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Solitary Extramedullary Plasmacytoma Presenting As Proptosis – A Case Report

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Abstract: *Solitary Extramedullary Plasmacytomas (EMPs) are nonepithelial neoplasms of plasma cell origin categorized among Non-Hodgkin lymphomas, without any bone marrow involvement or systemic spread as seen in Multiple Myeloma. They are uncommon tumors comprising 3% of all plasma cell neoplasms. We present in this report a case of EMP of left nasal cavity leading to left eyeball proptosis and movement restriction. Histopathological and immunohistochemical staining of the excised mass confirmed the diagnosis of Plasmacytoma.*

Keywords: *Extramedullary plasmacytoma, EMP, Nasal cavity,Proptosis.*

I. Introduction

Plasmacytoma is abnormal proliferation of monoclonal plasma cells in either the bone or soft tissue. It presents either as a single lesion (solitary plasmacytoma) or as multiple lesions localized in bone marrow (Multiple Myeloma). Solitary plasmacytomas most frequently occur in the bone but they can also be found in the soft tissues as solitary extramedullary plasmacytomas (EMPs). EMPs are rare forms of the Non-Hodgkin lymphomas. They represent about 3% of all plasma cell tumors [1]. The tonsils, nasal cavity, paranasal sinuses, nasopharynx,are the most commonly affected sites [2]. The median age at diagnosis is 55 to 60 years, and approximately two-thirds of the patients are males [3]. A combination of Surgery (for resectable tumors) and Radiotherapy is the treatment of choice for EMPs [4].

II. Case Report

A 60 year old male patient attended the outpatient clinic of ophthalmology with chief complaints of forward protrusion of left eyeball since 3 months, gradual in onset and nasal obstruction since 1 month with no complaints of epistaxis. Other history was nil significant.

Ocular Examination revealed eccentric proptosis(down and out) with fullness of both eyelids, restriction of movements in upgaze, with mechanical ptosis, absence of convergence and no change on valsalva (Figure.1). On palpation a non reducible mass is palpated in ethmoidal region extending upto 1/3 rd of the upper medial orbital margin, firm in consistency, Tenderness present. Anterior segment examination showed congested conjunctiva with a normal intraocular pressure. Posterior segment findings were nil significant.

Figure.1. Eccentric proptosis of left eye with fullness of both eyelids, restriction of movements in upgaze,

Anterior Rhinoscopy showed a Pinkish smooth mass in the left nasal cavity extending up to vestibule with DNS. On probing the mass is firm in consistency, sensitive to touch, not bleeding and probe cannot be passed laterally.

Other routine blood investigations were normal

CT Scan of Brain & PNS showed a lobulated soft tissue attenuating lesion of size7.6x7x4.5cm in the left nasal cavity with lateral extension into maxillary antra, superior extension to left orbit eroding the orbital floor and superior wall of maxillary sinus, with deviation of nasal septum to right . suggestive of a malignant lesion with intra orbital extension (figure.2).

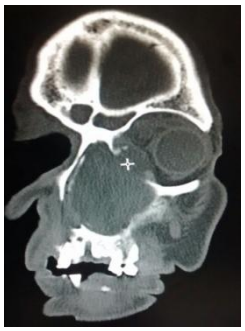

Figure.2. soft tissue attenuating lesion in the left nasal cavity with extension into maxillary antra, left orbit eroding the orbital floor, with deviation of nasal septum to right

MRI scan revealed a soft tissue intensity lesion involving left nasal cavity, extending along maxillary, ethmoid sinuses, orbit on left indenting the left medial rectus and displacing the globe anteriorly and laterally. suggestive of a malignant lesion with intra orbital extension (Figure3,4).

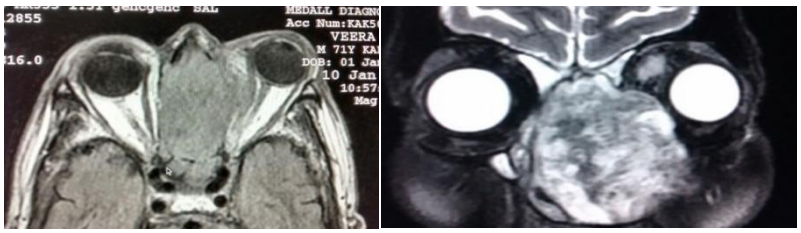

Figure3,4: soft tissue intensity lesion involving left nasal cavity, extending to orbit on left indenting the left medial rectus and displacing the globe anteriorly and laterally

Based on the above findings we proceeded for Excisional biopsy. The lesion was nonencapsulated and multilobular. Frozen sectioning of the mass was consistent with plasmacytoma. The tumor was completely excised from the sinus cavity. Histopathological and immunohistochemical staining of the mass confirmed the diagnosis of Plasmacytoma.

Postoperatively, the patient had a further diagnostic work-up, including bone marrow biopsy and aspiration, skeletal X-ray survey, serum electrophoresis, immunoglobulin quantification, serum and urine immunoelectrophoresis, chest X-ray, and abdominal ultrasonography. The bone marrow biopsy showed a plasma cell infiltration of less than 5% of all nucleated cells.The absence of hypercalcemia, renal failure, osteolytic bone lesions and other organ involvement in a patient with localized disease suggested the diagnosis of Solitary Extramedullary Plasmacytoma.

The patient was given adjuvant radiotherapy at a dose of 40 Gy. symptomatic relief and evidence of tumour regression was noted on repeat CT scans. He was disease-free at 12 -month follow-up with no progression to Multiple Myeloma.

III. Discussion

Extramedullary plasmacytomas (EMPs) are localized plasma cell neoplasms in tissues other than bone. Nonosseous EMPs arise most common in the mid facial region (72%), and sinonasal EMPs account for 75% of these tumours[4]. There is a male preponderance (3:1) of patients, and an average age of 60 years.[6]

The clinical presentation of paranasal sinus EMP depends on the size and site of involvement. Nasal obstruction (80%), soft-tissue swelling, epistaxis, nasal discharge, pain, and proptosis.in the present case the patient presented with proptosis and restricted ocular movements as a late presentation. Nasal obstruction has not been the chief complaint as with other cases. CT and MRI are complementary in evaluating the size, location and involvement of the adjacent structures [7]. CT and MRI images show soft tissue masses or infiltrative lesions with variable enhancement. Bony destruction is displayed depending on the expansion of the tumor. Isointense images on T1 and iso- to hyperintense images on T2-weighted sequences are well demonstrated on MRI. In the current case, CT examination was useful in demonstrating the bone erosions. MRI was superior to CT in defining the malignant character of the soft tissue mass, but it could not distinguish plasmacytoma from other probable causes (squamous cell carcinoma or adenocystic carcinoma).

The diagnosis of EMP was confirmed by histopathology. Our patient was referred to a hematology clinic for further diagnostic work-up[11]. Immunohistochemistry was done, and the tumor cells were CD20 , Cytokeratin negative, CD138 and MUM-1 positive, with lambda chain restriction. There was no M band or globulin fraction increase on serumprotein electrophoresis. Serum beta 2 microglobulin was slightly increased to

2400 µg/L (N=800-2200 µg/L). Biochemical investigations, and skeletal survey were normal. The bone marrow biopsy showed a plasma cell infiltration of less than 5% of all nucleated cells.

As plasma cells are radiosensitive, a dose of 40-50 Gy radiotherapy is advised in the treatment [10]. According to Alexiou et al., sinonasal EMPs with bone destruction should be treated with a combination of surgery and radiation [4]. Because of the bone destruction in the present case, our treatment of choice was surgery combined with a radiation dose of 40 Gy. In our opinion, complete surgery with postoperative radiotherapy is more promising in the treatment of solitary EMP.

In conclusion, One should always include the possibility of plasma cell tumors (EMPs) in older patients presenting with symptoms of nasal obstruction, or Proptosis. All elder patients with proptosis must be screened for extraorbital pathologies also. Long-term follow up for disease recurrence and progression must be considered.

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