



Case Report

Extra medullary Plasmacytoma of nasal cavity- A rare case report.

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Abstract

Extramedullary plasmacytoma is a rare neoplasm characterized by monoclonal proliferation of plasma cells and accounts for about 3 - 5% of all plasma cell neoplasms including bone and extramedullary tumors. Extramedullary Plasmacytomas are more predominant in males who are in the 5th – 6th decade. It is most often located in the head and neck region. It usually affects upper respiratory tract 80% cases and 15% spread to cervical lymph nodes. They usually present as rhinorrhea, epistaxis and nasal obstruction. Approximately 15% progress to myeloma and 25% have local recurrence, may spread to regional lymph nodes or metastasize to distant sites.

Keywords: Extramedullary plasmacytoma, rhinorea, nasal cavity, myeloma, immunohistochemistry markers.

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INTRODUCTION

Extramedullary plasmacytoma is a rare neoplasm characterized by monoclonal proliferation of plasma cells. It accounts about 3 - 5% of all plasma cell neoplasms including bone and extramedullary tumors [2]. Extramedullary Plasmacytomas are more common among males who are between 5th – 6th decade and is most often located in the head and neck region. It usually affects upper respiratory tract 80% cases and 15% spread to cervical lymph nodes. They usually present as rhinorrhea, epistaxis and nasal obstruction. Approximately 15% progress to myeloma and 25% have local recurrence, may spread to regional lymph nodes or metastasize to distant sites[3][4]. Radiologically, present as solitary bone lesions, which are usually lytic lesion with a narrow zone of transition to normal bone. MRI may be preferred radiological investigation to rule out additional bone lesions. Based on size of plasmacytoma, low polyclonal immunoglobulins, persistence of M protein following radiotherapy and osteopenia they are considered to have poor prognosis [5, 8].

Case Report

A 23year old male presented with a history of progressive nasal obstruction for past 1 year on right side and presented with history of recurrent sneezing, mild epistaxis and intermittent pain. On anterior rhinoscopy examination, showed a nasal mass in right side. On radiological examination (CT scan) of nose and paranasal sinuses showed a well-defined oval heterodensed soft tissue lesion in right posterior nasal cavity likely extending into choana with mild bony destruction (**Fig.1**). Surgical excision of lesion was done and on histopathological examination & immunohistochemical markers revealed Extramedullary plasmacytoma.

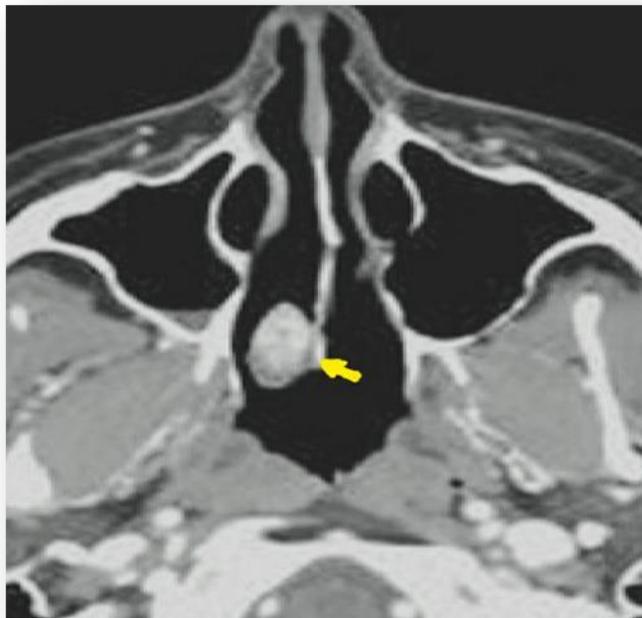


Fig.1: On radiological finding, well defined oval heterodensed soft tissue lesion in right posterior nasal cavity likely extending into choana with mild bony destruction.

On gross examination, the tumor was a gray-white nodular mass that measured 4 x 3.5 x 2 cm. A cut-surface analysis showed a pale brown with foci of congestion. On Microscopic examination, the mass was made up of sheets of neoplastic plasma cells with areas of congestion, lymphocytic infiltrate and scattered bony trabeculae (Fig.2a). Focal area shows a round cells with scant cytoplasm. Eccentric hyperchromatic nuclei with irregular chromatin distribution (Fig.2b) and binucleation were present (Fig.2c). On immunohistochemical marker study shows positive staining for CD 138 and negative for CD20 (Fig.3a & 3b).

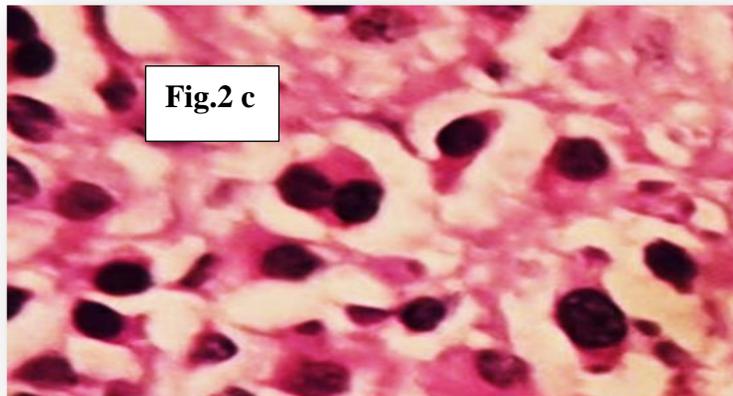
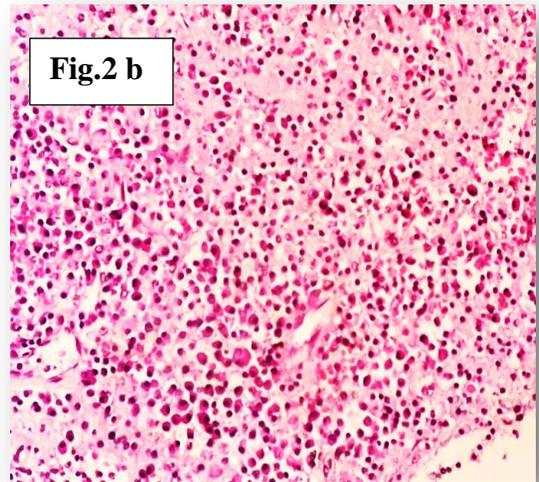
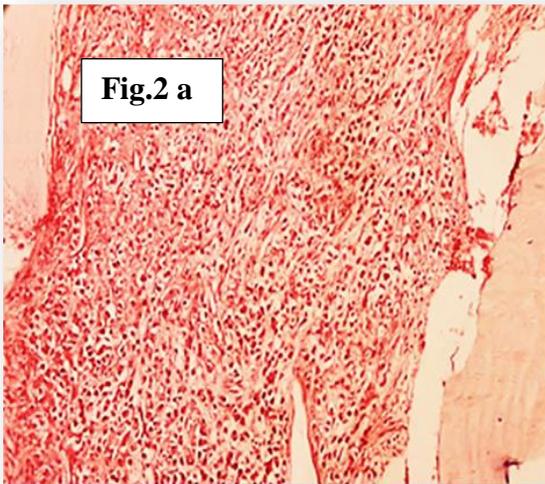


Fig.2: Histopathological sections revealing the presence of Neoplastic plasma cells with areas of congestion, lymphocytic infiltrate(a), Focal area showed round cells with scant cytoplasm. Eccentric hyperchromatic nuclei with irregular chromatin distribution (b) and binucleation were present (c).

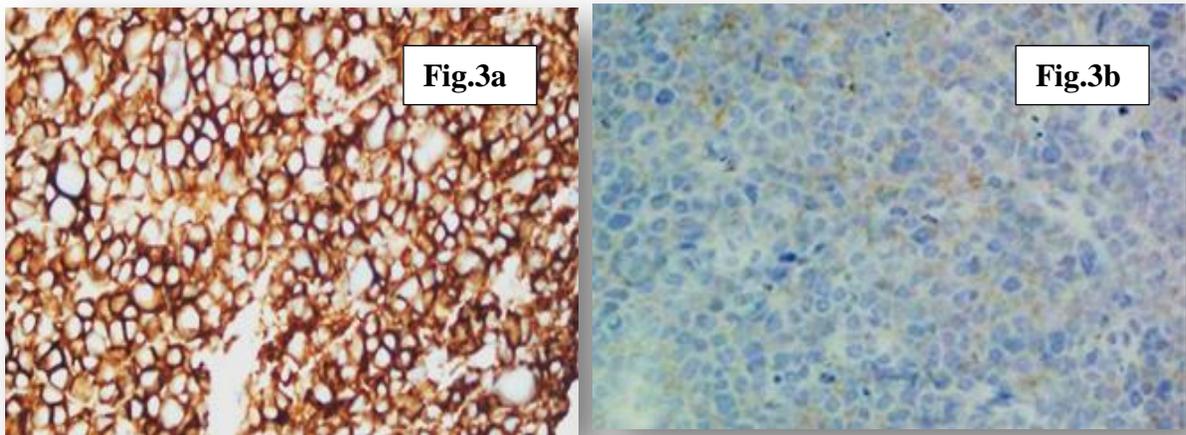


Fig 3: Immunohistochemical analysis shows the positive staining for CD 138 (a) and negative for CD20 (b).

Discussion

Extramedullary plasmacytoma (EMP), defined by the presence of clonal plasma cell proliferations extraneous to the bone marrow, is increasingly noticed as part of the constellation of multiple myeloma presenting symptoms. The incidence of EMP on diagnosis ranges from 7-18% and an observed association has been made with Ig D myeloma [4]. It was typically associated with advancing disease, relapse after being treated also. The mechanisms for extramedullary disease are multifactorial, including hematogenous spread and direct extension from skeletal plasmacytomas; having much to do with the cytogenetics, molecular mutations of the myelomatous cells [6]

The pathogenesis of extramedullary tumour growth is complex, and is probably due to loss of the interaction between plasma cells and the bone marrow microenvironment, which plays crucial roles in the processes of cell proliferation and migration. Among the causes of extramedullary plasma cell relocation, downregulation of adhesion molecule (CD56 and VLA-4) expression, dysregulation of chemokine receptors (CCR1, CCR2, and CXCR4), and an increase in heparanase-1 activity are suggested [7, 8]. These mechanisms are currently only suggested, since there are no scientific studies on a large number of patients, and the results of studies of small patient populations presented in the literature are contradictory

When these tumors occupy the nasal cavities, a differential diagnosis should be conducted in order to rule out other bleeding tumors, especially squamous cell carcinomas. So, an effective analysis must be performed to confirm the existing plasma cell disorder. On immunohistochemical marker study shows positive staining for CD 138 and negative for CD20 [11]

The prognosis of extramedullary plasmacytoma is much more favorable as compared to multiple myeloma as the tumor is radiosensitive, and surgery may occasionally be used to complement the treatment. Controlled clinical trials are needed to establish a definitive treatment of choice for the management of these patients. Though, extramedullary plasmacytoma occurs commonly in old age group, in our case it was observed in young adult hence, we presented this case.

Conflict of interest

There is no conflict of interest.

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Nil

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. The presented patient have given their written informed consent for publication of data of this patient.

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