Extramedullary fronto-Orbital plasmacytoma- A rare tumour

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Abstract

Purpose: To report a case of Extramedullary Fronto- orbital plasmacytoma.

Materials and Methods: A 55 year old female presented with swelling in left supraorbital region and ptosis of left eyelid. Mass was smooth, non-tender, non-pulsatile, non-compressible and approximately 3x4 cm in size. Visual acuity was counting fingers. Total excision of tumour was made.

Results: CT scan of brain revealed well defined hyper dense lesion with intense post contrast enhancement in left frontal region causing underlying destruction of left frontal bone, roof of left orbit, and left sphenoid bone, A biopsy was performed revealing that the tumour was composed of plasma cells positive with immunohistochemical stains and epithelial membrane antigen. The diagnosis of extramedullary fronto-orbital plasmacytoma was made.

Conclusion: Extramedullary Plasmacytoma a rare tumour is usually treated by total surgical excision or surgery combined with local radiotherapy and regular long-term follow-ups because disease has high potential to transform into multiple myeloma.

Keywords: Extramedullary Plasmacytoma, Multiple Myeloma, Plasma cells.

Introduction

Plasmacytoma is a neoplasm of proliferated monoclonal plasma cells in either bone or soft tissue and present as a single lesion (solitary plasmacytoma) or as multiple lesions in bone marrow (multiple myeloma). Solitary plasmacytoma most frequently occur in the bone but rarely found in the soft tissue as solitary extramedullary plasmacytoma.[1]

EMPs are categorized as very rare form of non-Hodgkin lymphoma and represent about 3% of all plasma cell tumours. The nasal cavity, tonsils, paranasal sinuses, nasopharynx and trachea are most commonly affected sites. Regional lymph node involvement occur in about 10% to 15% cases.[1]

Median age of diagnosis is 55 to 60 years and has male predominance.

A combination of surgery for resectable tumours and radiotherapy is treatment of choice. Chemotherapy is given in patient's refractory to radiotherapy. Herein we present the case of 55 year old female operated for left fronto-orbital mass presented with chief complain of swelling above left eye and drooping of left eyelid.

Case Report

A 55 year old woman referred to the Neurosurgery department with chief complain of swelling above the left eye and drooping of left eyelid since last 8 months. Apparently 8 months back she first noticed some bulge above her left eye which gradually increase and pushing her left eyelid downwards and creating facial asymmetry. She has no complain of any visual problems. No complain of vomiting, loss of consciousness, convulsions or nasal bleed.

On physical examination swelling in left supraorbital region and ptosis of left eyelid were noticed. Mass was smooth, non-tender, non-pulsatile, non-compressible and approximately 3x4 cm in size. Visual acuity and extra ocular movements were normal. No lymph node swellings were noted on palpation.

Contrast enhanced CT scan of brain revealed 37x38x35mm sized well defined hyper dense lesion with intense post contrast enhancement in left frontal region causing underlying destruction of left frontal bone, roof of left orbit, and left sphenoid bone (Fig. 1). Lesion extends extracranially into soft tissue of left frontal region, extends inferiorly into extraconal compartment of left eyeball and displacing it inferiorly.

The patient underwent left frontal craniotomy with osteoplastic bone flap and excision of tumour slightly extending into left orbit. Involved part of Dura and bone was excised, exposed left frontal sinus was packed with muscle and sealed with bone wax to prevent post-operative CSF rhinorrhoea and watertight duraplasty was done. Postoperative CT scan was done (Fig. 2). Tumour was completely excised and sent for histopathology and confirmed the diagnosis of plasmacytoma (Fig. 3). Postoperatively patient was better and improved in her complain of facial swelling and eyelid drooping. She was discharged after suture removal and advised for regular follow-up.

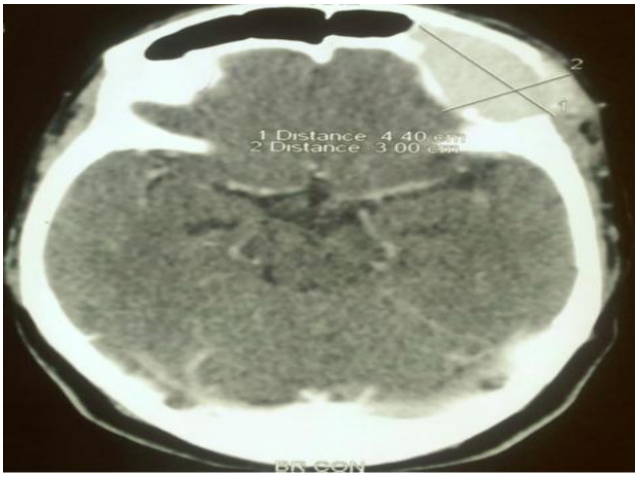

Fig. 1: CT scan displaying hyperdense lesion with extension into orbit.

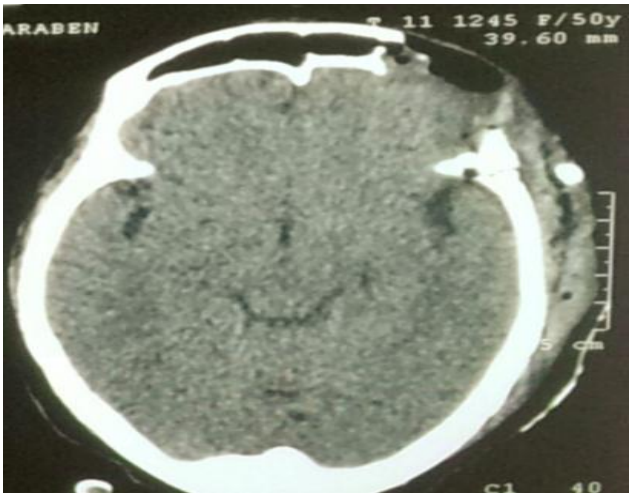

Fig. 2: CT scan displaying postoperative completely excised lesion.

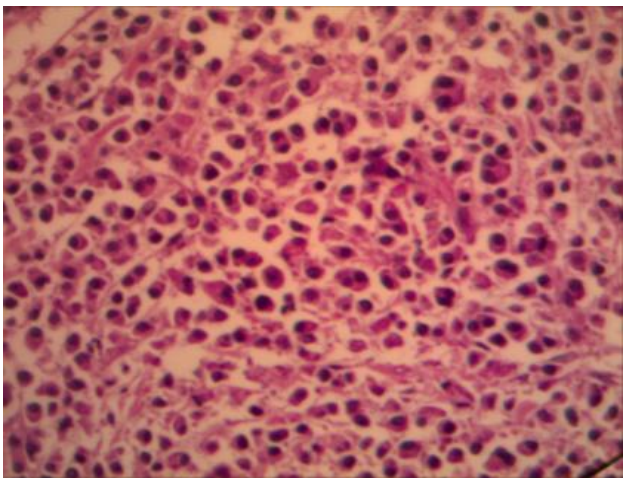

Fig. 3: Light microscopy showing a dense and monomorphic proliferation of atypical plasma cells with sparse stroma (Haematoxylin and Eosin. Total Magnification: ×100).

Discussion

Extra medullary Plasmacytoma (EMP) are seen in patients with age range of 17 to 80 years with male preponderance. Clinical presentation of EMP depends on the mass volume and site of involvement. Nasal obstruction, soft tissue swelling, epistaxis, proptosis, ptosis, diplopia and visual disturbances are main clinical findings observed.

Tumour has slow growing nature and become symptomatic when mass fills cavity several months later.

This condition may results in treatment delay and transformation into systemic disease (multiple myeloma). Early referral of this patient was due to obvious symptoms due to mass effect and pushing of eyeball down and ptosis.

CT and MRI are complementary techniques in evaluating the size, location and involvement of adjacent structures; however, the role of MRI in staging EMPs is not clear.Although non-specific, CT and MRI images show lobular soft tissue masses or infiltrative lesions with contrast enhancement. Bony destruction is displayed depending on expansion of tumour.

In our case CT scan was useful in demonstrating bony abnormalities and erosion along with the lesion. MRI was superior to CT scan in defining character of tumour but can't distinguish plasmacytoma from other causes like meningioma. Diagnosis of EMP depends on morphologic and immunophenotypic analysis of localised tumoral mass.

In our case microscopic description of tumour mass revealed sheets of plasma cells including few immature forms with focal perivascular amyloid deposition. Immunophenotyping with CD15, CD20, CD45RB, CD45RA, epithelial membrane, IgA, IgD, IgM, IgG, IgE, kappa light chain and lambda light chain antibodies are helpful in differentiating EMP from epithelial tumours. Once the diagnosis was confirmed by histopathological analysis patient was referred to haematologist. Based on well-known radio sensitivity of plasma cells, a dose of 40-50 Gy radiotherapy is advised in the treatment.

According to Alexiou et al., EMPs with bone destruction should not be treated with radiotherapy or surgery alone, but with rather combination of both modalities to ensure local and systemic control.

Galieni et al., reported surgery in case of easily resectable masses would be adequate to treat the disease without recurrence. They emphasized presence of monoclonal component at diagnosis was a poor prognostic factor.[2]

EMP demonstrates relatively low risk of progression to multiple myeloma (MM). Survival rate was observed of 70% in EMP cases over duration of 10 years.

Another study conducted by kilciksiz s et al in 2008, showed that Younger age group was to be an independent good prognostic factor predicting a lower rate of progression to MM.[3] Tumour size <5 cm, low M protein levels, patient age <40 years, absence of light chains, and disappearance of M protein after treatment were found to be good prognostic factors. [Hotz MA et al in 1999, Liu HY et al in 2010].[4,5]

Activation of bone morphogenetic protein 6 expression is independent favourable prognostic factor. In the case of persistent M protein after completion of radiotherapy or suppression of normal immunoglobulin classes and suppression of BMP6 expression by intensive methylation of genes at CpG sites of the BMP6 promoter region by plasma cells, with extremely high levels of serum lactate dehydrogenase, early intensive systemic therapy should be considered to prevent poor prognosis.[1]

In summary, solitary EMP usually develops in the head and neck soft tissues and also involve sites such as the thyroid and brain and may be observed in a large age range (17-80 years). Older Patients with symptoms of nasal blockage, facial mass or epistaxis should be considered for possibility of plasma cell tumour. This neoplasm is usually treated with combination of surgery and radiotherapy. After 3 to 8 years of treatment progression of MM may occur. Long-term follow-up in terms of disease recurrence and progression is mandatory.

Conflicts of Interest: None.

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