Multiple Myeloma Presenting As Primary Orbital Tumour – A Case Report

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Introduction

Multiple myeloma and its related plasma cell dyscrasias are characterized by an abnormal proliferation of a single clone of highly specialized lymphocytes engaged in the production of a specific immunoglobulin.

Mean age at presentation is between 40-70 yrs with peak in the 7th decade.

It accounts for about 10% of all haematological malignancies with an incidence of about 5.5 cases per 100,000 population.

Orbital involvement as a manifestation of multiple myeloma is very rare. It accounts for 0.1 – 0.5 % of all orbital tumours. Less than 50 cases of such involvement has been described in literature.

Recently we came across a case of multiple myeloma with orbital involvement.

Case Report

A 50 yrs old female presented in our OP with a history of painless, progressive drooping of her (R) upper lid of 2 ½ months duration and a swelling in the superolateral part of (R) orbit of 2 months duration (Fig.1). The swelling was painless, gradually progressive and no relation with posture or coughing. There was no history of diplopia, fever, weight loss, or trauma. She had chronic low back ache since 15 yrs and has been on treatment with analgesics. She also had a thyroid nodule excision at the age of 13 yrs, the details of which were not available.

Ocular Examination

Examination of the right eye showed an eccentric proptosis with a forward, downward and inward displacement of the eye ball. The proptosis was due to a swelling of about 8x5 cm over the superolateral part of (R) orbit (Fig.2). The skin over the swelling was normal. The swelling was firm, non tender, non pulsatile and non compressible. The posterior border could not be palpated and the overlying bone was eroded.

Elevation of the (R) eye ball was restricted minimally. Visual acuity and tension was normal and the pupil reacted briskly to light. Fundus examination showed choroidal folds. Left eye was within normal limits.

System examination revealed no abnormalities except for a scar over the front of neck (thyroid nodule excision) and a tenderness over L4-L5 region of spine.

Based on the clinical findings, a diagnostic possibility of a malignant tumour of the lacrimal gland or a metastasis from a primary (possible thyroid) was made. A CT scan of the head and orbit was ordered, which revealed an enlarged (R) lacrimal gland, destruction of the orbital plate of frontal bone with intracranial and scalp extension and compression upon the globe. Multiple lytic lesions in the skull were also seen suggesting the possibility of a malignant tumour of the lacrimal gland or metastasis (Fig. 3 & 4).
Fig. 1. Swelling in the superolateral part of the orbit displacing the globe

Fig. 2. Lacrimal gland pushed downwards

Fig. 3 & 4. CT showing swelling with overlying bone erosion

Fig. 5 & 6. Digital X-ray skull showing multiple punched out lytic lesions

Fig. 7. FNAC from the swelling showing plasma cells

Fig. 8. Bone marrow trephine biopsy showing plasma cells
Plain X-ray skull was taken which showed multiple, punched out, lytic lesions (Fig.5 & 6). Thus a differential diagnosis of multiple myeloma was also made.

FNAC from the swelling was performed using a 23 G needle. The aspirate was haemorrhagic. Microscopic examination revealed polygonal cells with moderate cytoplasm and a single, large eccentric nucleus (Fig.7).

A cytologic diagnosis of plasmacytoma was thus given and a detailed work up was advised to rule out systemic involvement.

Thus haematological investigations like ESR was repeated which showed 90 mm/ 1\textsuperscript{st} hr. (Initially – 49 mm/hr).

Hb was 12.4 g%

LFT, RFT, S.Calcium, S. Phosphorous were within normal limits. Urine Bence Jones Proteins was absent on repeated examination and serum electrophoresis was negative for myeloma proteins.

Plain X-ray pelvis showed sclerosis of L4-L5 area. The patient was then transferred to the haematology department where she underwent a bone marrow trephine biopsy. It revealed infiltration by group of plasma cells comprising about 20 % of the cell group (Fig.8).

A final diagnosis of multiple myeloma manifesting as orbital involvement at presentation was thus made.

The patient was put on CDT (Cyclophosphamide, Dexamethasone and Thalidomide) regime. She responded fairly well to the treatment.

Discussion

Orbital involvement in multiple myeloma is a very rare finding. Less than 50 cases of such involvement is described in the literature. Orbital involvement can occur in one of the following ways.

a. As a part of systemic multiple myeloma with local bone destruction from an isolated plasmacytoma.

b. Extramedullary plasmacytoma from orbital soft tissue.

c. Secondary extension to the orbit of a sinus.

In most cases, the onset is insidious with slowly progressing proptosis accompanied by pain, diplopia and visual impairment. Intracranial extension can lead to papilloedema and cranial nerve palsies. Other ocular manifestations include conjunctival and corneal crystalline deposits, scleritis, episcleritis, secondary glaucoma, uveal plasmacytoma, hyperviscosity retinopathy, retinal vasculitis etc.

Non specific symptoms like low grade fever, malaise, anorexia etc are common. Careful examination and performing the relevant investigations help in reaching the diagnosis earlier.

Imaging features of orbital multiple myeloma have been described in few cases. CT shows thinning of the overlying bone or causing marked bony expansion and destruction. MRI shows low signals on T1 weighted images and high signals in T2 images.

Since the clinical outcome is significantly worse in patients with systemic involvement as compared to those with solitary plasmacytoma an early cytologic diagnosis of extramedullary involvement in multiple myeloma helps in timely institution of appropriate treatment.

References


