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Case Report

# Multiple Nasal Swellings and An Unusual Diagnosis: Extramedullary Plasmacytoma

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## Abstract

Plasmacytomas are lymphoproliferative disorders characterized by monoclonal proliferation of plasma cells. Of these extramedullary plasmacytomas are rare tumors. They are mostly seen in the submucosa of the upper aerodigestive tract. Clinical manifestations vary depending on the location of the lesions. In the nasal cavity, it can cause nasal obstruction, anosmia/hyposmia, headache and epistaxis. The risk of conversion to multiple myeloma is 15%-20%. To rule out multiple myeloma, a complete systemic evaluation should be done in all patients with extramedullary plasmacytoma. Excision biopsy/IHC play an important role in differentiating and planning the management of these disorders. It is a highly radiosensitive tumor, so radiotherapy is the treatment of choice. We report a case of a 38 years old female presented to ENT department with nose block and diagnosed as extramedullary plasmacytoma without any systemic manifestation and no evidence of multiple myeloma, treated with surgery as sole modality of treatment and no recurrences on follow up to date, which makes this case different from others.

Even though radiotherapy is the treatment of choice, it can be treated with surgery alone if the lesion is small in size. Keeping the recurrence rates in mind, regular follow ups are required for early detection and treatment.

**Keywords:** Extramedullary Plasmacytoma; Multiple Myeloma; Immunohistochemistry; Serum Protein Electrophoresis; Serum Immunotyping

## Abbreviation

EMP: Extramedullary Plasmacytoma; MM: Multiple Myeloma; CT: Computed Tomography; RT: Radiotherapy; IHC: Immunohistochemistry; Ig G: Immunoglobulin G; PET: Positron Emission Tomography; IT: Inferior Turbinate;

## Introduction

Plasmacytomas are lymphoproliferative disorders characterized by monoclonal proliferation of plasma cells. Two subtypes of plasmacytoma are solitary bone plasmacytoma and extramedullary plasmacytoma [1]. Extramedullary plasmacytomas are rare tumors. Only less than 10% of all plasmacytic tumors and less than 1% of head and neck tumors are accounted for extramedullary plasmacytomas [2]. Extramedullary plasmacytomas most commonly seen in the submucosa of the upper aerodigestive tract. The risk of conversion to multiple myeloma is 15%-20% [3].

Approximately 3 of 100000 occur annually [4]. Plasmacytomas are 3 times common in males compared to females. These tumors usually occur in the fourth to fifth decade of life [5].

The exact etiology of extramedullary plasmacytoma is still unknown. Chronic stimulation, irritant inhalation, viral infection, radiation overdose or genetic disorders affecting the reticuloendothelial system play a role [5,6]. Clinical manifestations vary depending on the location of the lesions. In the nasal cavity, it can

cause nasal obstruction, anosmia/hyposmia, headache and epistaxis [5,6]. In the orbit, it can cause diplopia, ptosis, blurring of vision or impaired extraocular muscle movements. In the oropharynx, it can present as dysphagia and snoring. In the larynx, it can cause dysphonia, dyspnea and if it is quite large enough, might end up in stridor as well. In the gastrointestinal tract, it can cause epigastric pain, hematemesis, loss of appetite and chronic abdominal pain. These lesions present as friable, polypoidal swellings. It may vary in size from very small to big enough to cause complete airway obstruction [7]. Consistency of these are usually soft to firm, and they are non-tender to touch. To rule out multiple myeloma, a complete systemic evaluation should be done in all patients with EMP, which includes protein electrophoresis for detection of immunoglobulins in blood and urine, renal and hepatic function tests, biopsy of bone marrow and complete skeletal study with CT or PET scan [6]. Hypercalcaemia and anemia should also be ruled out [8]. If all of this is normal, we can rule out multiple myeloma and conclude this as a case of solitary plasmacytoma.

Differential diagnosis of plasmacytoma in nasal cavity are sarcoma, squamous cell cancer, meningioma, haemangioma, neurofibroma, esthesioneuroblastoma and lymphoma [9].

It is a highly radiosensitive tumor, so radiotherapy can be used as a sole modality of treatment, however surgery is essential for histopathological diagnosis [3,6]. Strict follow-ups with peripheral smear, protein electrophoresis, endoscopic examination and imaging are required for these patients since EMP has a risk of recurrence of 22%, risk of conversion into multiple myeloma of 15%-20% and the risk of distance metastasis of 35-50% [3], which can occur several years after the initial diagnosis. With all of this, global survival is 50- 70% at 10 years [8,10].

We report a case of a 38 years old female who presented to ENT department with nose block and was diagnosed as extramedullary plasmacytoma without any systemic manifestation or multiple myeloma, treated with surgery as sole modality of treatment and no recurrences on follow up to date.

## Case Report

A 38 years old lady with no known co morbidities presented to our tertiary care hospital with history of progressive right sided nasal block since 1 month. There was no history of nasal discharge, nasal bleed, or allergic symptoms. Diagnostic nasal endoscopy done showed multiple smooth surfaced polypoidal lesions in bilateral nasal cavities. In the right nasal cavity, lesion was seen originating from the floor of the nose anteriorly (Figure 1), right middle turbinate laterally, posterior end of inferior turbinate medially, septal wall at the level of middle turbinate. In the left nasal cavity, lesions of varying sizes were seen in the floor of the nose anteriorly (Figure 2), posterior end of inferior turbinate medially and on the inferior aspect (Figure 3). Lesions were firm in consistency, non-tender and not bleeding on touch.

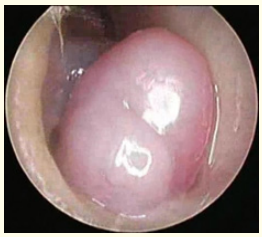

**Figure 1:** Smooth surfaced lesion in right nasal cavity floor.

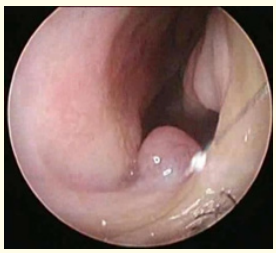

**Figure 2:** Small smooth surfaced lesion left nasal cavity floor.

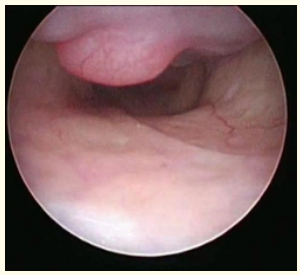

**Figure 3:** Lesion in the posterior end of left inferior turbinate inferior aspect

For further evaluation, CT scan Nose and PNS was done which showed a 1 x 1 cm, well-defined, mildly hyperdense, soft tissue density lesion in the anterior inferior aspect of the right nasal cavity (Figure 4 and Figure 5). This lesion was inseparable from the anterior portion of nasal septum medially and also the inferior turbinate laterally. Similar smaller lesion also seen in the inferior aspect of left nasal cavity (Figure 6).

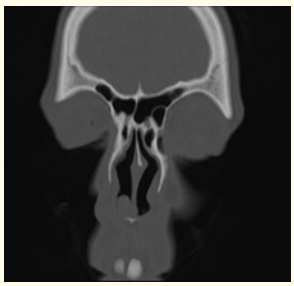

**Figure 4:** Lesion in the posterior end of left inferior turbinate inferior aspect.

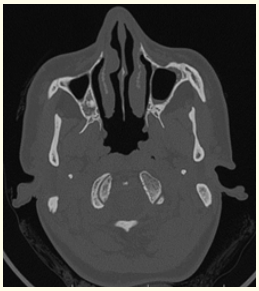

**Figure 5:** Soft tissue density lesion in the lateral aspect of the right nasal cavity inseparable from the IT.

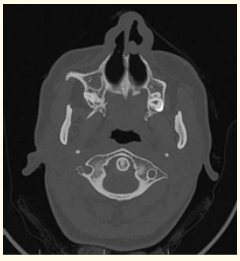

**Figure 6:** Lesion seen in the inferior aspect of left nasal cavity.

She underwent excision of the lesions via an endoscopic approach under general anaesthesia, all lesions were excised completely and sent for histopathological assessment. Post operatively she was relieved of nasal obstruction.

Histological sections examined showed polypoidal fragments of sheets of plasma cells including binucleated forms with overlying respiratory epithelium suggestive of plasmacytoma (Figure 7).

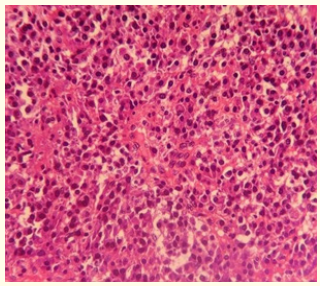

**Figure 7:** Section shows sheets of plasma cells (HE, x400).

Immunohistochemistry was done for further evaluation, which turned out to be positive for CD 138 and MUM-1. The cells showed lambda light chain restriction. The tumor cells did not express cytokeratin & CD 20.

In view of probable association of EMP and multiple myeloma, she was referred to the hematologist for further evaluation. Further hematological evaluation was performed to rule out multiple myeloma (MM). Peripheral smear showed mild Microcytic hypochromic anemia. Quantitative immunoglobulins, full blood count, urea, calcium, creatinine, and β2 microglobulin were all within normal ranges. Serum protein electrophoresis (Figure 8) and serum immunotyping (Figure 9) were normal and level of lambda and kappa light chain were normal. After ruling out MM, the patient was diagnosed as having EMP of the nasal cavity, which is of unknown origin. She is on regular follow-up with peripheral smear and diagnostic nasal endoscopy and there is no evidence of recurrence after 3 years following surgery and she is completely symptom free. Figure 10 shows nasal endoscopy finding 3 years post surgery.

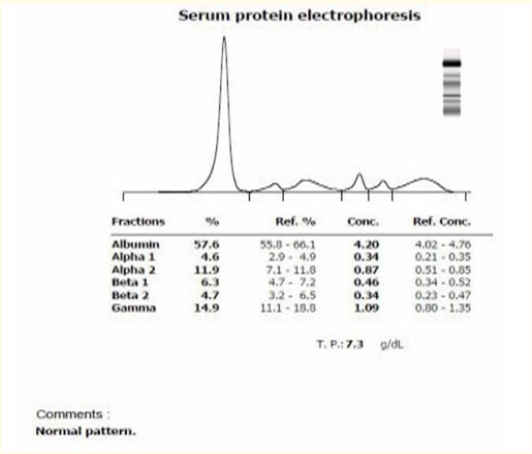


**Figure 8:** Serum electrophoresis showing normal pattern.

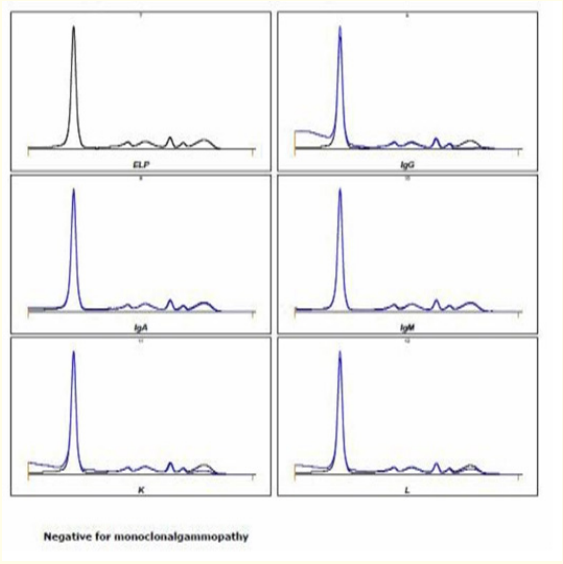


**Figure 9:** Serum immunotyping shows no evidence of monoclonal gammopathy.

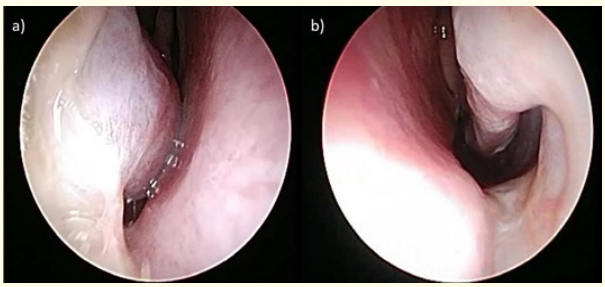


**Figure 10:** Post-operative image on 3 year follow up (a) right nasal cavity (b) left nasal cavity.

## Discussion

Extramedullary plasmacytomas originate from monoclonal proliferation of plasma cells [1,2]. These tumors are usually present submucosally, usually in the upper aerodigestive tract, that is the nasal cavity and paranasal sinuses, oropharynx, and followed by colon. This patient had extramedullary plasmacytomas that occurred in the multiple areas in the nasal cavity. EMP is 3 times more common in males compared to females, but our case was a female in her late 30s.

Symptoms of nasal EMP are usually non-specific, can include pain, nasal obstruction and epistaxis, which was similar to our patient's presentation. On examination, the lesion appears pink polypoidal friable mass, which is non tender.

The CT findings of EMP are non-specific; it is seen as solid growth with variable enhancement, with adjacent bone destruction, infiltration into adjacent soft tissues [11]. CT is important in determining the location, extent of lesion, and in determining adjacent bone and soft tissue involvement. Hu X., *et al*. reported a case of nasal EMP which showed bone resorption in the medial wall of the maxillary sinus [12]. Our case presented as multiple small lesions without bone and adjacent soft tissue involvement.

On gross examination, friable, polypoid soft tissue, with central necrotic zones can be seen.

The diagnosis of EMP can be concluded only with pathology and immunohistochemistry findings [12].

Histologically, it consists of a monoclonal proliferation of plasma cells arranged in sheets and clusters, as can see on HPE in our case. Sometimes, it can produce immunoglobulin, frequently IgG, which was absent in our case. 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 [10]. All the systemic work ups were normal in our patient so that we concluded this as a case of nasal extramedullary plasmacytoma.

The treatment of EMP is surgical excision, radiation, or a combination of both. Radiotherapy is the treatment of choice. Surgery

alone is indicated if the tumor is small. For larger tumors, both surgery and radiotherapy are required for a better prognosis. The 10-year survival rate is approximately 50-70% [8,10]. Chemotherapy is indicated only when there are multiple lesions that occur in areas other than the primary sites, therefore it was not required in our case.

Windfuhr., *et al*. reported a case of nasal EMP which was initially treated with radiotherapy to which the patient did not respond and later treated surgically [10]. Ashish G., *et al*. reported a case of sinonasal EMP involving orbit, in which they treated the patient with radiotherapy (40 Gy in 20 fractions) and at 2 years post radiotherapy patient was completely disease free [5]. Shreif. J. A., *et al*. reported a case of nasal EMP, in which the lesion was completely removed surgically and given post-op radiotherapy (4400 cGy over a 1-month period), patient was symptom free on 1 year follow up [14]. Merzouqi., *et al*. reported a case of EMP soft palate extending to nose treated solely with chemotherapy as he refused surgical management, he was completely disease free on 1 year follow up [15]. Ashraf., *et al*. reported 3 cases of EMP in nasal cavity in which one case is treated solely with surgery while other 2 cases with surgery and postop RT. All patients are completely disease free after treatment [8]. Hu H., *et al*. reported a case which was treated with surgery alone and 5 months post operatively, patient doesn't have any recurrence or conversion to myeloma [16].

Pinto JA., *et al*. reported a case of a solitary EMP larynx which was excised with a CO2 laser and no radiotherapy given [17]. Patient didn't develop MM nor had recurrence post treatment. Aslan I., *et al*. reported a case of EMP thyroid cartilage in which they removed the lesion through an external approach, and since it was locally destructed the cartilage, post op RT was given to the patient [18]. Nakashima., *et al*. reported two cases of EMP larynx, of which one was over the left arytenoid which was treated with excision and radiotherapy and the second case was an epiglottic EMP treated with surgical resection alone and patient was disease free 15 years post operatively [19]. Bazaadut S., *et al*. reported a case of EMP of palatine tonsil with level II lymph node involvement which was treated with surgical excision alone and patient remained disease free 6 months follow up [20]. Lorusso GD., *et al*. reported a case of nasopharyngeal EMP treated with surgery alone [21]. Our case was managed with surgical resection and patient was serially followed up, which showed no recurrence even after 3 years post surgery. If lesion is small, surgery alone is indicated as treatment. For larger tumors, both surgery and radiotherapy are required for a greater chance of survival [4].

Nearly 50% of extramedullary occur as a solitary mass, whereas 20% present in multiple areas. Solitary EMP has better prognosis than lesions in multiple areas [4].

## Conclusion

In a patient presenting with multiple lesions in the nasal cavity, it is important to keep in mind that there may be a manifestation of lympho-proliferative disorder like plasmacytoma. Excision biopsy/IHC play an important role in differentiating and planning the management of these disorders. Even though radiotherapy is the treatment of choice, it can be treated with surgery alone if the lesion is small in size. Keeping the recurrence rates in mind, regular follow ups are required for early detection and treatment.

## Conflict of Interest

None.

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