CASE REPORT

EXTRA OSSEOUS MULTIPLE MYELOMA DEPOSITS IN ORBITS AND PARANASAL SINUSES; MASQUERADING AS FUNGAL INFECTION ON MRI; A RARE PRESENTATION

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PJR July - September 2020; 30(3): 209-213

ABSTRACT

The extraosseous soft tissue deposits of multiple myeloma is very rare. The present report describes a case of 63 years old female , known case of multiple myeloma, on treatment, now consulted for left eye ptosis and visual loss. In this case report, we discuss the MRI findings of multiple myeloma in orbits and paranasal sinuses. The purpose of this report is to create awareness among radiologist about extraosseous presentation of multiple myeloma. There is paucity of locally published literature on extra osseous multiple myeloma deposits in orbits and paranasal sinuses.

Keywords: Multiple myeloma, extraosseous metastasis, orbital metastases, paranasal sinus metastases,, multiple myeloma metastases.

Introduction

Multiple myeloma is a systemic malignant disease of blood and accounts for 1% of all cancers and approximately 10% of all hematologic malignancies. It is slightly more common in men than women.it is characterized by malignant proliferation of monoclonal plasma cells in bone marrow leading to production of non-functional intact immunoglobulin or immunoglobulin chains. Important advances have now been evolved in diagnosis and staging and offer better treatment of multiple myeloma and increasing the overall survival of these patients. There has been a concurrent increase in unusual manifestation of multiple myeloma especially relapses or extramedullary disease. Approximately 1 to 2% of patients have extraosseous disease at the time of diagnosis while 8% develop later in the course of disease. Although imaging characteristics of multiple myeloma are non-specific, the presence of extraosseous soft tissue masses in patients with multiple myeloma should raise suspicion of extra osseous disease.

Case Report

A 63 years old female, k/c of multiple myeloma was presented to accident and emergency department of Liaquat national hospital Karachi with left eye ptosis for 3 months and vision loss for 4 days. Previously she was diagnosed as fungal infection in orbits and sinuses on MR imaging characteristics and had been treated with antifungals for presumed fungal infection, however she experienced detoriation of vision in left eye for which she had her biopsy done from maxillary sinus which showed respiratory mucosa fragments with underlying tissue showing infiltration by diffuse sheets and cohesive proliferation of cells. These cells

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Submitted 20 August 2020, Accepted 16 September 2020

are large in size with enlarged centric to eccentrically placed nuclei, prominent nucleoli and frequent mitosis. Proliferative index was high. The differential diagnosis of plasma cell dyscrasia (including plasma cell myeloma, plasmablastic or anaplastic type) and plasmablastic lymphoma were considered.

MRI revealed heterogeneous signal intensity mass in retro bulbar region of left orbit causing significant ptosis. It is extending from optic foramen to left parasellar region. It is abutting medial, inferior, and lateral recti muscles with loss of fat planes and encasing optic nerve. Anteriorly it is abutting posterior sclera , inferiorly it is extending in to maxillary antrum. It appears hypo intense on T1, intermediate on T2 and showed intense post contrast enhancement.

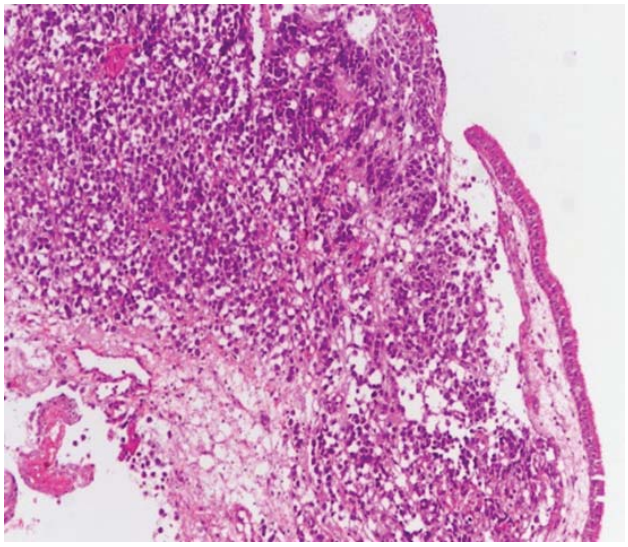

Figure 1A: Cohesive clusters and sheets of tumor cells with overlying respiratory mucosa.

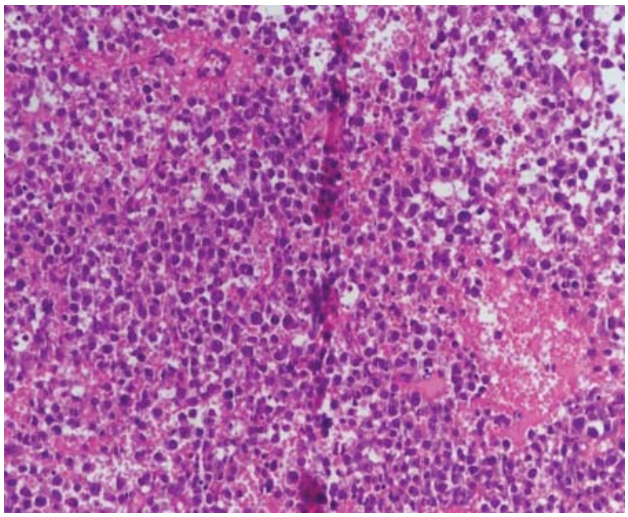

Figure 1B: Sheets of tumor cells with centric to eccentric nulei.

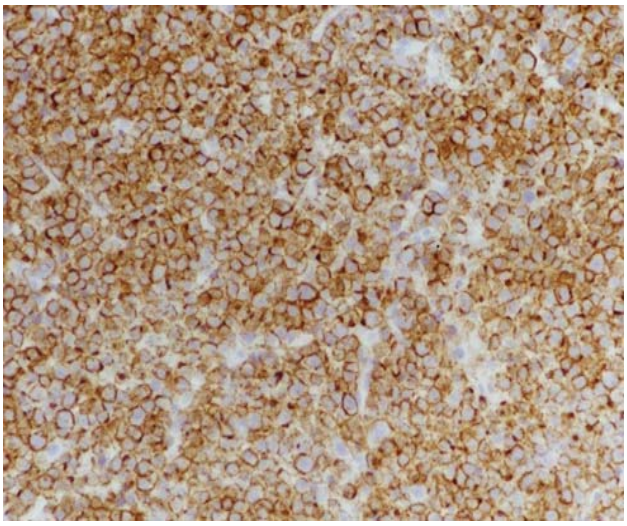

Figure 1C: Immunohistochemical marker CD138 diffusely positive in tumor cells

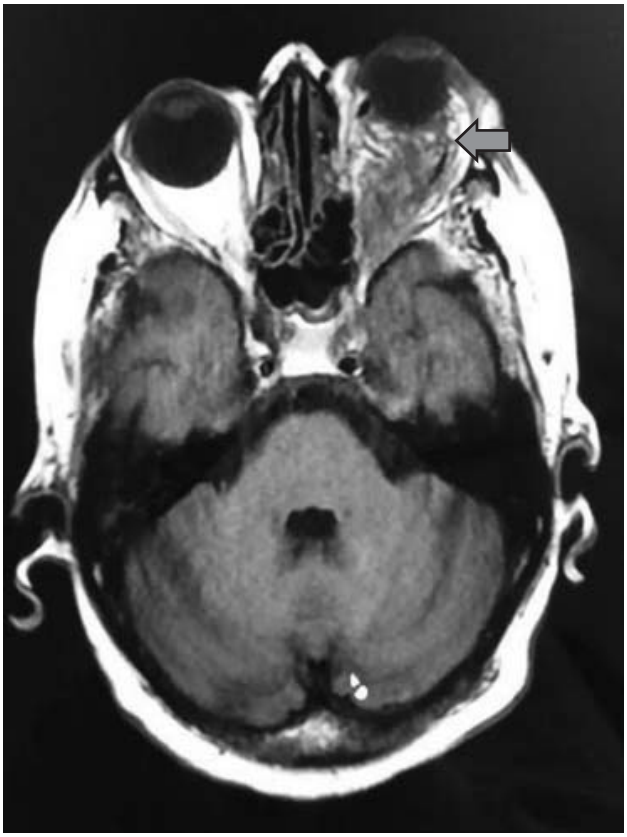

Figure 2A: Mass in retro bulbar region appears hypo intense on T1

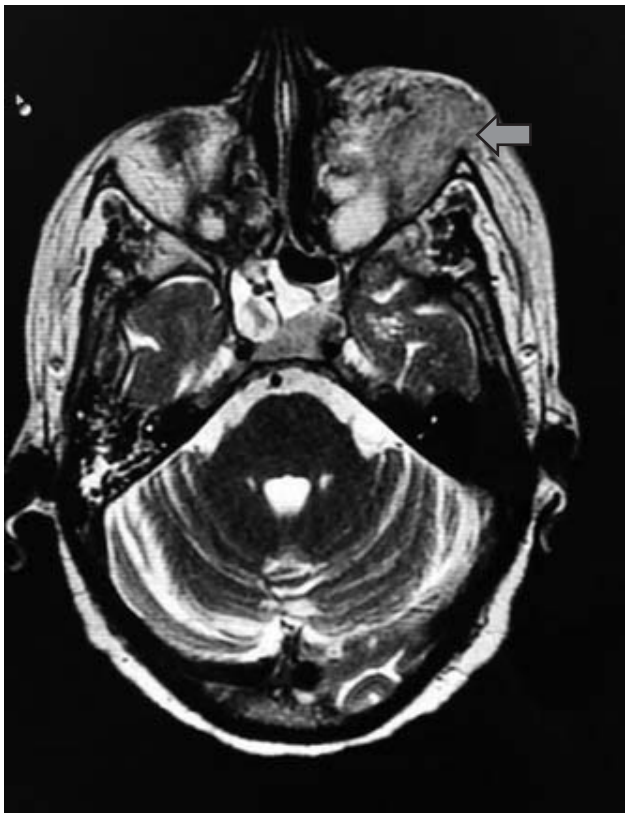

Figure 2B: It appears intermediate on T2WI

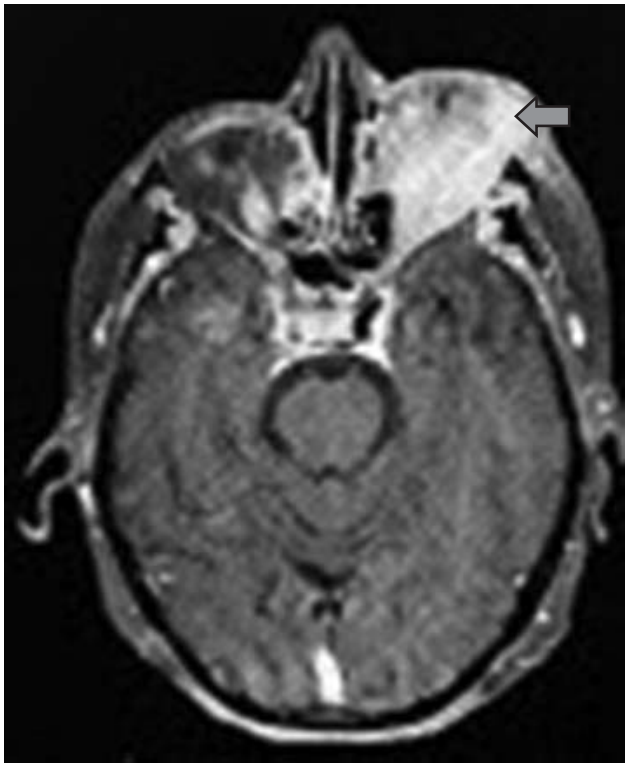

Figure 2C: It shows intense Post Contrast enhancement.

There is another similar signal intensity mass noted in right maxillary sinus extending inferiorly into hard palate. All these findings represent multiple myeloma deposits.

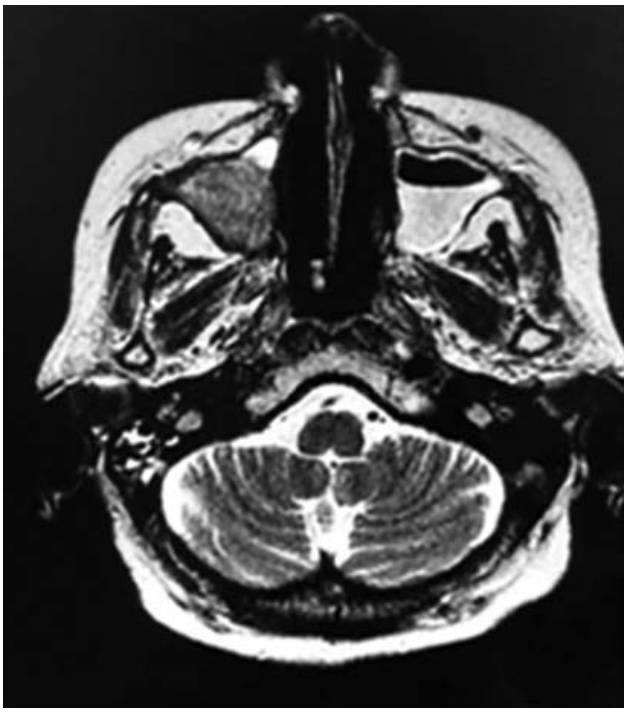

Figure 3A: Hypointense mass in right maxillary sinus on T2WI while air fluid level seen in left maxillary sinus likely due to sinusitis.

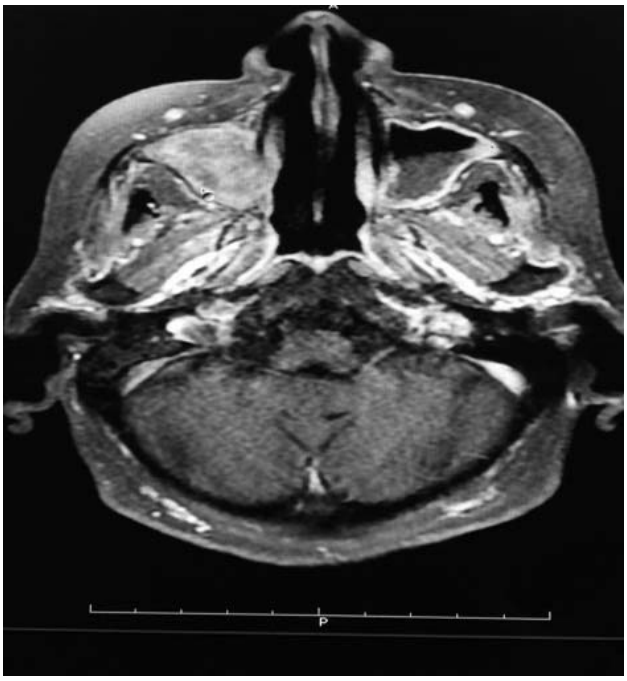

Figure 3B: Mass in right maxillary sinus shows enhancement on post contrast images.

Discussion

Multiple myeloma is a haematological malignancy characterised by monoclonal proliferation of plasma cells in the bone marrow.[1]

It manifests in a wide range of radiographic abnormalities.[1,2]

Bone disease is the main cause of morbidity and can be detected on skeletal survey. Although not standard, however additional imaging modalities include low dose whole body computed tomography, magnetic resonance imaging or flouro deoxyglucose positron emission tomography.[2,3] Other major clinical manifestations are anemia, hypercalcemia, renal failure, and increased risk of infection. Contiguous spread of disease is also common more than non-contiguous extra osseous spread, However in patients with known multiple myeloma, soft tissue deposits should be taken as high suspicion of extra medullary disease of multiple myeloma.[3,4]

Extra osseous deposits are occasionally found, amongst them most commonly seen in liver, spleen, lymph nodes, lungs, oropharynx.[3] The extramedullary plasmacytosis of the head and neck accounts for only 3% of all the plasma cell tumors.[3] It occurs more frequently in the upper respiratory tract, particularly in the nasal cavity and paranasal sinuses,[3] however orbital involvement is very rare.[4] Only few cases with orbital involvement have been reported.[4,5] Almost any structure in the orbit can be involved. Most common presentation include slowly progressive painful exophthalmos, diplopia and decrease in visual acuity. In our case patient presented with ptosis and vision loss.Orbital deposits in particular have worse survival rates compared to other extramedullary plasmacytomas.[4,5] It can also lead to intracranial extension.[5]

Examination of the skeletal system (either a positron emission tomography/computed tomography (PET/CT) scan or magnetic resonance imaging (MRI) of the entire spine and pelvis), and bone marrow study are necessary to establish a proper diagnosis.[6,7]

Due to superior soft tissue contrast, MRI is considered as best imaging modality to detect marrow signal changes in multiple myeloma before osseous destruction is seen at CT. It is recommended that more than one focal lesion >5mm should be considered a myeloma-defining event that would require therapy.[9]

Most extramedullary plasmacytomas are iso/hypointense to skeletal muscle on T1 and iso/hyperintense on T2-weighted image, with mild-to-moderate enhancement following intravenous gadolinium administration. Diffusion weight imaging demonstrates restricted diffusion, indicating high cellularity of the lesions.[10] Some extramedullary plasmacytomas have been described to demonstrate low T2 signal intensity.[10]

Our case found similar results with extramedullary plasmacytomas in orbits and paranasal sinuses demonstrating iso/hypointensity to skeletal muscle on T1 and mild-to-moderate hyperintensity on T2-weighted image with enhancement.[10]

Conclusion

Extraosseous metastasis of multiple myeloma are unusual, however it should be kept in mind and suspicion should raised under appropraite clinical conditions to aviod any possible catastrophy.

Conflict of Interest: None

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