

## Case Report

# A rare diagnosis of nasal mass: extramedullary plasmacytoma

Shrinath Kamath, Deepika Pratap\*, Kishore Chandra Shetty, Biniyam

Department of ENT, KSHEMA, Nitte University, Mangalore, Karnataka, India

**Received:** 20 February 2018

**Accepted:** 26 March 2018

### \*Correspondence:

Dr. Deepika Pratap,

E-mail: [pratap.deepika@gmail.com](mailto:pratap.deepika@gmail.com)

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## ABSTRACT

Extramedullary plasmacytomas (EMPs) are localized plasma cell neoplasms that occur within the soft tissue. Solitary plasmacytoma of the head and neck is a rare disease entity. They account for 1–2% of all plasma cell growths and have a great predilection for the upper respiratory tract. Males are more frequently affected during the fifth and sixth decades of life. We report a case of maxillary sinus plasmacytoma in an elderly man. Complete resolution of the tumour was seen with radiotherapy.

**Keywords:** Extramedullary plasmacytoma, Maxillary sinus, Radiotherapy

## INTRODUCTION

Plasmacytoma is an extremely rare plasma cell neoplasm. It was first described by Schridde in 1905.<sup>1</sup> Based on its location; plasmacytoma can be classified into two groups; solitary plasmacytoma of the bone (SBP) and extramedullary plasmacytoma (EMP). Solitary plasmacytoma of the bone mostly occurs in the axial skeleton, while extramedullary plasmacytoma occurs in the head and neck region.<sup>2,3</sup> EMPs comprises approximately 3% of all plasma cell neoplasms.<sup>4</sup> The estimated global incidence of the disease is 1 case per 500,000 population.<sup>5,6</sup>

More than 90% of EMPs originate in the head and neck and affects the nasal cavity, sinuses, oropharynx, salivary glands, and larynx.<sup>7-9</sup>

## CASE REPORT

A 63 year old man presented with history of swelling on the left cheek since 3 months (Figure 1). It was gradually increasing in size with one episode of bleeding from left nostril. On examination he had a 5x6 cm swelling over his left cheek extending from the nasolabial fold to 1cm in front of the tragus, which was firm and non tender.

Superiorly it extended up to zygomatic arch; inferiorly it extended from 2 cm lateral to angle of mouth to 2 cm anterior to angle of mandible. Anterior rhinoscopy showed a pale mass filling the left nasal cavity which was insensitive and did not bleed on touch. The septum was deviated to the right. There were no palpable neck nodes. Dystopia and reduced palpebral fissure was present due to the mass effect. Vision and eye movements were normal.

Computed Tomography scan of the paranasal sinus revealed a well defined expansile mass lesion measuring 6.6x4.3 cm in the left maxillary sinus. Anterior extension was to the premaxillary subcutaneous space, posteriorly to the pterygoid canal with minimal erosion of the pterygoid plate. Medially there was erosion of medial wall of maxillary sinus with extension into the left nasal cavity, pushing the septum to the right. Laterally, it extended into the masseteric space with partial erosion of root of zygomatic arch. Superiorly, there was erosion of floor of the orbit with intraorbital extension (Figure 2 and 3).

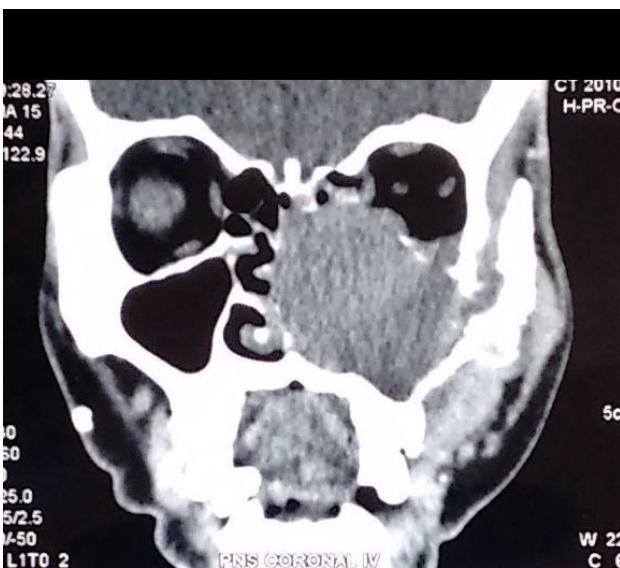
Histopathological study revealed cells with eccentric nucleus arranged in sheets and infiltrating the surrounding stroma. The diagnosis of plasmacytoma was

confirmed by immunohistochemistry (Figure 4). It was positive for CD138 and Kappa and negative for CK and CD 20. Bone marrow biopsy and aspiration was negative for bone involvement. The skeletal survey was normal. Urine analysis for bence jones proteins and serum myeloma protein was negative. Serum calcium and renal function was normal.

Patient received 25 fractions, 45 grays of radical radiotherapy over 4 weeks. A dramatic reduction in the size was seen within 1 week. There was complete disappearance of the tumor by the end of 4 weeks of radiotherapy (Figure 5). A follow up CT scan done for the patient after 6 months showed complete resolution of the tumour.



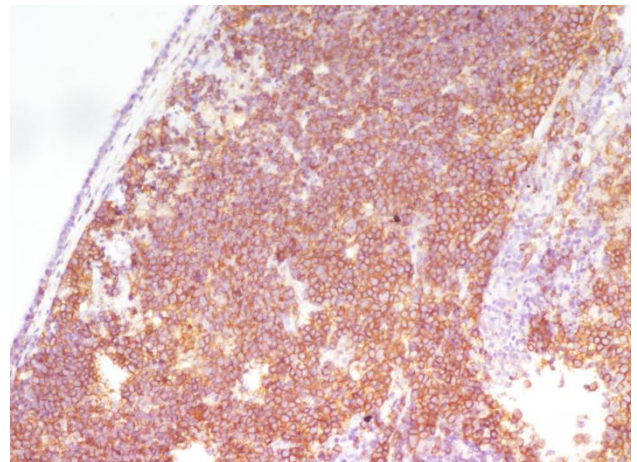
**Figure 1: Clinical photograph showing the swelling in left cheek.**



**Figure 2: Computed tomography-coronal view.**



**Figure 3: Computed tomography-axial view.**



**Figure 4: Immunohistochemistry slide.**



**Figure 5: Post treatment clinical photograph.**

## DISCUSSION

Extramedullary plasmacytoma represent less than 1% of head and neck malignancies.<sup>10</sup>

International myeloma working group diagnostic criteria of extramedullary plasmacytoma is as follows:<sup>11</sup>

- No M-protein in serum and/or urine
- Extramedullary tumour of clonal plasma cells
- Normal bone marrow
- Normal skeletal survey
- No related organ or tissue impairment

Approximately 80-90% of EMP involve the mucosa associated lymphoid tissue (MALT) of the upper airways. Of which, the nose and paranasal sinuses are involved in 75%.<sup>12</sup>

Plasmacytoma generally has an indolent course. Clinical symptoms are related to site of tumor development. Korolkowa et al reported that 40% occur in the nose and paranasal sinus, 20% in the nasopharynx, and 18% in the oropharynx. Multiple sites are involved in approximately 10% of extramedullary plasmacytomas.<sup>13</sup> The most common complaints in EMPs involving sinonasal region are nasal congestion, nasal discharge, and epistaxis.<sup>14</sup> Previous studies have reported the rare occurrence of EMP in the tonsils, minor salivary glands, posterior pharyngeal wall, thyroid gland, parathyroid gland, middle ear, colon, and liver.<sup>15</sup> Kapadia et al studied 20 patients with head and neck EMP and found that the most common complaint was the presence of a mass which was observed in 80% of the patients.<sup>16</sup> They found that airway obstruction was seen in 35% of the patients, epistaxis in 35%, localized pain in 20%, proptosis in 15%, regional lymphadenopathy in 10%, and abducens palsy in 5%.

Our patient presented with complaints of swelling in the cheek and epistaxis. Tumor was restricted to maxillary sinus and nasal cavity, with minimal intraorbital extension. There was no systemic involvement of the disease.

It is important to distinguish extramedullary plasmacytomas from non-neoplastic lesions like reactive plasmacytic hyperplasia, pseudolymphoma, plasma cell granuloma, and from malignancies like haematopoietic neoplasms, malignant melanoma, anaplastic carcinoma, olfactory neuroblastoma, and metastases. Immunohistochemical studies aid in the differentiation between plasmacytoma and polyclonal infiltrates of plasma cells.<sup>14</sup> EMP can be confirmed after excluding systemic disease by a serum and urine protein electrophoresis, immunoelectrophoresis, a skeletal survey and a marrow biopsy.

Previous studies and British Committee for standards in hematology recommend radical radiotherapy for

plasmacytoma encompassing the tumour volume with a margin of at least 2 cm and treating to a dose of 40 Gy in 20 fractions with a higher dose of 50 Gy in 25 fractions being considered for tumours >5 cm. The cervical nodes should be included if involved.<sup>17</sup>

The prognosis depends on tumour size and nodal involvement. The 10-years survival rate is 50–80%.<sup>18</sup>

## CONCLUSION

Extramedullary plasmacytoma is a rare head and neck malignancy. Immunohistochemistry aids in the diagnosis of EMP. It has a good prognosis with radiotherapy resulting in complete resolution.

*Funding: No funding sources*

*Conflict of interest: None declared*

*Ethical approval: Not required*

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**Cite this article as:** Kamath S, Pratap D, Shetty KC, Biniyam. A rare diagnosis of nasal mass: extramedullary plasmacytoma. *Int J Otorhinolaryngol Head Neck Surg* 2018;4:853-6.