



## Case report

## Successful endoscopic excision of nasal plasmacytoma: Lesson learnt

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## ARTICLE INFO

## Article history

Received 21 January 2016

Accepted 10 May 2017

Available online 29 June 2018

## Keywords

Extramedullary plasmacytoma

Sinonasal tumours

Multiple myeloma

## Doi

10.29089/2017.17.00060

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## ABSTRACT

**Introduction:** Extramedullary plasmacytoma is a rare entity which belongs to non-Hodgkin lymphoma category. Head and neck region is the most common site of manifestation, of which sinonasal area is of predominance which oftentimes remains undiagnosed.

**Aim:** To illustrate unusual presentation of nasal mass and its management.

**Case study:** Herein, we are reporting a case of sinonasal plasmacytoma in an elderly man who presented with a four month history of unilateral nasal blockage. Rigid nasoendoscopy revealed benign looking polypoidal mass occupying entire right nasal cavity with no evidence of any polypoidal mass seen over the left nostril. Computer tomography revealed homogenous soft tissue mass over right nasal cavity with minimal mucosal thickening in bilateral maxillary, ethmoid, sphenoid and left frontal sinus.

**Results and discussion:** Patient underwent endoscopic clearance of right nasal mass under general anaesthesia. Nasal mass was removed with the aid of microdebrider and Blakesley forcep without difficulty. Intra- and postoperative were unremarkable. Histopathological examination of the nasal mass along with immunohistochemical study was suggestive of nasal plasmacytoma. In addition, systematic workup for multiple myeloma was negative. Subsequent follow-up revealed no evidence or recurrence and patient has been asymptomatic till date.

**Conclusions:** Transnasal endoscopic surgical excision is an ideal treatment for small and localised extramedullary sinonasal plasmacytoma. Albeit rare, extramedullary sinonasal plasmacytoma should be considered as a differential diagnosis of sinonasal mass as this entity has tendency to develop into multiple myeloma as well as the different mode of management of this rare entity differs.

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## 1. INTRODUCTION

Extramedullary plasmacytoma (EMP) is a rare plasma cell neoplasm involving soft tissue, accounts for less than 1% of all head and neck tumours<sup>1</sup> and less than 0.5% of all aerodigestive tract tumours.<sup>2</sup> It commonly manifests in the upper respiratory tract, more so in the nasal cavity and paranasal sinus.<sup>1</sup> Approximately 80%–90% of EMP involve mucosa-associated lymphoid tissue of the upper airway, of which 75% involves sinonasal region.<sup>1</sup> In up to 33% of cases, multiple myeloma may occur, thus this warrants a life-long follow up of patients with sinonasal EMP. In this article, we describe the presentation, diagnosis and management of this entity.

## 2. AIM

Aim of this case report is to illustrate on unusual presentation of nasal mass, importance of histopathological examination for all nasal mass despite being benign looking and its management.

## 3. CASE STUDY

A 79-year-old Indian gentleman with underlying hypertension, was referred to Madras ENT Research Foundation, with a four-month history of right-sided nasal blockage. According to him, nasal blockage lead to difficulty in breathing which caused distress to him. Apart from that, there were neither any accompanying epistaxis, nasal discharge, facial tenderness, proptosis nor any inciting trauma or infection prior to that. There were also no prior symptoms suggestive of an allergic condition or any upper respiratory infections or any prior nasal surgery.

Upon review, patient was comfortable with no sign of respiratory distress. Anterior rhinoscopy revealed right-sided benign looking nasal mass with congested mucosa over right nasal cavity with no mass seen over left nasal cavity. Zero degree rigid nasoendoscopy revealed a benign looking polypoidal mass occupying entire right nasal cavity with no sign of an active infection which did not bleed upon probing. There was however, no evidence of any polypoidal mass seen over the left nostril. Other than that, intraoral examination was normal. His systemic examination and vital signs were also unremarkable and within normal range. Blood parameters were also normal.

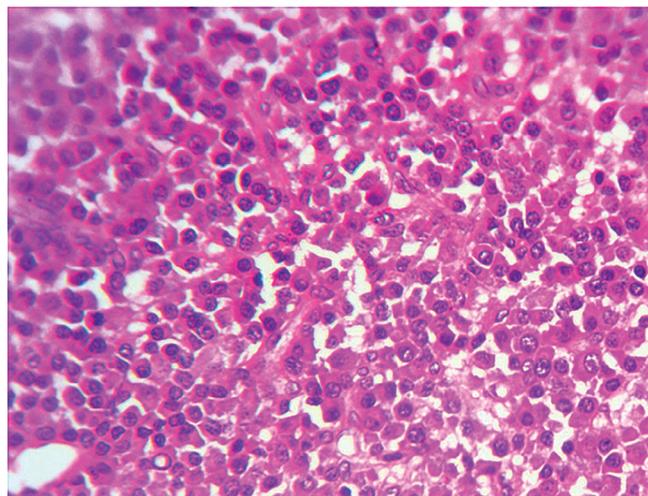
Computer tomography revealed homogenous soft tissue mass occupying right nasal cavity with no evidence of erosion, minimal mucosal thickening in bilateral maxillary, ethmoid, sphenoid and left frontal sinus (Figure 1). We proceeded with endoscopic clearance of right nasal mass under general anaesthesia. Intraoperatively, right nasoendoscopy revealed polypoidal mass arising from lateral nasal wall, occupying the entire right nasal cavity. The nasal mass was removed with the aid of microdebrider and Blakesley forcep without difficulty. As there was only minimal bleeding, no nasal packing was re-

quired. The excised mass was sent for histopathological examination. Post-operatively, there was no complication, patient was well and was discharged home the subsequent day with a one-week appointment. Patient was also prescribed home with a one-week course of antibiotics and analgesics.

Histopathological examination revealed respiratory epithelium with cells arranged as sheets showing oval cells with eccentrically placed nuclei, cloak faced chromatin, perinuclear clearing with few binucleate cells (Figure 2). Immunohistochemical study was diagnostic of plasmacytoma. Hence, a diagnosis of nasal plasmacytoma was made. Systematic workup for multiple myeloma was negative. Subsequent follow-up demonstrated no evidence of recurrence or any nasal mass and patient has been asymptomatic till date. Patient was planned for repeated follow-up to monitor recurrence.



**Figure 1. Homogenous soft tissue mass occupying right nasal cavity.**



**Figure 2. Respiratory epithelium with oval cells, eccentrically placed nuclei, cloak faced chromatin, perinuclear clearing with few binucleate cells.**

#### 4. RESULTS AND DISCUSSION

Plasmacytoma was first described by Schridde in 1905 as a discrete solitary mass of neoplastic monoclonal plasma cells. Plasmacytoma, basically can be divided into 3 variants: multiple myeloma, osseous solitary plasmacytoma and extramedullary plasmacytoma (EMP).<sup>3</sup>

EMP is derived from plasma cells with a single class of heavy and light chains in a monoclonal proliferation of B cells.<sup>4</sup> Upper respiratory tract is considered the most common site as the submucosa in this region are rich with plasma cells. Other rare affected sites includes urinary bladder, central nervous system, orbit, gastrointestinal tract, liver, spleen, pancreas, lung, breast, skin, testis, parotid gland, mediastinum and thyroid gland.

The global incidence of EMP is 1 case per 100 000 cases. There is a male preponderance of 3 : 1 with an average age of 60 years.<sup>5</sup> Our patient discussed above is a male in his 8<sup>th</sup> decade of life. The nasal mass appears as a solitary, submucosal, sessile or polypoidal mass. Apart from that, this entity has a slowly evolving nature.<sup>2</sup> Patients usually remain asymptomatic until the mass expands.<sup>6</sup> Patients usually turns up to the otorhinolaryngology clinic with complaints of nasal obstruction, epistaxis or facial pain although, cases with ulceration or bone invasion have been reported. Cervical lymphadenopathy has been documented in 20%–25% of cases with EMP in the head and neck region.<sup>7</sup> Our patient discussed above however, complained only of progressive unilateral nasal blockage.

Albeit myriad hypothesis that exists till date regarding aetiology of EMP including viral infection, overdose or inappropriate radiation and genetic alterations in the reticuloendothelial system,<sup>8</sup> chronic irritation from inhaled irritants remains to be the most sought out for and this may be the case in our patient as well.

Diagnosis of sinonasal EMP is based solely on histopathological examination as there are no pathognomonic clinical presentation. Thus, deep biopsies should be performed as the tumour is oftentimes submucosal and mucosa may be thickened as a result of inflammation.<sup>2</sup> Apart from histopathological examination of tissue biopsy, patients diagnosed with EMP should be further evaluated for serum electrophoresis and radiological skeletal survey with bone marrow aspirate to confirm diagnosis<sup>3</sup> and to rule out other systemic disease.

Differential diagnosis of this entity among others includes lymphoma, marginal zone B-cell lymphoma, plasma cell granuloma, poorly differentiated neoplasms and reactive plasmacytosis.<sup>9</sup>

Till date, there is no consensus on gold standard treatment of sinonasal EMP. Management of sinonasal EMP comprises of single-modality treatment including surgical, radiotherapy or chemotherapy and combined management. In many centres, combination of surgical excision with an adjuvant radiotherapy is favoured as this entity known to be radiosensitive.<sup>9</sup> Some authors also advocate radiotherapy as a sole treatment for this entity.<sup>2</sup> As for the dosage of radio-

therapy, no standard protocol exist and varies between 3000 rads and 8000 rads over a period of 3 to 6 weeks.<sup>2</sup> Chemotherapy should be considered for refractory or relapse cases. Having said that, small and localised tumour located in the head and neck region can be managed solely by surgical excision as this has been shown to have similar results as radiotherapy.<sup>10</sup> In our patient, we opted for transnasal endoscopic surgical excision as the mass was more localised and there was no signs of erosion.

Long-term follow up is mandatory for patients with sinonasal EMP as 15%–20% of EMP turns to multiple myeloma over the years especially when there is a bone involvement.<sup>5</sup> Not only that, local recurrence has also been reported by countless authors. Some authors, propose serum electrophoresis and imaging upon follow-up to detect recurrence.<sup>4</sup>

Prognosis of this entity depends on the dissemination of the disease, tumour size and nodal involvement. Ten-year survival rate have been reported in 70%. Hence, these patients ought to be kept under close surveillance with a life-long follow-up.

#### 5. CONCLUSIONS

Extramedullary sinonasal plasmacytoma albeit a rare entity, is often underdiagnosed. This article emphasizes on histopathological examination for all sinonasal tumours as negligence or underdiagnoses of EMP may lead to devastating consequences as these entity has tendency to develop multiple myeloma. As for surgical management, it should be tailored according to the tumour size and location.

#### Conflict of interest

None declared.

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