

Extramedullary Plasmacytoma of Nasal Cavity: A Case Report

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ABSTRACT

Plasmacytoma is a rare anomaly and discrete solitary mass of neoplastic monoclonal plasma cells. It was first described by Schridde in 1905. Extramedullary plasmacytoma (EMP) mostly occurs during the fifth and seventh decades of life and is rarely diagnosed in younger patients. A common clinical symptom includes epistaxis, rhinorrhea, a sore throat, dysphonia and hemoptysis and is usually managed by surgery with or without radiotherapy. Plasmacytoma is a malignant disease that present either in bone marrow (medullary plasmacytoma), within the bone (solitary plasmacytoma of bone), or outside of bone, as the EMP. EMP accounts for 3% of all plasma cell tumors and approximately 90% of EMP affect the head and neck region commonly affecting the nasal cavity, paranasal sinuses, tonsillar fossa and oral cavity.

Key words: Extramedullary, head and neck, nasopharynx, plasmacytoma

INTRODUCTION

A plasmacytoma is a disease of neoplastic monoclonal plasma cells. It can affect bone or soft tissue. Depending on the site of involvement, it's divided into plasmacytoma of the skeletal system (solitary bone plasmacytoma [SBP]) and extramedullary plasmacytoma (EMP). EMP constitutes 3% of plasma cell neoplasia.^[1] It mainly affects mucosal and submucosal tissue of nasal cavity, paranasal sinuses, nasopharynx, oropharynx, and laryngopharynx. Plasmacytomas are more common in males, with a male-to-female ratio of 3:1. These tumors usually

occur in the fourth to sixth decade of life.^[2,3] Though it mostly affects single site, multiple site involvement were reported in 10% of the patients. Radiotherapy was thought to be the treatment of choice and scope of surgery was limited only up to the biopsy. However in selected cases complete endoscopic transnasal excision with repeated follow-up can give similar result without any recurrence.

CASE REPORT

A 52-year-old male patient presented to us with chief complaints of nasal bleeding and discharge from past 2 months. He was also having right sided nasal blockage. On clinical examination with the nasal endoscope, a reddish-white mass was visualized involving the floor of the right nasal cavity, lateral wall and choana. Rest of the head and neck examination was normal. Contrast enhanced computed tomography (CT) of the paranasal sinuses was done including axial, coronal and sagittal view [Figure 1 and 2]. It

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showed irregular enhancing soft tissue lesion in the posterior part of right nasal cavity extending up to the nasopharynx. He was planned for complete excision and biopsy. Endoscopic complete excision of the right nasal mass was done.

Excised tissue (Figure 3) was sent for histopathology (Figure 4). The tissue was of dimension 4 cm × 3.5 cm × 1.5 cm. Section showed a nodular neoplasm composed of round to polygonal monoclonal plasma cells arranged in diffuse sheets with intervening fibrovascular stroma. Many tumor cells are binucleated and have eccentrically placed nuclei. Peripheral areas of the lesion show acellular matrix deposits with adjacent giant cell reaction. It was also positive for light lambda chains for which it was diagnosed as plasmacytoma.

To confirm the diagnosis and rule out multiple myeloma all other investigations were performed. X-ray of skull, CT scan of chest, abdomen and pelvis was done to rule out any other site of involvement. Urine examination showed absence of Bence Jones protein. His blood parameters, renal function test and bone marrow examination were normal. After the surgery and investigations EMP diagnosis was confirmed and he was kept under observation. There was no sign of recurrence after 2 years of follow-up.

DISCUSSION

EMP is uncommon frequency in about 0.5% in head and neck region. Plasma cell neoplasm is divided into disseminated multiple myeloma, multiple myeloma EMP and SBP. The former two affects multiple organs and the latter two affects single organ. As the name



Figure 1: Axial view of computed tomography scan nose and paranasal sinuses showing mass in the right nasal cavity extending to choana



Figure 2: Coronal view of computed tomography scan nose and paranasal sinuses showing mass in the right nasal cavity extending to choana



Figure 3: Surgical excision specimen from the right nasal cavity and choana

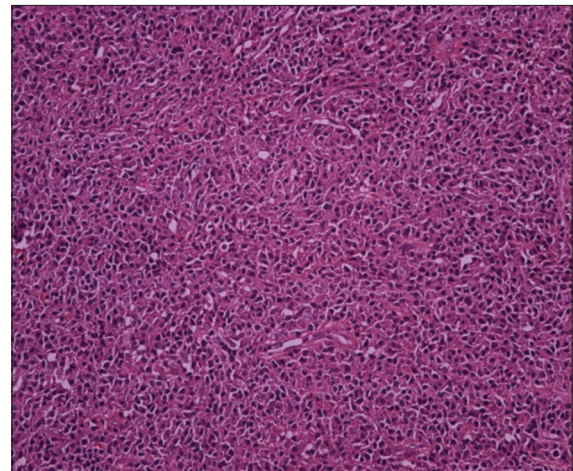


Figure 4: Round to polygonal plasmacytoid tumor cells in sheets with intervening fibrovascular stroma

suggests EMP affects soft tissues. It represents 3% of all the plasma cell neoplasm and mostly affect males. It mainly affects the mucosa associated lymphoid tissue of the nose, sinuses and nasopharynx.^[4] Rarely affected sites are urinary bladder, central nervous system, orbit, gastrointestinal tract, liver, spleen, pancreas, lung, breast, skin, testis, parotid gland, mediastinum, and thyroid gland.^[5,6] Though its pathophysiology is unclear but some studies relate it with chronic irritation due to inhaled agents and viral infection.^[7]

Symptoms depend upon the site of involvement. Presenting features of nasal EMP are nasal blockage, mass in the nose, nasal discharge, bleeding from nose and sometimes pain. Some patients also have cranial nerve palsy and cervical lymphadenopathy.^[8] For diagnosis of EMP complete chain of investigations are needed. These includes complete blood count, serum and urinary protein electrophoresis, peripheral blood smear, renal function test, liver function test, endoscopy of aerodigestive tract, X-ray of long bones, CT and magnetic resonance imaging scan of affected area and bone marrow study.

Wiltshaw^[7] classified soft-tissue plasmacytoma into three clinical stages, as follows:

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

The differential diagnosis includes: Lymphoma, marginal zone B-cell lymphoma, plasma cell granuloma, poorly differentiated neoplasms and reactive plasmacytosis. The different treatment options are surgical excision, radiotherapy, chemotherapy, and combination of surgery and radiotherapy. Radiotherapy is considered to be the treatment of choice for EMP of head and neck region. Previously surgery was only limited to the biopsy. But for small tumors of head and neck, surgical excision can be a good treatment option which provides same result as radiotherapy.^[9] Chemotherapy is reserved for EMP with distance metastasis. Combination treatment may provide the best results.^[5] The 10-year overall survival rate is 70%.^[10] The reported conversion rate of EMP to multiple myeloma is 15-30%^[8] and is associated with a poorer prognosis. Dissemination of the tumor takes place in 35-50% of EMP.^[11] Periodic follow-up every 6 weeks for first 6 months and then every 6 monthly is required to look for the development of lesion over the primary site

and its progression to multiple myeloma. Follow-up should include clinical examination along with hematological and radiological evaluation.

In our case as the disease was limited only to the nasal cavity and nasopharynx, we have opted surgical excision. Intra-operatively by using high end endoscopes and high definition camera we were able to remove the tumor completely. Though we have kept radiotherapy reserved for recurrence, but after 2 years of follow-up there was no evidence of disease.

CONCLUSION

EMP is a rare disease of solitary mass of neoplastic monoclonal plasma cells affecting mainly the soft tissues of head and neck region. Specific investigations are needed to differentiate it from other plasma cell neoplasms. The best management required for such type of lesions or tumors are surgical intervention combined with radiotherapy. For smaller lesions through radiation (radiotherapy, etc.) can be considered to be the treatment of choice but complete surgical excision with regular follow-up can also give the similar results with an excellent prognosis.

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