

# A Rare Incidence of Solitary Extramedullary Plasmacytoma of Nasal Cavity

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## Abstract

Extramedullary plasmacytoma (EMP) is a rare neoplasm characterized by monoclonal proliferation of plasma cells. They originate in either bone – solitary osseous plasmacytoma, or in soft tissue – EMP. EMP represents <1% of all head and neck malignancies. The nasal cavity and nasal septum are the common sites of occurrence. This is a report of EMP in a 47-year-old gentleman. He presented with a 3-month history of the left nasal blockage and epistaxis. Examination revealed a large reddish to blackish mass of the left nasal cavity. The first biopsy, however, showed benign sinonasal polyps. Biopsy was repeated in view of suspicious clinical and computed tomography features. Histopathology from the second biopsy reported as EMP, confirmed by immunohistochemical techniques. The patient underwent bone marrow aspirate and trephine, skeletal survey, and laboratory investigations was performed to exclude multiple myeloma. Radiotherapy was initiated.

**Key words:** Benign sinonasal polyp, Extramedullary plasmacytoma, Multiple myeloma

## INTRODUCTION

Extramedullary plasmacytoma (EMP) is an uncommon plasma cell neoplasm which occurs in soft tissue. It shows a preference for the upper respiratory tract, especially the nasal cavity and paranasal sinus (PNS). It corresponds to <10% of all plasmacytic tumors,<sup>[1]</sup> representing <1% of all head and neck tumors<sup>[2]</sup> and <0.5% of tumors of the aerodigestive tract.<sup>[3]</sup> It typically arises in submucosal soft tissues of the upper respiratory tract and it is destructive with a tendency to local recurrence.

The clinical symptoms: Tumor or local edema in 80%, nasal obstruction in 35%, epistaxis in 35%, localized pain in 20%, proptosis in 15%, rhinorrhea in 10%, regional lymphadenopathy in 10%, and paralysis of the VI cranial nerve in 5% of cases.<sup>[2,4]</sup> To exclude multiple myeloma (MM) or plasmacytoma of the bone, a systemic work-up and follow-up of the patient are mandatory, including serum protein electrophoresis, urinalysis for the Bence-Jones protein, skeletal survey, and bone marrow biopsy.

The treatment of choice for EP is surgery and radiation therapy (RT) with a dose of 40–50 Gy over a 4-week period, the disease is highly radiosensitive.<sup>[1,5]</sup>

Overall, most studies report high local control rates of approximately 80–100% with moderate doses.<sup>[6]</sup>

## CASE REPORT

A 42-year-old male underlying schizophrenia, presented with epistaxis over the left nostril for 3 months associated with nasal blockage. On physical examination, cranial nerve was intact, there was no external deformity. Nasoendoscope showed a dark reddish, well-lobulated mass occupying whole left nasal cavity [Figure 1], unable to visualize the origin of the mass. The right nostril was clear and bilateral for no mass seen. There were no neck nodes palpable. Biopsy was taken and reported as benign sinonasal polyp. Computed tomography (CT) PNS done and showed enhancing mass arising from the left nasal cavity measuring (4.7 × 3.6 × 4.8cm). Superiorly, the mass extended into the left anterior ethmoidal cells and frontal recess, left orbital wall intact, laterally into left maxillary ostium, and obliterates left ostiomeatal complex. Medially, it caused septal deviation to the right, inferiorly, until the level hard palate, with inferoposterior margin of the mass compresses onto the left torus tubarius. Anteriorly, the mass protruded into the left nostrils. Posteriorly, the mass extends to the posterior left ethmoidal air cells, obliterates

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the sphenoidal recess [Figures 2 and 3]. Biopsy was repeated under geometric algebra due to suspicious mass and reported as EMP as CD 138 positive cells with kappa light chain restriction. The patient was referred to medical team and a systemic work-up to exclude MM performed.

Renal profile was normal, blood profile shows no evidence of bone marrow infiltration by malignancy or plasma cell myeloma. Serum and urine protein electrophoresis shows IgA kappa paraproteinemia of 5.7 g/dL in the beta zone with no immunoparesis. Serum and urine free light chain showed no light chain detected. Bone marrow needle biopsy and skeletal survey were negative. CTNTAP showed no evidence of lesion/mass in the thorax or abdomen. Thus, a diagnosis of extramedullary nasal plasmacytoma was made.

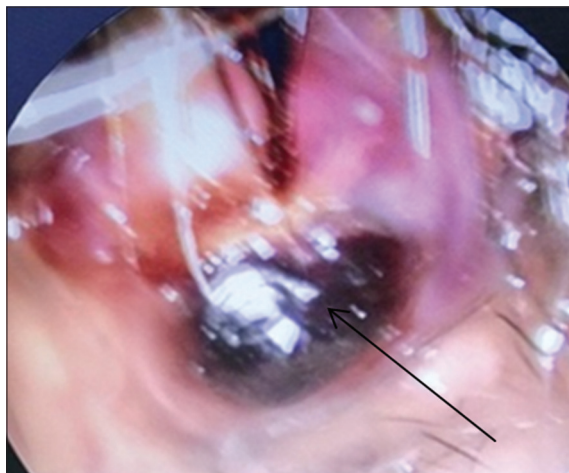
The patient received radiotherapy with a radiation dose of 40 Gy in 15 fractions. The tumor reduces in size post-radiotherapy and currently under surveillance follow-up

with nasoendoscope and CT scan. At present, 2-month post-radiotherapy, nasoendoscope showed the nasal mass remains the same size [Figure 4].

**DISCUSSION**

EMP is a rare neoplasm characterized by monoclonal proliferation of plasma cells outside bone marrow. It is defined as a soft tissue plasma cell tumor occurring in the absence of systemic signs of MM, such as bone osteolytic lesions, plasma cell infiltration in bone marrow, lytic bone lesion, or serum or urine myeloma protein (M-component). The most frequently affected areas in the upper aerodigestive tract are the nasal cavity or PNS (43.8%), followed by nasopharynx (18.3%), oropharynx (17.8%), and larynx (11.1%) due to its rich lymphatic tissue.<sup>[7]</sup>

Due to its presentation in the submucosa of the aerodigestive tracts, it is suggested that the etiology of



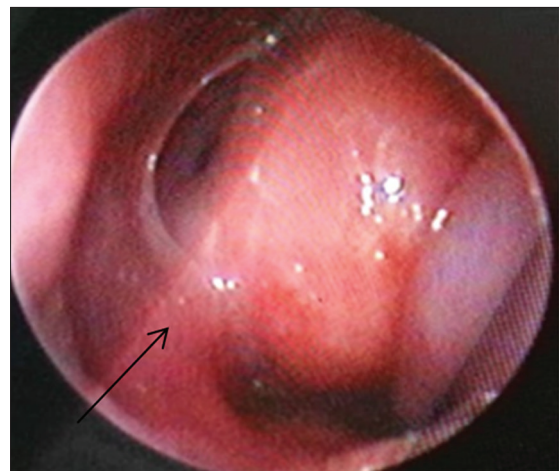
**Figure 1: Nasoscopy shows the left anterior nasal occupied with mass in red color – black arrow (before radiotherapy)**



**Figure 2: Computed tomography paranasal sinus shows homogenous enhancing mass (black arrow) arising from the left nasal cavity extends into the left maxillary ostium, anterior ethmoidal air cells, and frontal recess**



**Figure 2: Computed tomography scan axial view showing homogenous mass (black arrow) filling up the left nasal cavity. Orbital wall intact**



**Figure 4: Nasoscopy shows the left nasal mass confined over the inferior turbinate – black arrow (post-radiotherapy)**

EMP may be related to chronic stimulation caused by inhaled irritants or viral infection.<sup>[4]</sup> The most common clinical findings are as follows: Blocked nose, soft tissue mass (fleshy, yellowish-grey to dark red sessile, polypoid, or pedunculated), epistaxis, nasal discharge, pain, more rarely cranial nerve palsy, and neck lymphadenopathy.

The diagnosis of EMP depends on clinical suspicion such as unilateral nasal mass and biopsy to exclude other nasal tract malignancy. The differential diagnosis includes other nasal tract malignancies such as inverted papilloma, pleomorphic adenoma, squamous cell carcinoma, adenocarcinoma, adenoid cystic carcinoma, melanoma, esthesioneuroblastoma, rhabdomyosarcoma, lymphoma, sinonasal undifferentiated carcinoma, and Wegener granulomatosis.

In this case, the initial biopsy shows that inflammatory polyps could be due to superficial punch biopsy. Deeper biopsy under general anesthesia subsequently showed EMP. Deep biopsy should be performed because the tumor is submucosal and the mucosal lining can become thicker due to an inflammatory reaction. In cases, where clinical appearance is suspicious, a repeated biopsy is required.

A study using immunohistochemical methods confirmed the plasmatic nature of the cells with cell markers, for example, CD 138, which indicates the necessity of performing an evaluation for a differential diagnosis from other cancers such as melanoma, undifferentiated carcinoma, and pituitary adenoma.<sup>[8]</sup>

In all case of EMP, a systemic work-up including blood profile, renal and liver function, serum and urinary protein electrophoresis, serum immunoglobulin level, skeletal survey, and bone marrow examination must be performed to exclude a systemic disease such as MM.

RT is the treatment of choice in EMP localized in the head and neck, not extending through the floor of the anterior and middle cranial fossae and into the orbit. The British Society for Hematology recommends initial radiation treatment with 40 Gy in 20 fractions with a 2 cm margin for tumors smaller than 5 cm and 50 Gy in 25 fractions for larger tumors, whereas the role of surgery is usually limited to biopsy and to excision of residual disease.<sup>[1,5]</sup> The prognosis for patients with EMP is 5-year survival rates between 30 and 82% and 10-year rates 50–90%.

Chemotherapy is considered only in patients with tumors larger than 5 cm, high-grade tumors, refractory and/or relapsed disease, and in case of progression to MM.<sup>[5,9]</sup>

The rate of conversion of EMP to MM is lower than other plasma cell neoplasms, such as SPB, with rates reported to be

between 11 and 33% over 10 years.<sup>[9]</sup> Hence, it is recommended that patients receive regular follow-up after diagnosis of EMP due to the relatively high risk of conversion.

Myeloma guidelines by the Italian Association of Medical Oncology (AIOM)<sup>[10]</sup> suggest the first screening 45–60 days after radiotherapy by serum examinations, then every 3 months for the 1<sup>st</sup> year, subsequently, every 6 months by serum, radiological, and bone marrow examinations, if necessary. Due to the high risk of conversion, D'Aguillo *et al.*<sup>[9]</sup> proposed a regular screening for MM every 6 weeks for the first 6 months after diagnosis of EMP and then periodically, but without a specific timing. For these reasons, we propose a follow-up protocol consisting of nasal endoscopy and serum examinations every 3 months, and imaging study with magnetic resonance imaging (MRI) 3 months after radiotherapy and subsequently every 6 months/year for 5 years; after 5 years, we propose serum examinations and nasal endoscopy every 6 months and MRI every year. We recommend a biopsy only in cases of clinical and instrumental suspicion of recurrence.

In our patient, the location of tumor involving only the left anterior ethmoidal air cells and frontal recess. The left orbital wall is still intact with no tumor extension intraorbitally led us to performed only radiotherapy which the size of tumor significantly reduced. Currently patient under 3 monthly Otorhinolayngology follow up post radiotherapy and nasoendoscopy shows the mass significantly reduce in size as [Figure 4] with further systemic workout shows no multiple myeloma.

## CONCLUSION

EMP is a rare, aggressive tumor that mainly affects the submucosa of the nasal cavity and PNS. This tumor can remain in the area of the early lesion, advance to neighboring areas, or even spread. The otorhinolaryngologist must identify the lesion and refer the patient for hematologic monitoring; moreover, a multidisciplinary approach is required to differentiate between localized disease and blood dyscrasias with a poor prognosis, such as MM. Treatment with radiotherapy is effective because the tumor is radiosensitive, and surgery may occasionally be used to complement the treatment. Controlled clinical trials are needed to establish a definitive treatment of choice for the management of these patients. The patient should always be monitored for a long period of time.

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