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Nasal Extramedullary Plasmacytoma - A Rare Case

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Abstract

Extramedullary (extraosseous) plasmacytoma is a plasma cell tumor that grows within any soft tissue. It is a scarce neoplasm, characterized by monoclonal proliferation of plasma cells. Most of the lesions appear in the head and neck region, with the upper respiratory tract as the primary site. We here present a case of a young man who presented to us with similar presentation as of a patient with nasal polyps. Extra medullary plasmacytoma of the nasal cavity is relatively quite rare and should be in the back of our mind when we are dealing with unilateral nasal cavity masses.

Keywords: Plasmacytoma; Tumor of nasal cavity

Introduction

Plasma cells are mature B lymphocytes mostly found in various tissues and inflammatory foci and serve the function in producing immunoglobulin. Plasma cell neoplasms are represented by neoplastic proliferation of the plasma cells which can present as a single lesion or multiple lesion. Single lesions can present as solitary plasmacytoma of bone and extramedullary (extraosseous) plasmacytoma. The upper respiratory tract, due to its rich lymphatic tissue hence becomes the most common site for an extramedullary plasmacytoma, calculating up to 80% cases and this accounts for one percent of all tumors of the head and neck [1]. The etiology of this disease remains unknown, but chronic irritation from inhaled irritants and viral pathogenesis has been suggested. The approximated overall incidence of the disease is 1 case per 500,000 people [2].

Case Report

A 29-year-old gentleman presented with the complaint of left sided nose block and non-painful left nasal swelling for a total duration 2 years. Nasal endoscopy examination showed huge polyps occluding the entire left nasal cavity. Computer Tomography of paranasal sinuses showed soft tissue occupying the superior and middle meatus, extending to posterior

choanae, obliterating the left osteomethal complex (Figures 1-3).



Figure 1 CT scan axial view showing homogenous mass filling up the left nasal cavity and maxillary sinus.



Figure 2 Diagnostic nasal endoscopy showing residual mass at the inferior turbinate.

Patient underwent Functional Endoscopic Sinus Surgery (FESS), and the histopathology examination revealed

plasmacytoma. Further hematologic examination was carried out which includes blood and urine for electrophoresis; Bence-Jones proteinuria to exclude multiple myeloma. Patient subsequently was treated with 25 cycle of radiotherapy. One month post radiotherapy, his nasal endoscopy examination revealed a small mass located medially to the left inferior turbinate who which he was planned for endoscopic left medial maxillectomy for tumor clearance. Histopathology examination from the inferior turbinate mass revealed inflamed nasal polyps with amyloid deposits. Till day, patient remained under multiple discipline follow-up and has not showed any evidence of systemic manifestations.



Figure 3 CT scan post-surgery showing complete tumor clearance.

Discussion

Nasal extramedullary plasmacytoma is very scarce and thus usually missed especially in younger group of patient. It is prevalent in male around the age of 40 [1,3].

The diagnosis of this disease depends initially on clinical suspicion. Deep targeted biopsy should be carried out as the tumour is submucosal. Due to the scarcity of this tumor, most common misconception is squamous cell carcinoma since the clinical presentation is similar. Even so, the histopathology examination alone isn't sufficient to exclude multiple myeloma from an extramedullary plasmacytoma and further investigation is needed to exclude the systemic manifestations. Waldenstrom macroglobulinemia should also be considered in dealing with this kind of patients. Plasmacytoma should be differentiated from other lesions having plasma cells such as plasma cell granuloma, chronic granulomatous inflammation and rhinoscleroma. To achieve the accurate diagnosis, immunohistochemistry is needed especially with immunohistochemical studies with Cd38 antibody staining.

Imaging should include a metastatic bone survey and either a positron emission tomography/computed tomography

(PET/CT) scan or magnetic resonance imaging (MRI) of the entire spine and pelvis to rule out systemic involvement.

There is no unanimity regarding the treatment choice of nasal extramedullary plasmacytoma. The option of radiotherapy alone or surgery solely is still disputed in the published data. Some authors suggest the use of radiotherapy solely as it yield better outcome in most cases. Chemotherapy is only initiated in disseminate disease and the amount of dose for therapy is debatable.

Wax3 proffered surgery as the prime treatment in the cases of isolated lesions that could be removed with minimal morbidity in 1993. In 1988, Abemayor favored the use of radiotherapy as the main stream of treatment and surgery is only reserved for residual disease clearance [4]. In this reported case, a smaller tumor remains after receiving radiotherapy and hence was resected.

Amyloid deposits may be seen in 15-38% of extramedullary plasmacytoma [5]. This was seen in the reported case.

Wiltshaw categorized soft tissue plasmacytoma into 3 clinical stages [6]:

- **Stage I** - Limited to an extramedullary site
- **Stage II** - Involvement of regional lymph nodes and
- **Stage III** - Multiple metastasis

Following this classification, this patient is classified into Stage 1.

Prognosis of plasmacytoma, with no doubt is superior to of multiple myeloma. However, the likeliness for conversion of extramedullary plasmacytoma to multiple myeloma prevails. As the occurrences rate varies from 15-20% [7], every case need long term monitoring because extramedullary plasmacytoma can return as disseminated multiple myeloma and the transformation of this tumor is uncertain.

The patient in the study has been on follow-up for 30 months from initial diagnosis without reporting tumor recurrence.

Conclusion

Nasal extramedullary plasmacytoma is very rare. Otolaryngologist should identify the lesions and refer to hematological monitoring. However, if suspected; a histopathology examination is needed to confirm the diagnosis and followed up with radiological examination to differentiated plasmacytoma from multiple myeloma. A multidiscipline approach is needed to differentiate extramedullary plasma plasmacytoma with the poor prognosis multiple myeloma. Although mainstream of treatment for plasmacytoma is still radiotherapy as the tumor is radiosensitive but surgical intervention is also needed for complete clearance of the disease. A lifelong evaluation of the disease progression is also necessary.

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