

CASE REPORT

A Rare Case of Sinonasal Extramedullary Plasmacytoma with Orbital Involvement

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ABSTRACT

Extramedullary plasmacytoma (EMP) is a rare entity in the head and neck region. It is phenotypically and immunologically different from multiple myeloma. Chronic transformation to multiple myeloma or its disseminated form is well known. It may even be the first manifestation of multiple myeloma also popularly known as secondary EMP.

We present a 43-year-old male patient with history of persistent progressive nasal obstruction with occasional epistaxis. Endoscopy revealed a right sinonasal mass which on excision biopsy was consistent with features of EMP.

Extramedullary plasmacytoma is a rare differential diagnosis for sinonasal mass. However, multiple myeloma should be ruled out via bone marrow examination, bone scan and serum and urine electrophoretic evaluation before arising at a diagnosis of EMP.

In spite of EMP of nasal cavity being a rare entity, it should be considered as one of the rare differential diagnoses.

Keywords: Extramedullary plasmacytoma, Multiple myeloma, Sinonasal tumors.

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INTRODUCTION

Plasmacytomas are rare neoplasms which originate from B cells. They have been categorized into three variants namely multiple myeloma (MM), osseous solitary plasmacytoma and extramedullary plasmacytoma (EMP).¹

Solitary EMP is the rarest of all the three varieties. In order to establish a diagnosis of solitary EMP, evidence of bone destruction or occult disease anywhere in the body must be ruled out.²

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Extramedullary plasmacytoma account for approximately 4% of the nonepithelial sinonasal tumors and 0.4% of the head and neck tumors. These tumors are generally found in the 4th decade of life with male to female preponderance 3:1.³

The various etiologies proposed to explain the occurrence of for EMP are chronic stimulation, overdose or inappropriate irradiation, viral infections, and genetic alterations in the reticuloendothelial system.⁴

These tumors have varied clinical presentation depending of the site of affliction. Extramedullary plasmacytoma of sinonasal origin usually presents as progressive nasal obstruction and intermittent epistaxis, as was in our case.

Olfactory neuroblastoma, lymphoma, anaplastic carcinoma and metastatic deposits stand as potential candidates for differential diagnosis of such sinonasal neoplasms.⁵ We present the above case aiming at developing insights toward early diagnosis and appropriate treatment of this rare sinonasal tumor.

CASE REPORT

A 43 years old male patient presented to us with complaints of right-sided persistent progressive nasal obstruction, epistaxis for the last 10 months. He also gives history of associated diplopia and proptosis in right eye for last 6 months.

Anterior rhinoscopy revealed grayish, polypoidal, exophytic nonsensitive, friable mass in the right nasal cavity. Posterior rhinoscopy was suggestive of the mass reaching till choana with normal nasopharynx. Visual acuity and color vision was normal in both eyes. Nasal endoscopy confirmed above findings, mass was firm in consistency and bled on touch.

His lab reports revealed the following: Hb: 11.9 gm%, TLC: 10500/cc, DLC: P67, L23, M7, platelets 194 × 10³/L, ESR 140 mm/hr, blood urea: 45 mg%, S creatinine 1.07 mg%, S uric acid 5.7 mg/dl, S calcium 8.46 mg/dl, total protein 8.0 gm/dl, S albumin 5.32 gm/dl. Fungal stains and Xpert TB test were negative.

Computed tomography (CT) and magnetic resonance imaging (MRI) of neck and paranasal sinuses showed a heterogeneously enhancing well-defined mass in the right maxillary sinus and nasal cavity bulging into the orbit

along the floor of the orbit, retromaxillary fat pad and infratemporal fossa and ethmoid air cells (Figs 1 to 4).

Endoscopic biopsy was done. Histopathological features included solid sheets, vague nests and diffuse infiltrates of immature plasma cells displaying round to oval nuclei with moderate pleomorphism, occasional prominent nucleoli and moderate amounts of amphophilic cytoplasm (Fig. 5). Immunohistochemistry showed MUM-1: nuclear positivity in immature and mature plasma cells (Fig. 6).

Bone marrow aspiration and biopsy was normal. Serum electrophoresis was noncontributory. Myeloma band was not detected and urine examination showed no Bence Jones proteins. Final diagnosis was EMP.

The case was discussed in the multidisciplinary tumor board. He was administered adjuvant radiotherapy (40 Gy in 20 fractions). At 2 years postradiotherapy on follow-up, the patient exhibited no signs or symptoms of recurrence or progression to multiple myeloma.

DISCUSSION

Dalrymple and Bence-Jones first described plasma cell dyscrasias in 1846. This was illustrated in a patient

who presented with diffuse bone pain and marked proteinuria. Later in 1873 Rustizky et al attributed the above clinical entity to a histopathological diagnosis of multiple myeloma.

Plasma cell neoplasms are further subclassified as solitary plasmacytoma, multiple myeloma and plasma cell leukemia. Solitary plasmacytoma can be either intramedullary solitary bone plasmacytoma (SPB) or EMP.

Extramedullary plasmacytoma frequently arises from soft tissues. This clinical entity was first illustrated Schridde in 1905. These tumors may be either single or multiple. Most commonly, they present as a solitary lesion in the head and neck region. These have also been reported to occur in various other sites, such as lung, pleura, stomach, small bowel, colon, ovary, uterus, testes, kidneys, skin and breast.⁶

The course of the disease is usually indolent, and it remains symptomless till it achieves a significant size. Extramedullary plasmacytoma arising in sinonasal region often presents as gradually increasing hard and painful swelling, resulting in progressive nasal obstruction,

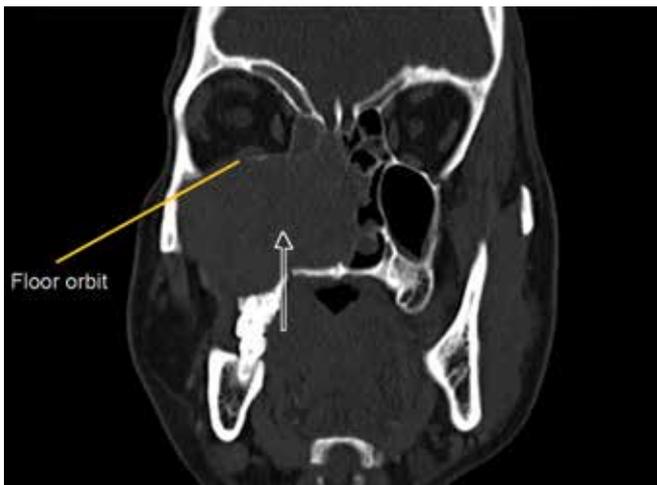


Fig. 1: Coronal view of CT scan



Fig. 2: Sagittal view of CT scan



Fig. 3: Magnetic resonance imaging coronal view

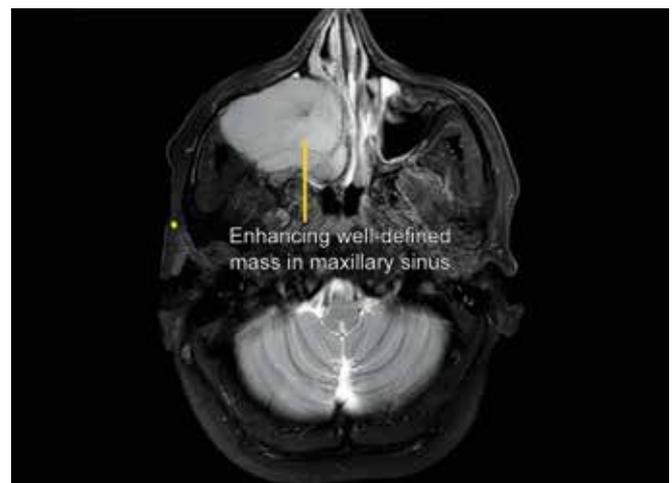


Fig. 4: Magnetic resonance imaging axial view

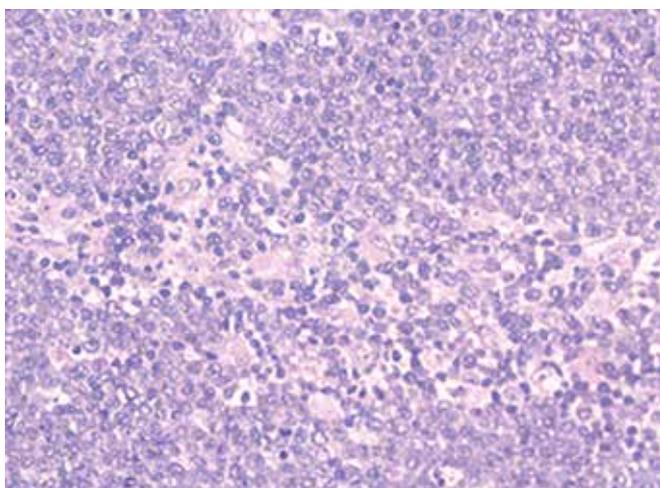


Fig. 5: Histopathological features included solid sheets, vague nests and diffuse infiltrates of immature plasma cells displaying round to oval nuclei with moderate pleomorphism

recurrent epistaxis and occasionally proptosis if orbital involvement is present.⁷

United kingdom myeloma forum has outlined broad guidelines to diagnose EMP which include: solitary extramedullary mass of clonal plasma cells, histologically normal bone marrow aspirate, normal skeletal survey, no evidence of anemia, hypercalcemia, or renal impairment due to myeloma, and low or absent serum and urinary monoclonal immunoglobulin.⁸

Universal absence of cyclin D1 expression and the frequent absence of CD56 in EMP, proves to be promising diagnostic markers for EMP as per a study by Lomeo et al.⁶

Histopathological gradation is based on cellular atypia EMP which can be categorized as low (grade 1), intermediate (grade 2) or high grades (grade 3).

Stage I is disease confined to one site. Stage II includes tumors with local lymph node involvement and stage III has metastatic diffuse spread. Our case was scored under grade 1 and stage 1 because of minimal evidence of cellular atypia with maximum resemblance to the parent cells and no evidence of local or distant spread.⁹

About 15 to 20% of EMP convert to MM over the years especially if the EMP involves the adjacent bone. The conversion rate of EMP to SPB is 48% which has a poorer prognostic outcome.¹⁰

The optimal management of EMP is controversial, however, reasonably good outcomes have been attained with combined surgical excision and adjuvant radiotherapy due to the good radiosensitivity of these tumors.^{11,12}

United Kingdom myeloma forum has recommended radiotherapy dose of 40 Gy in 20 fractions for tumors <5 cm and up to 50 Gy in 25 fractions for tumors ≥5 cm with at least a 2 cm margin encompassing the primary tumor. Positive nodes should be included in the radio-

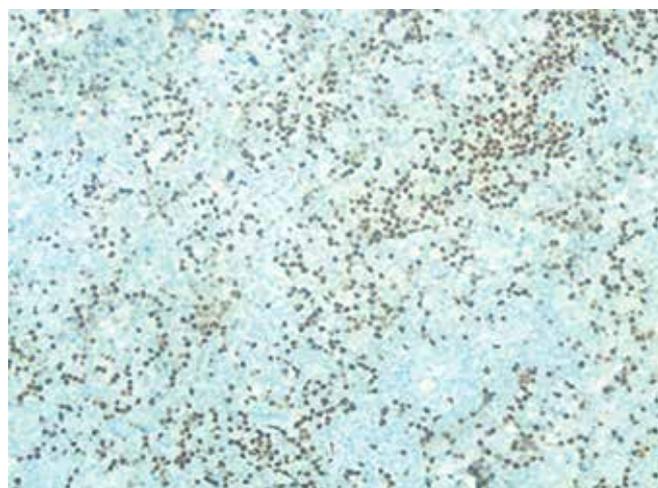


Fig. 6: Immunohistochemistry showed MUM-1: nuclear positivity

therapy field. Surgery should be avoided in cases where extensive disfiguring procedures are warranted or if the tumor lies in the vicinity of vital structures.¹³ Secondary EMP and/or disseminated cases of primary EMP are potential candidates for treatment with steroids and other medical treatment.²

CONCLUSION

Extramedullary plasmacytoma is rare sinonasal tumors which require fulfillment of specific histological, phenotypic and radiographic criteria. Management of such tumors requires multidisciplinary participation. Management is controversial, however, surgical excision along with adjuvant radiotherapy gives promising results. These patients need long-term follow-up as rare transformation to multiple myeloma is a possibility.

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