



Intracranial Plasmacytoma with Unilateral Vision Loss: An Unusual Initial Presentation of Multiple Myeloma

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ABSTRACT

Plasma cell neoplasms are clonal proliferation of plasma cells that produce a single class of immunoglobulin. Among the plasma cell neoplasms, both solitary plasmacytoma of bone and extra-osseous plasmacytoma can progress to multiple myeloma. Here, we described a rare case of multiple myeloma with initial presentation as plasmacytoma with both intracranial and extracranial component, sphenoid sinus involvement and left sided vision loss. Serum protein electrophoresis revealed presence of M spike. Histopathological and immunohistochemical analysis detected plasmacytoma. Extramedullary plasmacytoma is a rare presentation of multiple myeloma. Intracranial involvement and involvement of sphenoid sinus of plasmacytoma with pressure effect mimicking other intracranial neoplasm is extremely rare. Clinicians should consider this entity in any solitary intracranial neoplasm with rapid progression of clinical course.

Keywords: Multiple myeloma, Intracranial plasmacytoma, Sphenoid sinus, Unusual presentation.

Introduction

Plasma cell neoplasms are clonal proliferation of plasma cells that produce a single class of immunoglobulin. According to the World Health Organisation 2016 classification of lymphoid neoplasms, plasma cells tumours are classified into plasma cell myeloma (multiple myeloma), solitary plasmacytoma of bone, and extraosseous plasmacytoma.^[1] Extramedullary plasmacytoma are rare and accounts for 3% of plasma cell tumours.^[2] Very infrequently intracranial plasmacytoma with sphenoid sinus involvement become initial presentation of multiple myeloma. Here, we report on a 52-year-old woman, who presented with left-sided forehead swelling and

loss of vision in left eye caused by intracranial plasmacytoma.

Case Report

A 52 years old female patient presented to our hospital with headache, vertigo, gradually increasing swelling over left side of forehead and loss of vision with proptosis of left eye ball for last 3 & ½ months. Except Hepatitis B infection, she had no suggestive medical history. Routine blood analysis revealed Hb% 4.6g/dl, ESR 170 mm in 1st hour, blood urea 157 mg/dl, serum creatinine 3.9 mg/dl and serum calcium 11.5mg/dl. Serum protein electrophoresis shows presence of M spike.

CT scan noted a hyperdense soft-tissue mass measuring 67 mm X 43 mm in the frontal lobe eroding the left frontal bone, left supra-orbital ridge and extending to left orbit. Another hyperdense lesion measuring 29 mm X 21 mm also noted in suprasellar region involving diaphragmasella. CT scan gives the diagnosis of meningioma or lymphoma.

MRI revealed a large, lobulated margined lesion measuring about 47mm X 56mm X 71mm in left frontal region having intracranial and large extra-cranial component with associated destruction of adjacent calvarial bones and extending inferiorly into superior part of adjacent orbit. Another lesion measuring about 44 mm X 36 mm X 31 mm with intracranial extension into adjacent sphenoid sinus seen along floor of anterior cranial fossa and adjacent sellar and suprasellar regions with associated bone destruction. The buckling of brain seen in left frontal lobe with effaced adjacent sulci and mild contra- lateral shift of anterior midline. MRI gives the diagnosis of invasive carpet meningioma.

The tumour has been excised completely. During operation, it was found that the tumour is highly vascular and has destroyed the left frontal and orbital wall. Tumour not affected the dura.

Histopathological study revealed, the tumour composed of the mature and immature plasma cells arranged in diffuse sheets and cords. Individual cells have distinct cellular outline with large eccentrically placed nuclei, sometimes multinucleated, containing diffuse or clumped chromatin at periphery giving rise to characteristic cart-wheel appearance, often with prominent nucleoli and perinuclear halo.

Immunostaining revealed strong granular cytoplasmic positivity for CD 138. But the expression of CD 20, MUM 1 and CD 56 are negative.

From the histopathological study, Immunohistochemical study, routine blood tests a diagnosis of multiple myeloma presenting as intracranial plasmacytoma with sphenoid sinus involvement is made. Immunofixation study and Bone marrow

aspiration could not be done as the patient expired within three days after diagnosis.



Fig. 1. CT scan showing a large soft tissue mass eroding frontal bone and greater wing of sphenoid and affecting Left eye ball

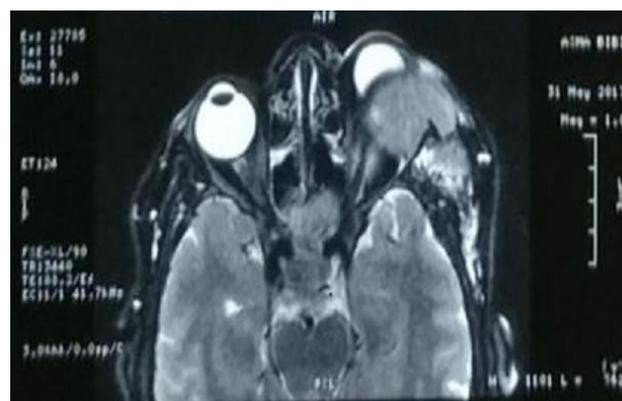


Fig.2. MRI showing tumour mass in left frontal region and over sphenoid sinus

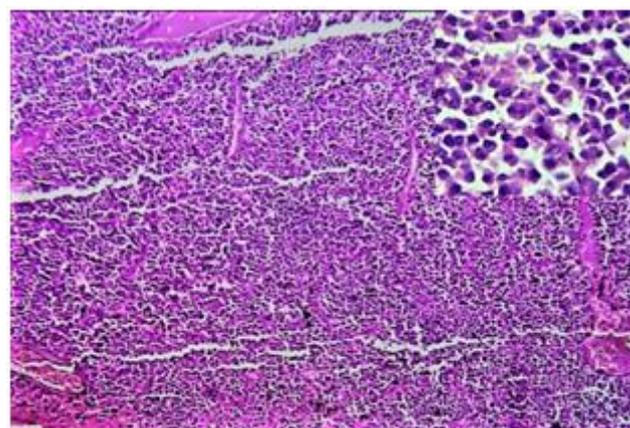


Fig.3. HPE showing Plasmacytoma low power (10X) and high power (40X)(inset)

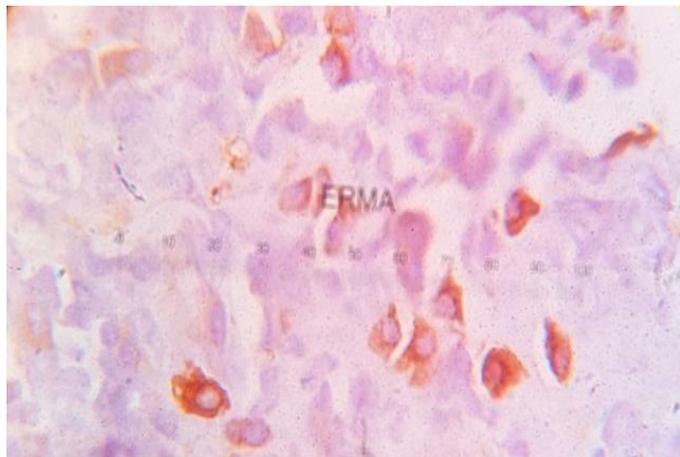


Figure 4: Plasmacytoma showing CD 138 positive cells

Discussion

Plasma cell neoplasm are more common in the 4th and 7th decades with male to female ratio 1.3:1. Extramedullary plasmacytoma is an infrequent initial presentation of multiple myeloma. In a series of 822 patients with plasma cell neoplasm, only 3% had Extramedullary plasmacytoma as their initial presentation.^[3] A plasmacytoma with both intracranial and extracranial component along with sphenoid sinus involvement is extremely rare as initial presenting symptoms, as our case.

During normal B cell development, cells acquire expression of CD138, a heparin sulphate proteoglycan, which is highly specific for terminally differentiated normal plasma cells as well as multiple myeloma cells.^[4] CD138 cells control tumor survival, growth, adhesion and bone-cell differentiation in multiple myeloma.^[5] CD56 is a neural adhesion molecule that expressed in 70% - 80% cases of multiple myeloma.

CD 56 negative patients had higher β 2-microglobulin levels and a higher incidence of Extramedullary disease, Bence

Jones protein, renal insufficiency and thrombocytopenia than CD 56 positive patients.^[6] Multiple myeloma patients with CD 56 negative plasma cells have more aggressive disease and a poor prognosis.^[7]

CD 20 is a transmembrane phosphoprotein that acts as a calcium ion channel in the cell

membrane and retained on mature B cells until differentiated into plasma cell. Loss of CD 20 expression during the disease course correlates with significant worsening of overall survival and event-free survival.^[8]

MUM 1 is a transcriptional factor and a nuclear stain which is used to differentiate germinal center large B cell lymphoma from non-germinal center phenotype.

Sphenoid sinus occupies the body of the sphenoid and is surrounded by various important structures like, olfactory tract, optic chiasma, optic, abducent and trigeminal nerves and internal carotid artery. This anatomic location largely accounts for the wide diversity of symptoms and difficulty in early diagnosis when sphenoid sinus is involved.^[9] By the time of diagnosis, the disease advances much.

Due to rarity of presentation, there is no fixed treatment protocol and treatment modalities vary amongst institution. A multidisciplinary approach consisting of Chemotherapy, Radiotherapy, Immunotherapy and Stem-cell transplantation has been considered important for overall improvement of survival. The overall prognosis of multiple myeloma depends on a variety of factors including patient's performance status, renal function, tumour burden and tumour biology.^[10]

In conclusion, intracranial plasmacytoma with sphenoid sinus involvement is a rare initial presentation of multiple myeloma and the early diagnosis is challenging unless there is a strong clinical suspicion. The clinicians should keep in mind this clinical entity while dealing with a patient with intracranial tumor.

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