Solitary Extramedullary Plasmacytoma of the Sinonasal Region

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Abstract Less than 10% of the patients with plasma cell neoplasms present with a solitary plasmacytoma. Though the nasal cavity is a common extramedullary site, the occurrence is extremely rare. Two cases of solitary extramedullary plasmacytoma of the sinonasal region are reported. The first of which is sinonasal plasmacytoma with concomitant HIV, an association that has been reported rarely in literature to date and is matter of much debate. In the second case report, we present an instance of surgical excision of the tumor using KTP 532 laser. The diagnosis was established using immunohistochemical techniques and multiple myeloma workups were negative in all cases.

Keywords Plasmacytoma · KTP 532 laser · HIV

Introduction

The upper respiratory tract, due to its rich lymphatic tissue is the most common site for an extramedullary plasmacytoma, accounting for 80% cases. Extramedullary plasmacytoma of the head and neck region is a rare malignant tumour comprising approximately 3% of all plasma cell tumours. They account for 1% of all tumors of the head and neck and 4% of all nonepithelial tumors of the nasal tract [1]. Approximately 80–90% of extramedullary plasmacytomas involve the Mucosa-Associated Lymphoid Tissue of the upper airways, 75% of these involve the nasal and paranasal regions [2]. 10% of these lesions on evaluation are multiple myelomas.

In 17–33% of patients with extramedullary plasmacytomas, there is a possibility of developing multiple myeloma which necessitates life long follow up. Plasma cells are derived from the clonal proliferation of B lymphocytes. On immunoperoxidase staining methods, the neoplastic plasma cells show a monoclonal staining pattern with one light chain (type) and one heavy chain (class). This helps differentiate from reactive plasma cell infiltrates with polyclonal staining patterns. There are only few cases of extramedullary plasmacytoma of the head and neck region associated with HIV positive patients reported in literature. None of the reports involved the sinonasal region.

Case Report

A thirty-one year old male patient, who was a diagnosed case of HIV, presented with swelling in the right maxillary region involving the upper alveolus of 2 months duration. It was associated with right sided nasal obstruction and foul smelling nasal discharge. On examination, he had a highly vascular swelling in the region of the right upper alveolus and hard palate, extending into the right nasal cavity. A computerized tomographic scan of the nose and paranasal sinuses with contrast showed a moderately contrast enhancing soft tissue density lesion in the right maxillary antrum with bony erosions extending into the right ethmoid sinuses and sphenoid sinus. It was also extending into the nasal cavity and nasopharynx. A diagnostic nasal endoscopy and biopsy of the lesion was done under local anaesthesia. On histopathological examination, the sections showed respiratory epithelium overlying an encapsulated tumor composed of sheets of plasma cells, bi and multi-nucleated forms separated by numerous proliferating, congested and dilated capillaries and foci of necrosis.

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Immunohistochemistry was diagnostic of plasmacytoma. Multiple myeloma workup was negative. Protein electrophoresis showed increase in globulin fraction but no M band. The serum values of IgG and IgA were raised. He was planned for external beam radiotherapy, using 30 Gy in ten fractions using lateral parallel opposed field technique. The patient discontinued treatment after six fractions due to mucositis. On follow up after 5 months he had symptoms of post nasal drip and no evidence of primary. The patient was put on melphalan and prednisolone for 5 days. This case was lost on follow up.

The second case was a sixty year old man, a known diabetic and hypertensive, who presented with a four year history of right sided nasal obstruction and recurrent epistaxis. On diagnostic nasal endoscopy, a highly vascular swelling was seen arising from the posterior end of inferior turbinate, filling the right choana. A computerized tomography imaging of the nose and paranasal sinuses revealed a moderately contrast enhancing soft tissue density lesion arising from the posterior end of the nasal septum and going into the right side of nasopharynx, without evidence of bony erosion (Fig. 1). Under general anaesthesia, using KTP 532 Laser utilizing 7 W power set on continuous mode, the mass was excised in toto under nasal endoscopic vision. The adjacent mucosa was vaporized with laser and a piece of surgicel was placed over the raw area. On histopathological examination of the mass, ulcerated hyperplastic respiratory epithelium was noted with overlying dense infiltrate of mature plasma cells, plasmacytoid cells with vesicular prominent nucleoli, Russel bodies and neutrophils. These features were suggestive of plasma cell dyscrasia. The stroma showed abundant homogeneous eosinophilic material resembling amyloid (Fig. 2). The hemoglobin level was normal and the erythrocyte sedimentation ratio was 50 mm in 1 h. Urinary Bence Jones

Fig. 1 A computerized tomography image showing soft tissue density lesion arising from the posterior end of inferior turbinate

Fig. 2 Histopathological picture diagnostic of plasmacytoma

proteins were absent. Protein electrophoresis showed the presence of alpha and gamma heavy chains. Serum values of IgG and IgM were raised. The patient underwent post-operative radiotherapy (30 Gy in ten fractions). On follow up after two months, endoscopy showed a soft tissue swelling at the posterior end of the nasal septum and roof of choana. The computed tomography studies showed features similar to the initial lesion, although smaller in size. A biopsy was confirmative of a residual disease. An endoscopic laser assisted excision of the residual lesion was done under general anesthesia. Patient has been asymptomatic over a follow up period of 8 months, without any evidence of recurrence on CT scan. The repeat multiple myeloma workup done was also negative.

Discussion

Nasal plasmacytoma though rare, is the most frequent site for extramedullary non osseous plasmacytoma [3]. There is a male preponderance of 3:1, mainly in the fifth and sixth decades of life [4]. Patients present mainly with nasal

symptoms on the same side of the tumors [5]. The common presentations are a soft tissue swelling and nasal obstruction. Other clinical features are epistaxis, nasal discharge, pain, proptosis and cranial nerve palsies. Cervical lymphadenopathy has been noted in 20–25% of head and neck solitary plasmacytomas [6]. The diagnosis is usually as a result of biopsy. Extramedullary plasmacytomas should be distinguished from non-neoplastic lesions like reactive plasmacytic hyperplasia, plasma cell granuloma, pseudolymphoma and from malignancies like haematopoietic neoplasms, malignant melanoma, olfactory neuroblastoma, anaplastic carcinoma and metastases. Differentiation between plasmacytoma and polyclonal infiltrates of plasma cells requires immunohistochemical studies [7]. The solitary plasmacytoma can be confirmed after excluding systemic disease by a serum and urine protein electrophoresis, immunoelectrophoresis, a skeletal survey and a marrow biopsy. There are three reported cases of plasmacytoma involving the gingiva in HIV positive patients. These patients had prior risk factors for HIV infection. A case of plasmacytoma involving the larynx has been reported in a HIV patient without any prior risk factors. The HIV status was noted in these cases subsequent to the diagnosis of extramedullary plasmacytoma [8]. There is no reported association between HIV infection and sinonasal solitary extramedullary plasmacytoma. Solitary extramedullary plasmacytoma in HIV positive patients may occur at a younger age. They tend to be more aggressive with a poor prognosis. This may be due to the poor immunity. A patient diagnosed to have solitary extramedullary plasmacytoma should have their HIV status evaluated, due to the possible association and poor prognosis [8]. Radiotherapy is considered the treatment of choice, surgery being limited to biopsy and to excision of residual disease [2]. Radiotherapy has been widely used as a treatment modality, but little has been written about surgery as a single management modality. However, such an option assumes importance in a developing nation, where patient follow up is erratic and treatment costs must be kept low [1]. There is not much difference in terms of survival following surgery or radiotherapy or

combined surgery and radiotherapy. Radiotherapy would also cover the regional lymphatics. KTP 532 laser can be used to excise small lesions endoscopically in the nasal cavity thus radical procedures can be avoided. The prognosis in solitary plasmacytoma without HIV infection is good. There is a possibility of developing multiple myeloma in 17–33% of these cases [4]. Chemotherapy is reserved for disseminated disease. Survival depends on dissemination of the disease [7, 9]. The prognosis depends on tumour size (>5 cm) and nodal involvement. The 10-year survival rate is 50–80% [10]. In view of high incidence of progression to multiple myeloma in due course the patients should be kept under constant surveillance.

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