



Extramedullary Plasmacytoma of the Jugular Bulb- A Rare Case Report

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Abstract

Introduction: Extramedullary plasmacytoma (EMP) of jugular bulb is a very rare plasma cell proliferative disorder arising outside the bone marrow. EMP represents less than 1% of head and neck malignancies. More than 90% of EMPs are diagnosed in the head and neck with the soft tissues and upper aero digestive tract being the most common sites. EMPs should be distinguished from other plasma cell dyscrasias for prognosis and treatment. There are very scarce previous documented reports on plasmacytoma arising from jugular bulb. Hence, we present here a rare case report of EMP arising from the jugular bulb masquerading as an extradural space occupying lesion

Case Report: A 56 year old man presented to our hospital with history of neck stiffness, torticollis and progressive nuchal pain since 3 months. Radiological findings identified an extradural expansile mass lesion in the left jugular foramen bulb causing bony erosion. Intraoperative squash cytology suggested a differential diagnosis of small round blue cell tumor. Surgical excision of the mass was performed and sent for histopathological evaluation, which showed features of plasmacytoma. Immunohistochemistry was positive for CD 138 confirming the diagnosis of EMP.

Keywords: Extramedullary Plasmacytoma, Squash Cytology, IHC.

Introduction

EMP is an extremely rare B-lymphocytic plasma cell dyscrasia originating from soft tissue accounting for <1% of all head and neck malignancies which was first described by Schridde in 1905.^[1,2] Plasma cell neoplasms can present as a solitary plasmacytoma or multiple lesions can present as a bone lesion or soft tissue lesion.^[3] It is known that SPB and EMP arise from the analogous cell types confined to a single area,

the former has high tendency for multiple myeloma (MM), while the latter consist of plasma cell infiltration with no sign of MM and hence, the pathogenesis of these two diseases are not alike.^[4] EMP represent for 3% of all plasma cell neoplasia. Plasmacytomas are more common in males, with a male-to-female ratio of 3:1 occurring in fourth to sixth decades in life.^[5] Almost 80% of EMP occur in submucosal lymphoid tissue in head and neck, commonly

affecting the nasal cavity, paranasal sinus, tonsillar fossa and oral cavity but may also occur in the gastrointestinal tract, urinary bladder, gland, lymph node and skin.^[6]

Here, we present a rare case of EMP arising from the jugular bulb masquerading as an extradural space occupying lesion.

Case Report

A 56 year old man, farmer by occupation, visited our hospital with chief complaints of torticollis (positive towards the left side), neck stiffness and nuchal pain since 3 months. The neck pain and stiffness was gradually progressive. There was no history of trauma or any other associated systemic illnesses.

Radiological Investigation with computed tomography (CT) scan of brain revealed a well defined enhancing mass lesion in the left jugular bulb causing bone erosion. (Figure.1a)

Magnetic resonance imaging (MRI) showed a large expansile mass lesion in the left jugular foramen with erosion and widening of the jugular foramen extending to the C2 vertebral body at the anteroinferior aspect (Figure.1b).

A diagnosis of extradural space occupying lesion (SOL) was made and patient was planned for excision of the mass. Intraoperative squash cytology with Hematoxylin and Eosin(H&E), Giemsa and Papanicolaou (PAP) stain was performed (Figure.2a,b,c) which revealed cellular smears of round to oval tumour cells with eccentrically placed nucleus, condensed chromatin & scant cytoplasm arranged in sheets in a haemorrhagic background. The differentials considered were small round blue cell tumor-plasmacytoma, meningioma, and poorly differentiated carcinoma.

The excised SOL specimen was sent for histopathological evaluation.

Macroscopy consistent of multiple grey white to grey brown soft tissue bits approximately amounting to 2 cc.

Microscopy revealed small round to oval cells with mild pleomorphism having eccentric round

to oval nucleus arranged in sheets, cords and clusters separated by thin fibrovascular core. At places many dilated vascular spaces surrounded by these tumor cells with intervening fibrocollagenous tissue was observed (Figure.3). The diagnosis was narrowed down to differentials of Meningioma & Plasmacytoma.

IHC

Immunohistochemistry was strongly positive for CD138 & kappa (Figure.4a&b). Focal positivity was seen for lambda (Figure.4c) which confirmed the monoclonal nature of plasma cells.

Multiple systemic and radiological studies were performed in order to rule out systemic plasmacytoma. Serum protein electrophoresis, beta-2 microglobulins were normal. Urine examination for Bence Jones proteins and for light chain assay was negative. Full blood count and levels of serum glucose, calcium, liver function tests and renal function tests were within normal limits. Bone marrow aspiration from the iliac crest was normal and showed no evidence of plasma cell infiltration. A radiological skeletal survey was performed which showed no evidence of lytic skull lesions in the skull, vertebral column or bones of chest wall or pelvis.

Diagnosis

After ruling out the possibility of Multiple myeloma (MM), a final diagnosis of Extramedullary plasmacytoma of jugular bulb was given.



Fig 1: (a) CT Scan



(b) MRI

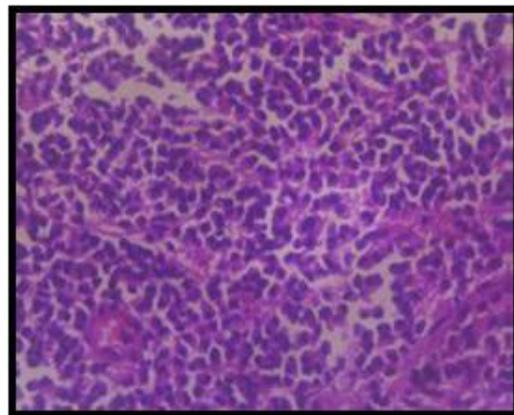


Fig 3: H&E Section

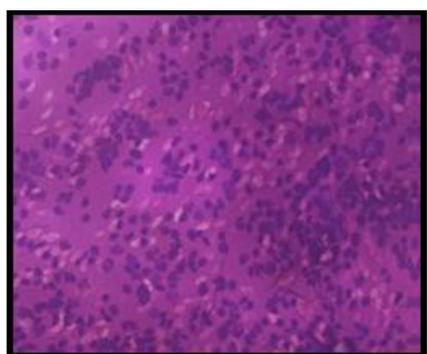


Fig 2 Squash cytology: (a) H&E Stain

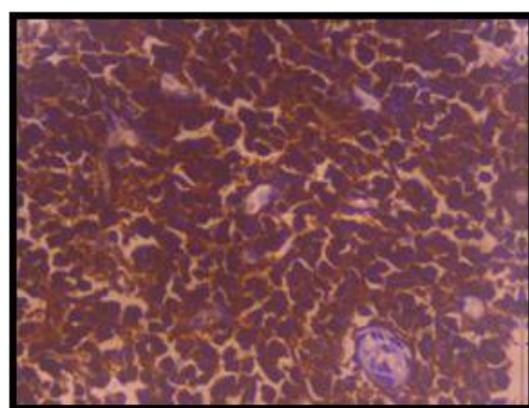
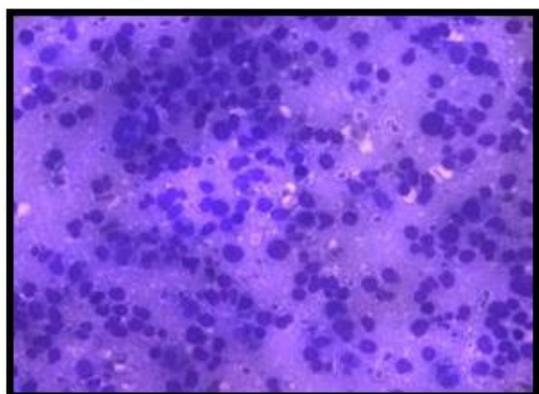
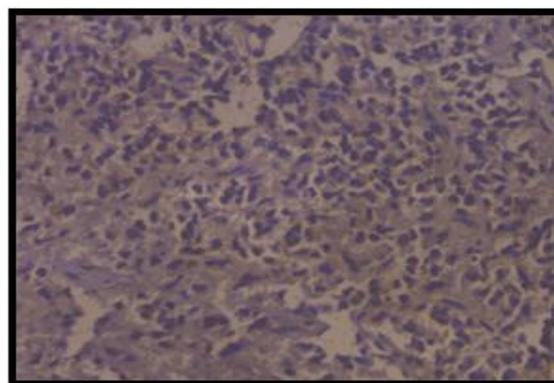


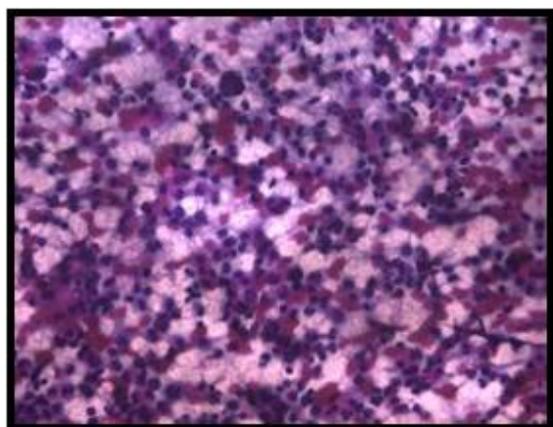
Fig 4 IHC: (a) CD138



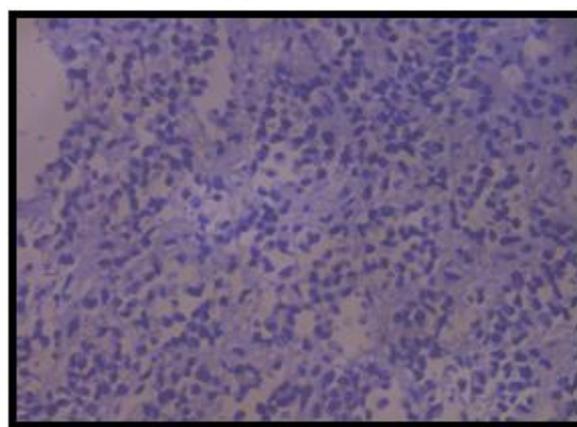
(b) GIEMSA Stain



(b) KAPPA



(c) PAP Stain



(c) LAMBDA

Discussion

Solitary plasmacytoma is a rare neoplasm of monoclonal plasma cells. EMP originates from plasma cells with a single class of heavy and light chains in a monoclonal proliferation of B cells.^[7]

EMPs occur in men approximately three times more than in women.^[2,8] The diagnosis of EMP depends on histopathological examination. The clinical symptoms are usually in relation to the specific location of lesion than to the nature of tumor. We hereby report a case of extramedullary plasmacytoma arising from Jugular bulb, rarely reported in world literature.

EMP of the jugular bulb clinically presented as torticollis and progressive neck stiffness in our case. Other patients can also have cranial nerve palsy and cervical lymphadenopathy.^[7]

No lymph node involvement was found in our patient. The aetiology of this disease remains unknown, but factors such as viral pathogenesis have been previously indicated.^[2]

The recommended diagnostic criteria of EMP of soft tissue are^[6]: (i) Pathological tissue evidence of monoclonal plasma cells involving a single extramedullary site. (ii) no bone marrow involvement (iii) no anemia, hypercalcemia or renal impairment caused by plasma cell dyscrasias (iv) negative skeletal survey results and (v) low serum or urinary levels of monoclonal immunoglobulin.

For diagnosis of an EMP through set of investigations are required. These include blood count serum and urinary protein electrophoresis, peripheral blood smear, renal function test, liver function test, tissue biopsy, radiological survey, CT and MRI scan of the affected area and bone marrow biopsy. Since the morphologic characteristics of EMP may resemble those of extramedullary invasion of a well differentiated MM, the possibility of MM should be omitted before confirming the diagnosis of EMP.

Wiltshaw classified soft tissue plasmacytoma into 3 clinical stages as follows:^[8]

Stage 1	Limited to an extramedullary site
Stage 2	Involvement of regional lymph nodes
Stage 3	Multiple metastasis

The Differential diagnosis for EMP includes Lymphoma, Poorly differentiated carcinoma, Meningioma. Squash cytology with histopathology aids in diagnosis of EMP. Histopathology showing monoclonal plasma cells and Igs on IHC can confirm the diagnosis. CD 138 is a specific marker to confirm the plasmacytic nature of the cells which differentiates it from a carcinoma which stains for CK and CD 40.^[9] Light chain positive for Kappa and focal positive for lambda helps us in determining the process is monoclonal.

The mainstay of treatment for EMP of head and neck as described by United Kingdom Myeloma Forum (UKMF) recommends radiotherapy with surgical excision while EMPs in other sites recommends surgery excision initially.^[10] The most important prognostic factor determining the outcome post therapy is progression to MM. The reported conversion rate of EMP to Multiple Myeloma is 15-30%.^[6]

Conclusion

Extramedullary plasmacytomas are a rare neoplasm of the head and neck but should be included in the differential diagnosis of Extradural space occupying lesions for early detection, favourable prognosis and outcome. Squash cytology and routine histopathological examination helps in detection of these lesions. Immunohistochemistry is of vital significance to establish clonality and neoplastic nature of these tumors and definitive diagnosis.

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