Case report

An Extramedullary Plasmacytoma of the Nasopharynx: A rare entity

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Abstract:

A 64 year old female with no underlying medical illness presented with 3 months history of unilateral nasal blockage and rhinorrhoea. Nasopharyngeal examination revealed a smooth surface mass in the posterior nasal space. Endoscopic biopsy confirmed the mass to be Extramedullary Plasmacytoma. Investigation pertaining to multiple myeloma revealed no evidence of the disease. The mass was excised endoscopically and the patient completed 25 cycles (50Gy) radiotherapy. Patient had no evidence of recurrence after 6 months follow up.

Key words: nasal blockage; nasopharynx; biopsy; extramedullary plasmacytoma

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Introduction:

Extramedullary plasmacytoma (EMP) is a rare tumor belonging to the category of non-Hodgkin lymphoma. It makes up 4% of all plasma cell neoplasm and occurs mainly in the upper aerodigestive tract (UAD) [1] .Patient with EMP may develop multiple myeloma in 17-30% of cases. [2] Male to female ratio is 2:1 [2] . The median age of patients is 55 years [2] . The following case illustrates a rare case of EMP diagnosed in a 64 year old lady.

Case summary:

A 64 year old malay female presented with frequent left side nasal blockage associated with rhinorrhoea for three months. She had no epistaxis, neck nodes and ear symptoms. Clinically she was stable. Anterior rhinoscopy, intraoral and ear examination revealed no significant findings. Rigid nasoendoscopy revealed smooth surface reddish mass at the centre of the nasopharyngeal wall and continued with the nasal septum . Biopsy taken showed nasopharyngeal mucosa partly lined by respiratory and metaplastic squamous epithelium. The stroma revealed diffuse infiltration by neoplastic plasma cells. The cells display fairly uniform eccentrically located nuclei and moderate amount of eosinophilic cytoplasm (Figure 1 & 2). These cells were positive to CD 79a, CD56 and CD138 with evidence of light chain restriction. They are negative to CD3, CD20 and CKAE 1/3 indicating malignant plasma cells consistent with as extramedullary plasmacytoma.

CT scan showed a homogenously enhancing lobulated soft tissue lesion arising from the posterior aspect of the left nasal cavity extending into the nasopharynx and it is abutting the nasal septum which look eroded and left pterygoid bone and inferiorly it is abutting the hard and soft palate. It is measures 4.3cm x 2.6cm (Figure 3). No significant cervical lymph nodes noted.

Several investigations to rule out multiple myloma (i.e Urine assays for Bence Jones protein, bone marrow biopsy and skeletal survey) was performed and all revealed negative.

The mass was excised via endoscopic approach. Intraoperative findings showed a well define smooth surface mass arising from roof of nasopharynx extending to posterior part of septum. The overlaying mucosa look healthy with no increase in vascularity and no contact bleeding.

Radiotherapy was given postoperatively for 25

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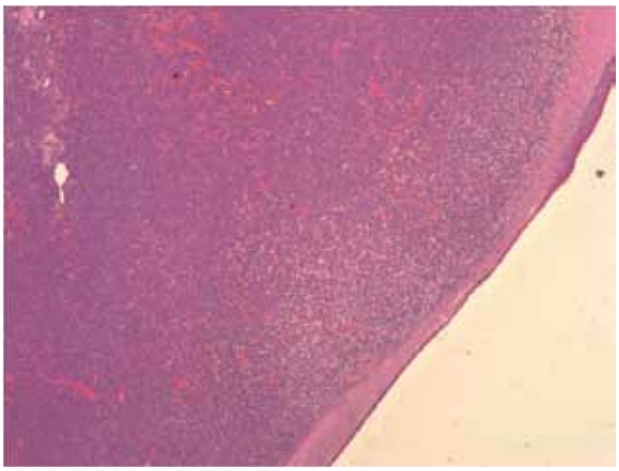

Figure 1: A circumscribed nodule covered by attennuated benign stratified squamous epithelium.

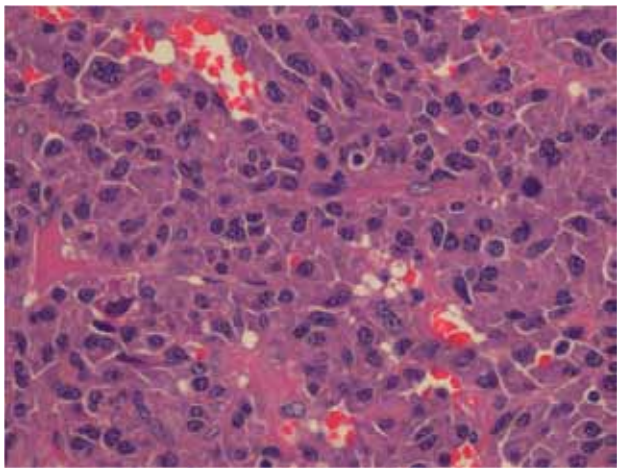

Figure 2: A neoplastic plasmacytoid cells, which display fairly uniform eccentrically located nuclei with moderate amount of eosinophilic cytoplasm.

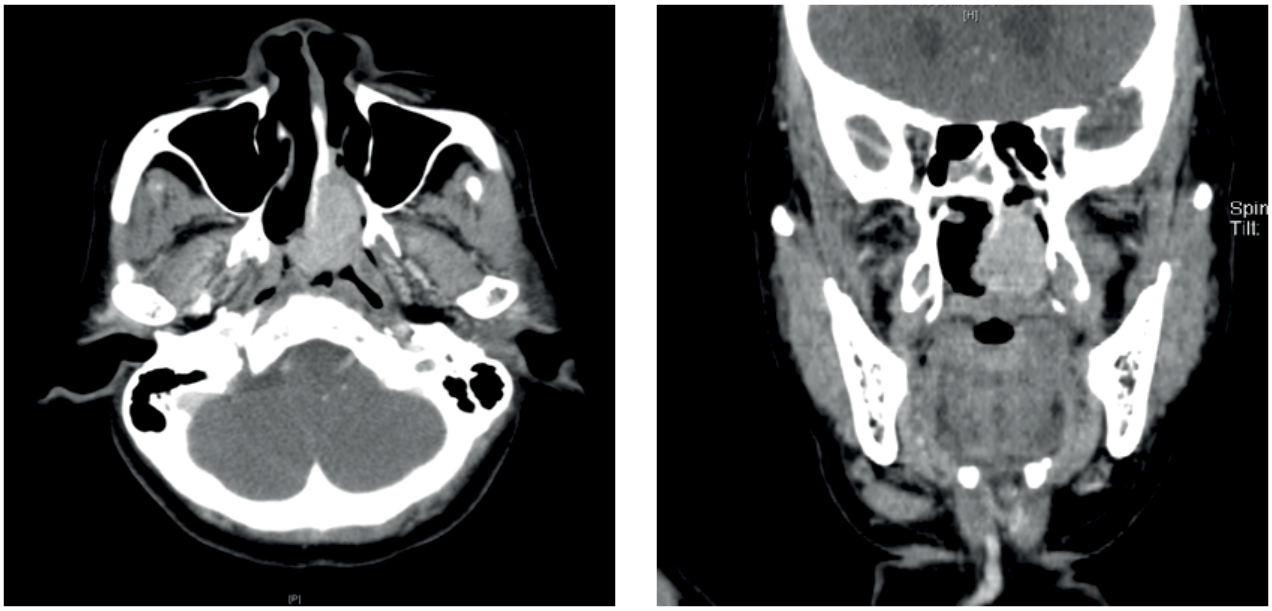

Figure 3 : Axial and coronal view of the patient showed homogenous enhancing mass occupied posterior aspect of left nasal cavity abutting the septum (partially eroded) and extended to nasopharynx.

cycles (50 gy). Patient was well after 6 months follow up and a subsequent whole body CT scan showed no evidence of recurrence.

Discussion:

Extramedullary plasmacytoma (EMP) is a rare entity belonging to the category of non-Hodgkin lymphoma. It as a rare monoclonal neoplastic proliferation of plasma cells characterized by an extramedullary tumor of clonal plasma cells with no or small amounts of monoclonal protein in the serum and/or urine, normal bone marrow, normal skeletal survey, and no end-organ damage from the plasma cell proliferative process. [3]

EMPs make up 4% of all plasma cell tumors and occur mainly in the upper aerodigestive tract (UAD) [1]. EMPs account for only 1% of all tumors of head and neck region and only 4% of all nonepithelial tumor of the nasal tract [1]. Typical location of EMPs in head and neck region are pharynx (21.5%), nasal cavity (19.3%), oral cavity (14.7%) and paranasal sinus (13.0%). [4,5]

Male to female ratio are 2:1 [2]. The median age of patients with EMPs is 55 years. [2] Multiple myeloma(MM) may developed in 17 -33% of EMPs [2]

Genetic factors, radiation exposure, smoking , occupational exposures, chronic stimulation caused by inhaled irritants or viral infection have been

implicated as possible etiologic agents [6,7,8].

Diagnostic criteria for solitary EMP include monoclonal plasma cell histology, bone marrow plasma cell infiltration showing less than 5% of all nucleated cells, absence of any osteolytic bone lesion, absence of hypercalcemia or renal failure and low levels of paraprotein concentration if present. [9] By immunohistochemical techniques, a monoclonal staining pattern may demonstrate either one heavy chain class, one light chain type or both. [6,7] .CD 138 has been recognized as a marker for neoplastic plasma cells[10]. Like in this case, immunohistochemical technique revealed positive for CD 138, CD 79a and CD56 with evidence of light chain restriction.

Systemic work-up such as serum protein electrophoresis, urinalysis for the Bence-Jones protein, skeletal survey and bone marrow biopsy is vital to exclude multiple myeloma or plasmacytoma of the bone in EMPs [11,12] For this case, bone marrow aspiration cytology, urinalysis for Bence Jones protein and skeletal survey all were negative. There was no evidence of renal failure. In addition, EMP shows nonspecific CT and MR imaging features.[5]

Radiotherapy is the treatment of choice for EMPs which is localized in the head and neck region. [13] Radiation therapy of 4000-5000 cGys assure local (95- 100% control rate) and regional tumor control. [14, 15]

Five year survival rates between 30 to 82% and 10 year rates are 50 to 90%.[14] Long-term follow-up is essential because local recurrence and development of disseminated disease into multiple myeloma or radiation-induced malignancy can occur many years after diagnosis. [3]

Conflict of interest: None

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