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# Extramedullary plasmacytoma involving nasal cavity and orbit

*Eranga URR*[1], *Eleperuma DD*[2], *Kirihene KDRA*[3]

[1-]Registrar in Otorhinolaryngology, National Hospital Sri Lanka, Sri Lanka

[2-]Senior Registrar in Otorhinolaryngology, Teaching Hospital Karapitiya, Sri Lanka

[3-]Consultant ENT Surgeon, National Hospital Sri Lanka, Sri Lanka

## Abstract

### Introduction

Extramedullary plasmacytoma (EMP) is a disorder involving plasma cells, leading to the development of neoplastic masses outside the bone marrow. While EMP can occur in different body regions, its predominant manifestation is observed in the head and neck area.

A 44-year-old female presented with nasal obstruction, left-sided proptosis, and infraorbital swelling. Rigid nasal endoscopy revealed a polypoidal mass filling both nasal cavities, and imaging showed a uniform density mass lesion involving bilateral sinuses extending into the left orbit. The lesion was resected endoscopically, with subsequent histopathological examination confirming the diagnosis of EMP. Ancillary studies ruled out multiple myeloma (MM). The intraorbital component of the tumour was successfully treated using primary radiotherapy, leveraging the tumours' inherent radiosensitivity to minimize surgical morbidity.

### Conclusions

This case underscores the importance of including EMP in the differential diagnosis of sinonasal and other head and neck tumours. The favourable outcome achieved through a combination of surgery and radiotherapy highlights the significance of adopting a multidisciplinary approach in navigating challenging clinical scenarios, ensuring improved patient outcomes.

**Keywords**: Nasal Plasmacytoma, Extramedullary Plasmacytoma, Sinonasal Tumours



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**Correspondence**: Dr Udubokke Rathnayakage Rajith Eranga (raitheranga@med.ruh.ac.lk)

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## Introduction

Plasmacytomas are characterized by the abnormal proliferation of monoclonal plasma cells, presenting as either solitary osseous plasmacytoma or extramedullary plasmacytoma (EMP), with the latter accounting for less than 1% of head and neck malignancies[1]. Symptoms of nasal EMP include obstruction, epistaxis, rhinorrhea, facial swelling, and pain. EMP typically arises in the submucosa of the upper respiratory tract and has the potential to progress to multiple myeloma if not managed properly[2]. The rarity of nasal EMPs, coupled with the possibility of clinical resemblance to benign polyps, poses a diagnostic challenge for ENT surgeons

## Case Report

A 44-year-old female presented with a progressive nasal obstruction predominantly on the left side and swelling below the lower eyelid over a period of 6 months. She denied any history of epistaxis, epiphora, diplopia, anosmia, hyposmia, nasal discharge, headache or facial pain.

She had left proptosis, yet her visual acuity and colour vision remained normal, with no signs of ophthalmoplegia. Rigid nasal endoscopy (RNE) revealed bilateral hypertrophied turbinates filling the nasal cavity, without definitive mass lesions, purulent secretions or mucosal necrosis.

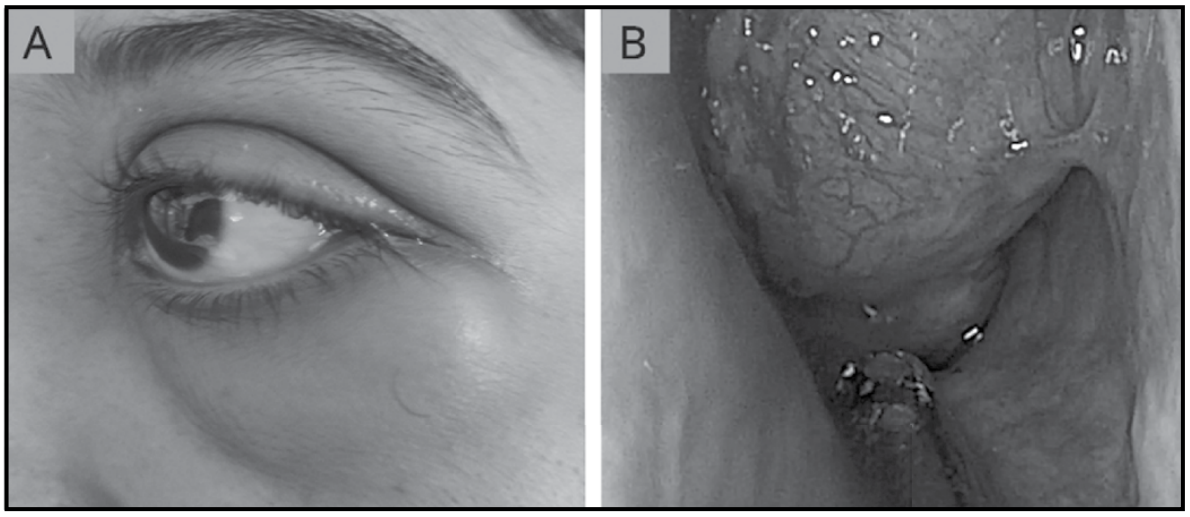


**Figure 1:** A. Infraorbital swelling and proptosis of left eye; B. Endoscopic appearance of left nasal cavity

Contrast-enhanced computed tomography (CECT) of the nose and paranasal sinuses revealed a uniform-density polypoidal mass involving the frontal, maxillary, ethmoid, and sphenoid sinuses. The lesion was found to have eroded the roof of the left maxillary sinus, extending into the extraconal compartment of the left orbit compressing the extraocular muscles. Ultrasonography (USS) showed no cervical lymphadenopathy.

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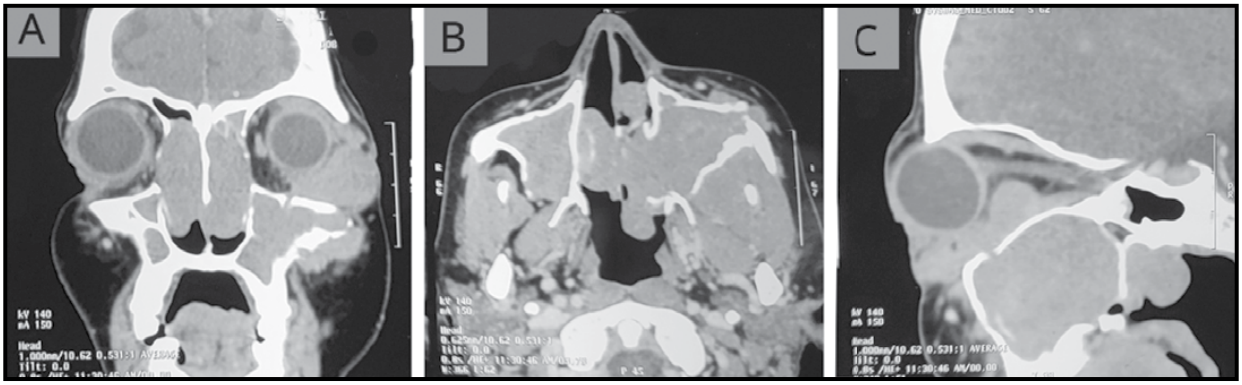


**Figure 2:** CECT of nose and paranasal sinuses showing nasal tumour; A. Coronal view B. Axial view C. Sagittal view

Considering the potential diagnoses of a malignant sinonasal tumour with orbital extension or invasive fungal sinusitis, the decision was made to proceed with endoscopic resection of the tumour on urgent basis. Multiple tissue samples were collected during the surgery for histopathology, special fungal staining, fungal culture, and tuberculosis PCR. Histological examination revealed an infiltrating tumour composed of sheets of plasmacytoid cells, including mature and immature plasma cells suggesting EMP in the nasal cavity. Direct microscopy did not reveal any fungal hyphae or granulomata, and tuberculosis studies yielded negative results.

Further ancillary studies were conducted to exclude multiple myeloma. The full blood count revealed normal RBC and WBC counts, with a blood picture showing normochromic normocytic red cells and no rouleaux formation. Negative results were obtained for urine Bence Jones protein (BJP). Imaging through whole-body low-dose CT showed no lytic lesions, and a bone marrow aspirate from the iliac spine indicated normal cellularity, red and white cell maturation, with only 2% plasma cells and 5% lymphocytes. These findings collectively supported the exclusion of multiple myeloma in the diagnostic evaluation.

Trephine biopsy demonstrated normal lamellar bone with normocellular marrow space, active trilinear haematopoiesis, and no evidence of granuloma formation, lymphoid aggregation, necrosis, or fibrosis. Immunohistochemistry revealed scattered CD138 positivity (6-8%), indicating no evidence of abnormal plasma cell proliferation, confirming the diagnosis of EMP without systemic marrow infiltration.

In the subsequent management of the patient, a collaborative effort among specialists in oncology, radiology, ophthalmology, and haematology was employed to address the infraorbital portion of a lesion left in situ during endoscopic surgery. It was decided to avoid surgical further intervention due to the high risk of surgical morbidity, considering the patient had good vision. Instead, the patient was referred for radiotherapy, given the general radiosensitivity of EMP, and had a positive outcome.

## Discussion

Extramedullary plasmacytoma in the nasal cavity is a rare, slow-growing tumour primarily located in the head and neck region, encompassing nasal and oral cavities, with its exact cause remains unknown[2,3]. EMP originates from mucosal plasma cells, and its histological grade correlate with an increased risk of progressing to multiple myeloma in two to three years3. While cross-sectional imaging is plays a crucial role in evaluation of tumour involvement and staging, the definitive diagnosis relies on histopathology and immunohistochemistry[3].

The cornerstone of treating plasmacytoma involves a combination of radiotherapy (RT) and surgery. Extensive surgery for head and neck EPM is often discouraged due to the high radiosensitivity of these tumors[3]. In cases involving high-risk regions, as outlined in this case report, primary radiotherapy (RT) is a viable option. However, the preferred approach typically involves surgery with adjuvant RT, as depending solely on RT heightens the risk of recurrence[3]. Adjuvant chemotherapy may be considered in cases of large or RT non-responsive tumours to delay the progression to multiple myeloma3. Due to the limited number of cases, prognostic factors for nasal EMP remain unclear, emphasizing the need for lifelong follow-up[3,4].

## Conclusion

Despite their rarity, comprehensive understanding of head and neck extramedullary plasmacytomas is essential for ENT surgeons. Considering the diagnostic and therapeutic challenges, adopting a case-based management strategy that combines surgery and radiotherapy, facilitated by a multidisciplinary team (MDT) approach, is crucial for improving patient outcomes.

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