Case Report
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Nasopharyngeal Solitary Extramedullary Plasmacytoma: A Rare Case and Review of Literature

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Abstract

Introduction:

Solitary extramedullary plasmacytoma (EMP) of the nasopharynx is a rare plasma cell neoplasm, representing a small subset of head and neck malignancies. Its nonspecific symptoms often lead to diagnostic delays.

Case Report:

A 50-year-old female with non-specific progressive nasal obstruction and intermittent epistaxis, underwent complete endoscopic surgical excision using coblation. Post operative histopathology revealed Nasopharyngeal Extramedullary Plasmacytoma. The patient then received adjuvant radiotherapy (Volumetric Modulated Arc Therapy - VMAT), leading to a successful outcome with no recurrence at the one-year follow-up.

Conclusion:

This case underscores the importance of clinico-histopathological evaluation and a comprehensive, multimodal treatment approach, with nasopharyngeal extramedullary plasmacytoma (EMP) considered as a differential diagnosis in nasopharyngeal masses. Given the potential for progression to multiple myeloma, long-term surveillance is crucial. Increased awareness of this rare entity can facilitate early diagnosis, timely intervention, and improved patient outcomes.

Keywords: Extramedullary plasmacytoma, Nasopharyngeal mass, Plasma cell neoplasm, Solitary plasmacytoma, Volumetric Modulated Arc Therapy (VMAT).

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Introduction

Solitary plasmacytoma is an extremely rare, well-defined mass of neoplastic monoclonal plasma cells.

Plasma cell neoplasms are diseases characterized by the abnormal proliferation of monoclonal plasma cells (B cells) that produce monoclonal immunoglobulins.

These neoplasms can present as either: (a) multiple lesions, known as multiple myeloma (MM), a disseminated disease associated with abnormal M protein and lytic bone lesions; or (b) a single lesion, classified as solitary plasmacytoma (SP), which may occur in the bone (osseous plasmacytoma) or soft tissue (extramedullary plasmacytoma- EMP). Extramedullary plasmacytomas (EMP) account for <5% of all plasma cell neoplasms, with over 80% occurring in the head and neck region. EMPs represent approximately <3% of all nasal cavity tumors (1), and primarily affects males in their 5^{th} to 7^{th} decade of life.

Due to non-specific symptoms, nasopharyngeal EMP can be misdiagnosed as hemangiomas, angiofibroma, or lymphomas.

Diagnosis requires a combination of clinical evaluation and histopathology, and immunohistochemistry.

Standard treatment involves surgery, radiotherapy, or both, depending on tumor characteristics and location.

This report contributes to the limited literature on nasopharyngeal EMPs emphasizing the significance of early recognition and optimal treatment selection.

Case Report

A 50-year-old female from Mahasamund, a district in Chhattisgarh, presented to the Department of Otorhinolaryngology at AIIMS Raipur with a 1.5-year history of bilateral nasal obstruction that was insidious in onset and was gradually progressive.

In the past two months, the patient experienced intermittent nasal bleeding. The bleeding was scanty and stopped spontaneously after each episode.

No additional allergic, systemic or constitutional symptoms (such as weight loss, night sweats, nerve palsies or fever) were reported. Nasal endoscopy revealed a globular, mobile, vascular, encapsulated mass filling nasal choanae (Figure 1) with no bleeding on touch.

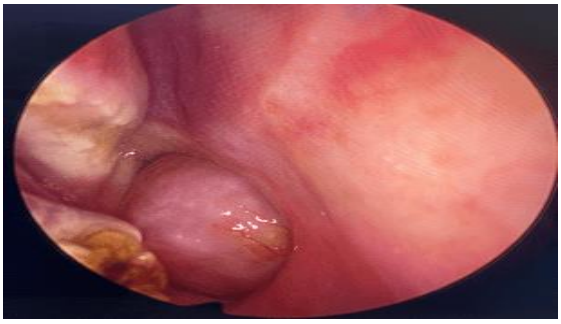

Fig. 1: Fleshy globular mass obstruction bilateral choanae

The examination of the ears, throat, and systemic systems was unremarkable, with no evidence of cervical lymphadenopathy, cheek swelling, nerve palsy, or paresis or any eye symptoms.

To further evaluate the condition, a *contrast enhanced computed tomography (CECT)* scan of the nose and paranasal sinuses was performed. It revealed a well demarcated, enhancing soft tissue mass measuring 2.6 × 1.9 × 1.8 centimeters, located on the posterior nasopharyngeal wall (Figure 2). The mass was abutting the soft palate, extending up to the C1-C2 vertebral level.

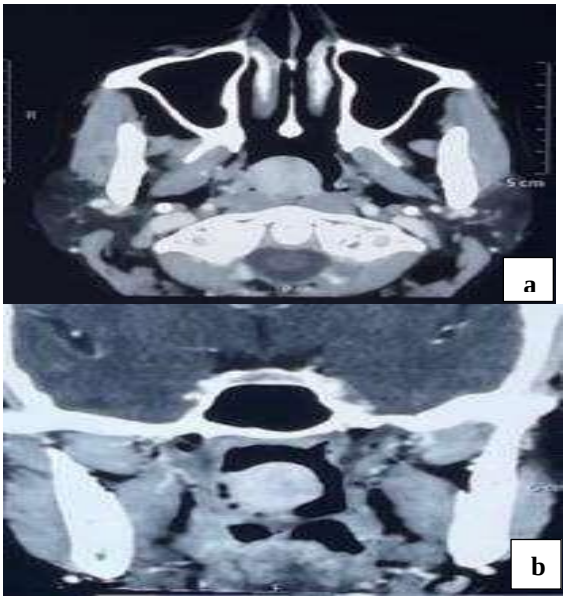

Fig. 2. Preoperative **(a)** axial and **(b)** coronal CECT showing well organized enhancing soft tissue mass in posterior wall of nasopharynx.

Further evaluation with diagnostic *digital subtraction angiography* (DSA) revealed a mild vascular blush, with blood supply to the lesion provided by the bilateral ascending pharyngeal arteries. Based on these findings,

the initial differential diagnosis included nasopharyngeal hemangioma, angiomatous polyp, and other neoplastic masses.

Surgical Management-

The patient underwent endoscopic surgical excision of nasopharyngeal mass under general anaesthesia using coblation. Intraoperatively, the lesion was found to have a pedunculated attachment on right lateral wall of nasopharynx behind posterior attachment of inferior turbinate. A reddish-grey globular mass was retrieved in toto from the oral cavity and sent for biopsy. Intraoperative bleeding was very minimal and postoperative period was uneventful.

Histopathological examination of excised tissue revealed dense sheets of atypical plasma cells (Figure 3) with high nuclear-cytoplasmic ratio, and very rare binucleated cells, including immature plasmablasts.

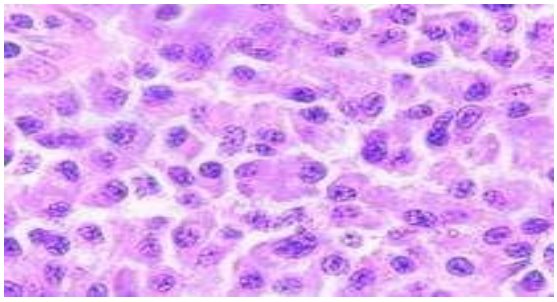

Fig. 3. Dense sheets of atypical plasma cells

Immunohistochemical analysis confirmed positivity for CD 138 (Figure 4), MUM-1, confirming plasma cell origin. Myogenin, Melan A, HMWCK, and chromogranin markers were negative, excluding other tumours. Hemangiomas and angiofibromas were excluded due to lack of vascular endothelial markers. These findings were consistent with a diagnosis of *Solitary Plasmacytoma*.

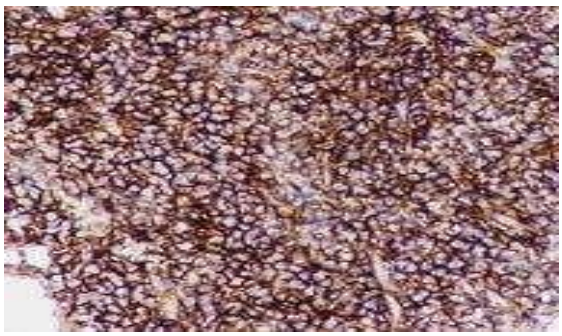

Fig. 4. Positive staining of CD 138 immunohisto-chemical marker

A comprehensive systemic evaluation was conducted to rule out multiple myeloma and other systemic plasma cell disorders. Ultrasonography of the whole abdomen showed no significant abnormalities. The assessment of beta-2 microglobulin levels, serum calcium, creatinine, and uric acid were within normal limits. A peripheral blood smear showed normal features. Serum and urine electrophoresis revealed no monoclonal gammopathy. A positron emission tomography PET scan was performed to identify possible osteolytic lesions (Figure. 5), along with bone marrow aspiration, which did not reveal any clonal plasma cell infiltration. Based on these findings, a final diagnosis of *Nasopharyngeal Solitary Extramedullary Plasmacytoma* was established.

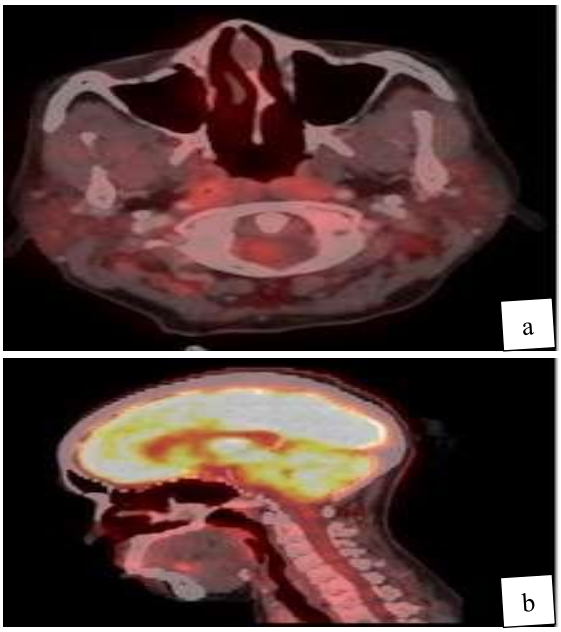

Fig. 5. Postoperative PET scan **(a)** axial cut, **(b)** coronal cut, showing no residual mass in nasopharynx without any other osteolytic lesion

Final diagnosis made as *Nasopharyngeal Solitary Extramedullary Plasmacytoma,* showing clonal plasma cells with no evidence of additional lesions or signs of systemic disease.

Adjuvant Therapy and Follow-Up

Following surgical excision, the patient underwent adjuvant radiotherapy, receiving a total of 28 cycles with cumulative dose of 50.4 Gray, delivered using Volumetric Modulated Arc Therapy (VMAT) technique to head and neck over a period of three months. No complications were observed post-radiotherapy.

At the 3-month follow-up, nasal endoscopy (Figure 6(a)) and computed tomography (Figure 6(b)) revealed no evidence of residual disease or recurrence. Subsequent endoscopy at 1-year follow-up (Figure 6(c)) showed a clear nasopharynx, indicating a favorable treatment response and a sustained remission with confirmed no signs of recurrence.

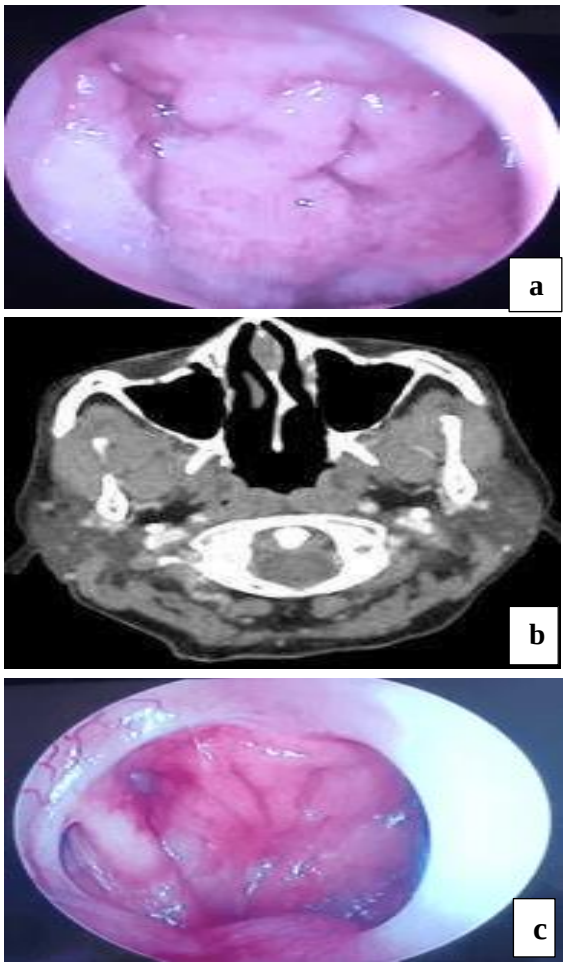

Fig. 6. Serial follow-up images demonstrating no residual or recurrent lesion in the nasopharynx. **(a)** Nasal endoscopy at 3 months post-treatment. **(b)** Axial cut of contrast-enhanced CT scan at 3 months. **(c)** Nasal endoscopy at 1-year follow-up confirming sustained remission

Discussion

Solitary extramedullary plasmacytoma (EMP) of the nasopharynx is a rare form of plasma cell neoplasm, accounting for less than 3% of all plasma cell tumors (2) and only 0.4% of head and neck malignancies (3). Approximately 80% of EMPs occur in head and neck region (2). EMPs predominantly affect males in the 5th to 7th decades of life, with a male-to-female ratio of approximately 3:1.

Although the exact pathogenesis remains unclear, chronic antigenic stimulation, environmental toxins, and viral infections such as Epstein-Barr virus (EBV) have been proposed as potential triggers. The most common presenting symptoms include progressive nasal obstruction, recurrent epistaxis, nasal discharge, and, in some cases, cervical lymphadenopathy. Due to its nonspecific presentation, EMP can be mistaken for other nasopharyngeal tumors, such as hemangiomas, angiofibroma, or lymphomas.

Wiltshaw's classification (3) is commonly used to stage extramedullary plasmacytomas:

- Stage I: Disease limited to a single extramedullary site
- Stage II: Regional lymph node involvement
- Stage III: Multiple metastases or systemic involvement

The accurate diagnosis of EMP requires a combination of imaging, histopathology, and immunohistochemistry. Microscopic examination typically reveals sheets of monoclonal plasma cells, while immunohistochemistry confirms positivity for markers such as CD138 and MUM-1. Given that approximately 15-30% of EMPs progress to multiple myeloma (3), systemic workup—including serum protein electrophoresis, urine Bence Jones protein analysis, and bone marrow biopsy-is essential to rule out disseminated disease. While radiotherapy (40-50 Gy) over four weeks remains gold standard (4) as these tumors are highly radiosensitive (1), our case used a multimodal approach (surgery + VMAT) to optimize local control and reduce recurrence. Surgical excision may be an alternative for well-circumscribed lesions, particularly in anatomical regions where RT may cause significant morbidity.

In our patient, the nasopharyngeal mass was well-circumscribed and allowed complete excision during the initial surgery, eliminating the need for a separate biopsy. The diagnosis of extramedullary plasmacytoma was confirmed postoperatively, following which systemic evaluation was performed and adjuvant VMAT radiotherapy was administered. This combined approach provided immediate mass removal while addressing potential microscopic residual disease, thereby optimizing local control and long-term outcomes.

The five-year survival rate ranges from 40–85%, with 7% of patients experiencing local

recurrence and 10–15% progressing to multiple myeloma. Long-term follow-up is essential, with evaluations every three months for two years, followed by every six months for the next three years and finally as annually for at least five more years. Several case reports have documented successful management of sinonasal EMPs using endoscopic resection followed by adjuvant radiotherapy. For instance, a 64-year-old male with nasal obstruction and epistaxis underwent endoscopic excision, with postoperative histopathology confirming EMP. Adjuvant radiotherapy was administered, and the patient remained disease-free on short-term follow-up (7). By Sarkar et al. (5), a small nasopharyngeal EMP treated with precisely targeted VMAT (45 Gy/25 fractions) achieved complete remission and remained relapse-free for 3 years with minimal toxicity (5). Another case treated by endoscopic excision followed by 45 Gy radiotherapy also demonstrated durable local control without recurrence on surveillance (6). Gupta et al. (3) and Grover et al. (2) emphasized radiotherapy as the primary treatment, whereas Hazarika et al. (1) explored surgical excision using KTP 532 laser. Unlike prior cases, our report highlights a multimodal approach combining surgery and radiotherapy, reinforcing its efficacy in preventing recurrence.

While Gupta et al. (3) and Grover et al. (2) reported predominantly male cases, our female patient represents a rarer demographic. This highlights the need for greater awareness and more extensive research into the epidemiology and behavior of this disease across different demographics. All studies confirmed monoclonal plasma cell proliferation; notably, Hazarika et al. (1) reported an HIV-positive case, suggesting immunological considerations. Our case utilized VMAT (Volumetric Modulated Arc Therapy) for radiotherapy, which offers better dose precision and tissue sparing—a technical advancement absent in prior studies. Unlike Sarkar et al. (2022) (4) and Serin et al (2020) (5), where radiotherapy alone was used, our case involved a well-circumscribed, accessible lesion, making surgical excision feasible. Surgery allowed immediate mass removal, reducing tumor burden and enabling better radiation targeting, minimizing toxicity to surrounding structures. Grover et al. (2) discussed socioeconomic barriers to radiotherapy, justifying surgery as a standalone treatment in resource-limited settings whereas our patient received adjuvant radiotherapy, emphasizing a comprehensive approach in a well-equipped center. Hazarika et al. (1) also reported a patient discontinuing radiotherapy due to mucositis, emphasizing treatment limitations in real-world settings.

Conclusion

This case underscores the efficacy of a multimodal treatment approach managing nasopharyngeal extramedullary plasmacytoma (EMP). The combination of surgical excision and advanced radiotherapy techniques like Volumetric Modulated Arc Therapy (VMAT) led to excellent local control with no recurrence at one year.

Given the potential for progression to multiple myeloma (10–30% of cases), long-term monitoring remains crucial.

Further research is needed to clarify genetic, immunological, molecular predictors and gender-specific factors influencing EMP behavior. Ultimately, this case underscores that even rare conditions can significantly impact patients' lives. Rare malignancies, when diagnosed early and managed appropriately, can have favorable outcomes. Maintaining a high index of suspicion and ensuring timely intervention are essential to improving outcomes in this rare but treatable malignancy.

Declaration

This case report did not require ethics approval as per institutional policy for single case reports. Consent for Publication: Written informed consent was obtained from the patient for publication of this case report and accompanying images.

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