



*Case Report*

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**Extramedullary Plasmacytoma of the Nasopharynx: A Case Report**

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**Abstract**

*Plasmacytoma is a rare and discrete solitary mass of neoplastic monoclonal plasmacells. Extramedullary plasmacytoma(EMP) tends to occur during the 5th and 7th decades of life and is rarely diagnosed in younger patients. Here we report a 38yr old female patient presenting with longterm nasal obstruction; who was found to have emp of nasopharynx. The patient was treated with radiotherapy with a satisfactory outcome.*

*Key Words: plasmacytoma, extramedullary ,head and neck, nasopharynx.*

**Introduction**

Plasmacytoma is a rare and discrete solitary mass of neoplastic monoclonal plasmacells, which was first described by schridde in 1905(1). Emp has been reported rarely and accounts for 4% of all non epithelial tumors of upper respiratory tract(2). while occasionally localised to git, lungs, testes and skin. It has been previously reported that 80% of Emp are localised in H&N region(3,4). Common clinical symptoms include epistaxis, rhinorhea, a sore throat dysphonia and hemoptysis(5,6). EMP is usually managed through radiotherapy with or without surgery. The current presents the case of a middle age female patient with Emp of nasopharynx who was treated successfully with radiotherapy. Written informed consent was obtained from the patient.

**Case Report**

A 38yr old female was reported to the Department of Radiotherapy in the month of July at MNJ Cancer Hospital due to chief complaints of intermitent epistaxis from nose since 3months, In addition, the patient reported difficulty in swallowing ,persistent nasal obstruction and snoring. A physical examination revealed a mass seen in nasopharynx. on clinical examination of neck, a node is present on the left side of neck level 2 was seen. Magnetic Resonance Imaging of head and neck showed evidence of Nasopharynx-T1/T2 isointense moderately homogenously enhancing soft tissue density lesion in nasopharynx along the midline

approximately m/s 32x30mm ,superiorly fat planes lost with planum sphenoidale.Inferiorly fatplanes lost with distal aspect of hardpalate.Anteriorly fat planes lost with superior and middle turbinate(Right>left). There is evidence of few enlarged level IB, 2 nodes on the left largest m/s 18x15mm at level 2a.

On HPE: Microscopically The tumor shows cells arranged in sheets and individually scattered. Individual cells are round to oval with moderate amount of cytoplas; round hyperchromatic nucleus and prominent nucleolus focal areas of hyalinization and congestive blood vessels are seen f/s/o undifferentiated Nasopharyngeal carcinoma.

Specimen -> Nasopharyngeal mass biopsy -> Block received.[2nd opinion)

Microscopillay -> Sections are infiltrated by sheets of uniform polygonal cells with eccentric nucleus with cart wheel chromatin and a rim of cytoplasm with a perinuclear clearing in some cells.Few of these have central nuclei and occassional binucleate forms are seen.Homogenous Esoinophilic material is seen between.The material is congophilic with reddish green birefringence under polarized light confirming it as amyloid.



Figure 1

On Magnetic Resonance Imaging of head and neck showed evidence of Nasopharynx-T1/T2 isointense moderately homogenously enhancing soft tissue density lesion in nasopharynx along the midline approximately m/s 32x30mm ,superiorly fat planes lost with planum sphenoidale.Inferiorly fatplanes lost with distal aspect of hardpalate.Anteriorly fat planes lost with superior and middle turbinate(Right>left).e/o few enlarged LN level 1b; 2LN on left;largest m/s 18\*15 at level 2a.(fig:1)

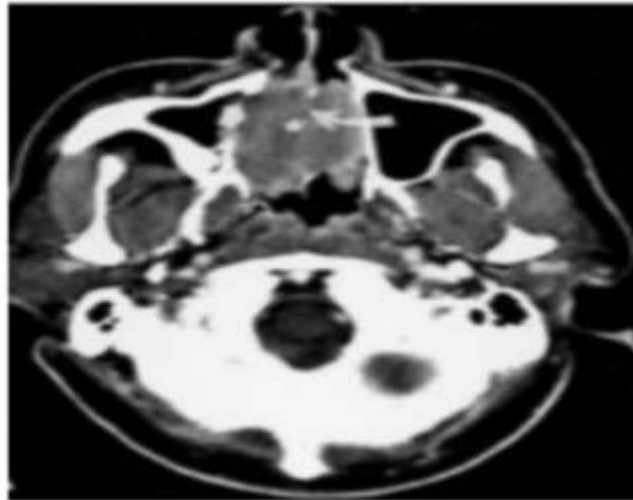


Figure 2

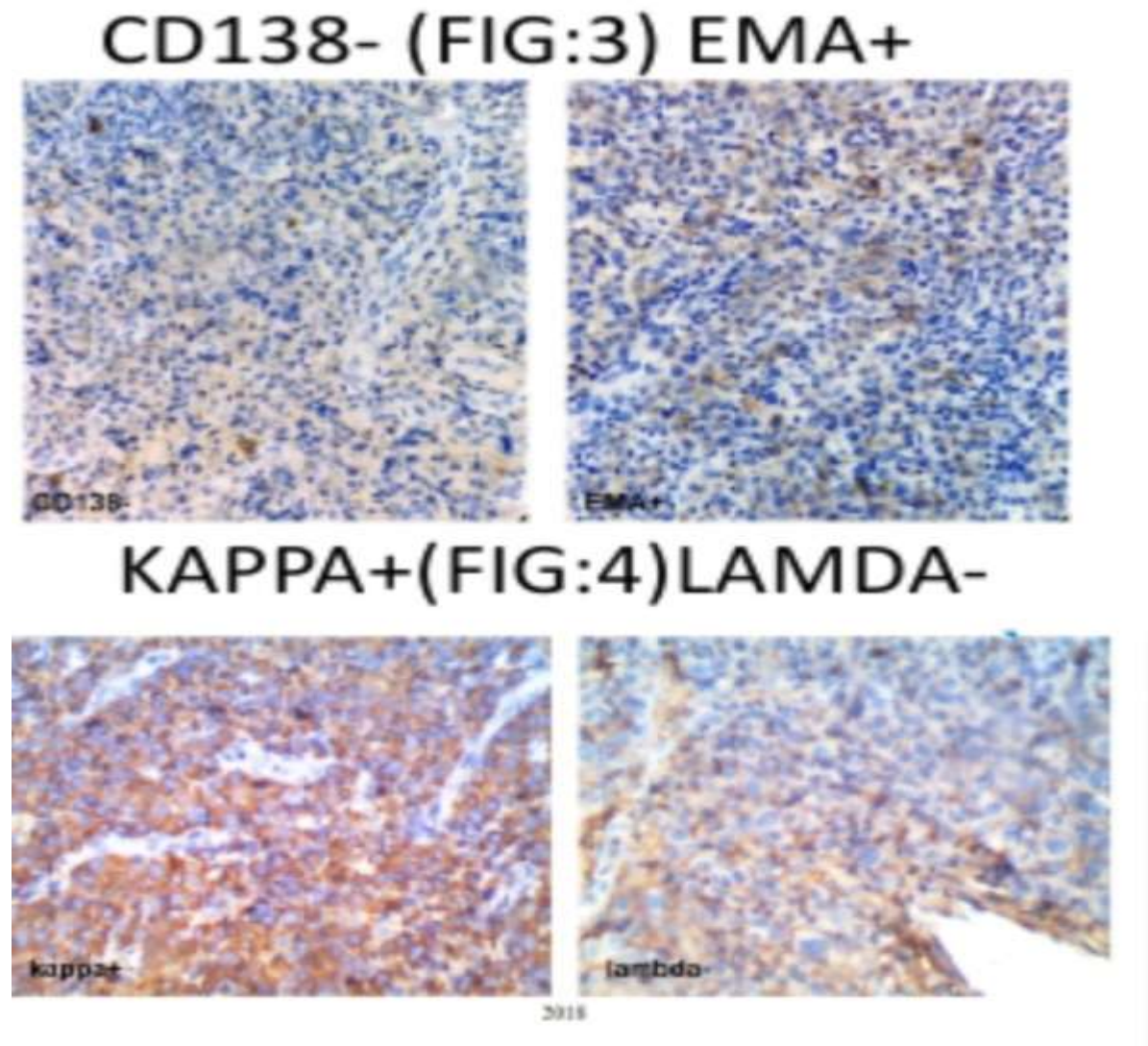


Figure 3

## Diagnosis

The above features are consistent with a plasmacytoma with amyloid deposits.

The positivity for vimentin and EMA and negativity for pancytokeratin reported elsewhere are consistent with the immune profile of plasma cells.

IHC report showed: Kappa - Positive

Lambda - Negative

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IMP: Morphology is correlation with Immune stains consistent with plasma cell Neoplasm.

FNAC from left cervical L. N showed: Plasma cell Neoplasm

In addition; expression was +ve IgG, K+ve; m band present; i.e monoclonal gammopathy was seen in IgG and Kappa region. The CBP; sr. calcium creatinine and uric acid within normal limits. On Electrophoresis of sr. protein; m band was seen in gamma region; B2 globulin was reduced; gamma globulin was increased and whole body bone survey revealed no detectable osteolytic lesions. A bone marrow aspiration was in normal limits. A final diagnosis of Extramedullary plasma cell of Nasopharynx was determined and IMRT was planned for the patient.

## Discussion

Plasmacytomas are rare tumors that often appear in the H&N region and are characterised by monoclonal proliferation of plasma cells. These are classified into 3 subtypes. The most common type is multiple Myeloma; which is usually a disseminated disease and is characterised by abnormal M protein. The other two types; Solitary plasmacytoma of bone and EMP of soft tissue, are considerably less common. On both clinical presentation and pathologic examination these tumors may be confused with more common tumors of head and neck. Medullary plasmacytoma has more tendency for conversion to multiple Myeloma when compared to extramedullary plasmacytoma. The diagnosis of EMP of soft tissue has been based on following Inclusion criteria. (1) Pathological tissue evidence of monoclonal plasmacells involving a single extramedullary site. (2) No bone marrow involved. (3) Negative skeletal survey results. (4) No anemia; hyperkeratotic cornea or renal impairment caused by plasmacell dyscrasia and (5) low serum or urinary levels of monoclonal immunoglobulin (8). There is greater male preponderance (M:F IS 3:1) and EMP tends to occur during the 5th and 7th decades of life; rarely being diagnosed in younger patients. Only four cases of EMP have been previously reported in relatively young patients:

(1) Two 3.5yr old males with unexpected EMP following adenoidectomy for chronic rhinosinusitis. (2) A 12yr old female who presented with progressive hoarseness and was subsequently found to have EMP coexisting with localized involving the larynx (10) and (3) A 11yr old male who presented with an EMP of orbit (11). (4) In present patient the tumor was localized in the nasopharynx. There was no involvement of bony structure or bone marrow and the diagnosis of solitary EMP of nasopharynx was confirmed. The etiology of this disease remains unknown; but factors such as viral (EBV) pathogenesis and chronic irritation from inhaled

irritants have been previously indicated(12-14) Despite the typical presentation as a locally destructive tumor, plasmacytomas are highly radiosensitive; 70-80% survival may be obtained with the use of radiotherapy. Radiotherapy remains the main stay for management of EMP.

The biochemical nature of amyloid deposits was also shown to be of kappa Ig light chain; suggesting the pathogenic relationship between Plasmacytoma and Amyloid deposition in the nasopharynx of this patient. There was no other evidence of malignancy or amyloid else where. This case also supports the concept that localized Nasopharyngeal Amyloidosis may be manifested of low grade B – cell lymphoma(10)

Previously; Susnerevala et al proposed a pathological grading system based on multiple myeloma grading criteria; tumors classified into low, intermediate and high grades, which have been found to correlate closely with outcomes. The study recommended the use of adjuvant chemo in patients with higher grade disease (14). The general EMP; are considered radiosensitive with a local control rate of 90-100% (15).

A radiation dose of 40-50Gy delivered to the primary site of EMP in the nasopharynx is usually recommended(8). Irradiation to the neck is required only in cases with clinically positive cervical node mets. In a recent study it was found that the primary modality of EMP is radiotherapy. Moreover, RT combined with surgery produced an improved outcome, as determined by survival rates(12). Although the rate of chemotherapy in EMP treatments has not been established, chemo is usually considered for EMPs with high risk factors for local treatment failure (tumor size >5cm) and in case of refractory disease.

Followup radiological and electrophoresis assessment is required following treatment to detect recurrence and progression to multiple myeloma, which occurs in 10-30% of cases. The overall 10 yr survival rate is 70%. (7, 8) A literature search revealed no publications supporting the use of surgery alone to treat EMP. In the current case, the patient was treated with IMRT.

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