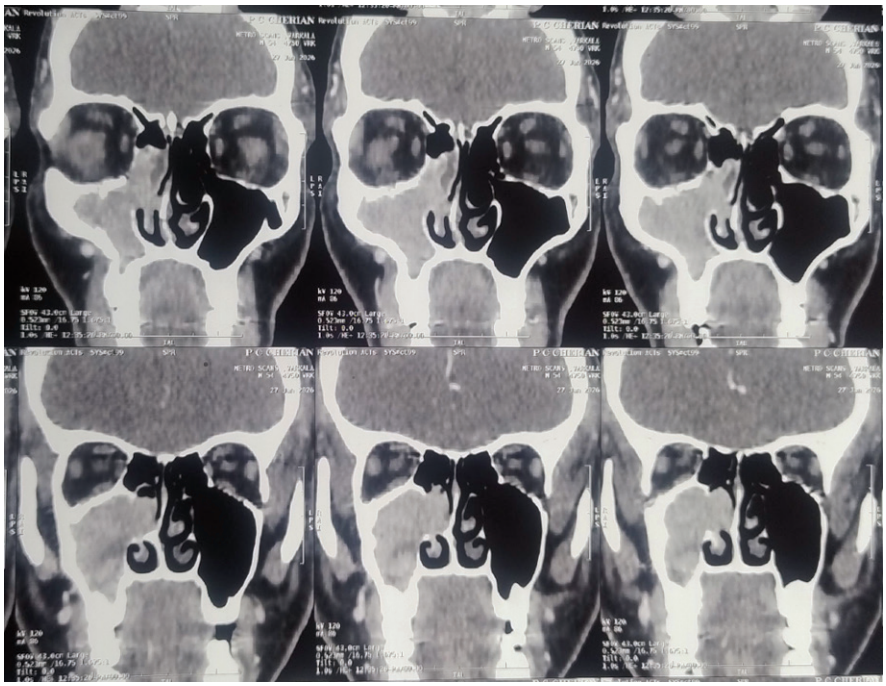

Figure 3: Contrast-enhanced computed tomography of paranasal sinuses

Mitotic figures, including atypical mitosis, were seen. The surrounding fibrous stroma shows inflammatory infiltrate composed of lymphocytes, histiocytes, and neutrophils with focal areas of apoptotic debris and hemorrhages. In Immunohistochemistry, tumor cells are strongly positive for CD138, kappa-restricted, and lambda-negative. Hence, morphology and IHC studies were consistent with plasmacytoma.

The patient was referred to Oncology for further management and is planned for chemoradiotherapy. In view of Extramedullary Plasmacytoma associated with Multiple Myeloma, local radiotherapy is planned for the maxillary sinus lesion along with chemotherapy for the management of multiple myeloma.

DISCUSSION

Extramedullary plasmacytoma is an uncommon Plasma cell neoplasm comprising approximately 3–5% of Plasma cell dyscrasias. About 80% of these arise in the head and neck region, with 38% which is seen in sinonasal cavity.[4] Owing to its nonspecific clinical presentation, including nasal obstruction, nasal mass, rhinorrhoea, and epistaxis, Extramedullary Plasmacytoma of the sinonasal tract can resemble a variety of inflammatory, infective, and neoplastic lesions. Important differential diagnoses in this case include Invasive Fungal Sinusitis and Metastatic deposit.[4] Previous case reports have shown osteolytic punched-out lesions in the mandible and rarely in the maxilla, accompanied by tooth mobility and localised swelling.[5] Even among reported maxillary lesions, clinical presentation as a soft tissue mass is extremely rare.

Because clinical and radiological findings may mimic other sinonasal conditions, histopathological examination is the cornerstone of diagnosis. The presence of sheets of plasma cells, along with CD138/CD38 positivity and monoclonal kappa or lambda light chain restriction, helps confirm the diagnosis.[6,7]

In Extramedullary Plasmacytoma associated with Multiple Myeloma, management includes surgery, usually limited to diagnostic biopsy, followed by radiotherapy for local control of the plasmacytoma. Chemotherapy for the management of Multiple Myeloma includes bortezomib-, lenalidomide-, and cyclophosphamide-based regimens. Supportive care is essential and includes treatment of anemia, renal dysfunction, infections, and metabolic abnormalities.[8-10]

In the aggressive terminal phase of Multiple Myeloma, malignant plasma proliferates autonomously outside the bone marrow microenvironment.[6,11] Loss of Cell adhesion receptors may enable Myeloma cells to evade control of the bone marrow, and hematogenous dissemination may lead to the development of isolated extramedullary plasmacytomas without direct anatomical association with bone.[6] At this stage, the disease is typically refractory to conventional chemotherapy, and hence, overall survival remains poor despite the use of high-dose chemotherapeutic regimens.[11]

From an Otorhinolaryngological perspective, this case highlights the importance of considering Plasma cell neoplasms in the differential diagnosis of vascular or atypical nasal masses. Histopathological examination remains the gold standard for definitive diagnosis, and a multidisciplinary approach is crucial for appropriate treatment planning and prognostication.

CONCLUSION

Extramedullary Plasmacytoma of the sinonasal tract is a rare plasma cell dyscrasia. In patients with Multiple Myeloma, the development of a sinonasal mass should raise suspicion of Extramedullary Plasmacytoma and possible disease progression.

END NOTE

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