

Case Report

Unilateral Proptosis as the initial presentation of Multiple Myeloma

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ABSTRACT

A 75 year lady presented with the complaints of double vision on up gaze since 1 year and gradually progressive painless protrusion of the left eye since 2 months. There was a history of repeated admission to the hospital in past 1 year for weakness. On systemic examination patient had pallor, pedal edema and a borderline splenomegaly. Ocular examination revealed eccentric proptosis. CT scan of head and orbit showed an isointense ill-defined mass in superior antero-lateral extraconal space extending to the medial side which was eroding the fronto-zygomatic bones. Fine Needle aspiration cytology showed binucleate plasma cells and plasma blasts suggestive of multiple myeloma. Serum electrophoresis showed 2 abnormal bands of paraproteins in the gamma region.

Though uncommon, proptosis could be the initial presentation of multiple myeloma. A high clinical suspicion helps us to make a diagnosis of multiple myeloma. Early diagnosis and timely treatment would help to prolong the patient's life and to improve quality of life.

Key words: Multiple myeloma, Unilateral proptosis, Plasma cells

INTRODUCTION

Multiple myeloma is a type of plasma cell dyscrasias of the immune system, characterized by neoplastic proliferation of plasma cells or their precursors. Multiple myeloma can involve practically every ocular structure. Ocular findings may be the first manifestations of the disease.^[1] Cysts of the ciliary body have been reported in 33-50% and retinal vascular lesions up to 66% of myeloma patient.^[2] Corneal and orbital

involvement is less common. Unilateral proptosis is a well-recognized, presentation of multiple myeloma as unilateral proptosis is a well recognized but rare presentation in ophthalmic practise. Proptosis may be either the initial manifestation of the disease or occur as a terminal event also it may alert us about insufficient chemotherapy.^[3]

CASE REPORT

A 75 years old female presented to the department of Ophthalmology, M.S Ramaiah Medical College, Bangalore with the complaints of double vision on up gaze since 1 year and protrusion of the left eye since 2 months. The protrusion was gradually progressive and painless There was no history of defective vision, head ache and trauma. There was a history of loss of appetite since one year. There were repeated admission to the hospital in the

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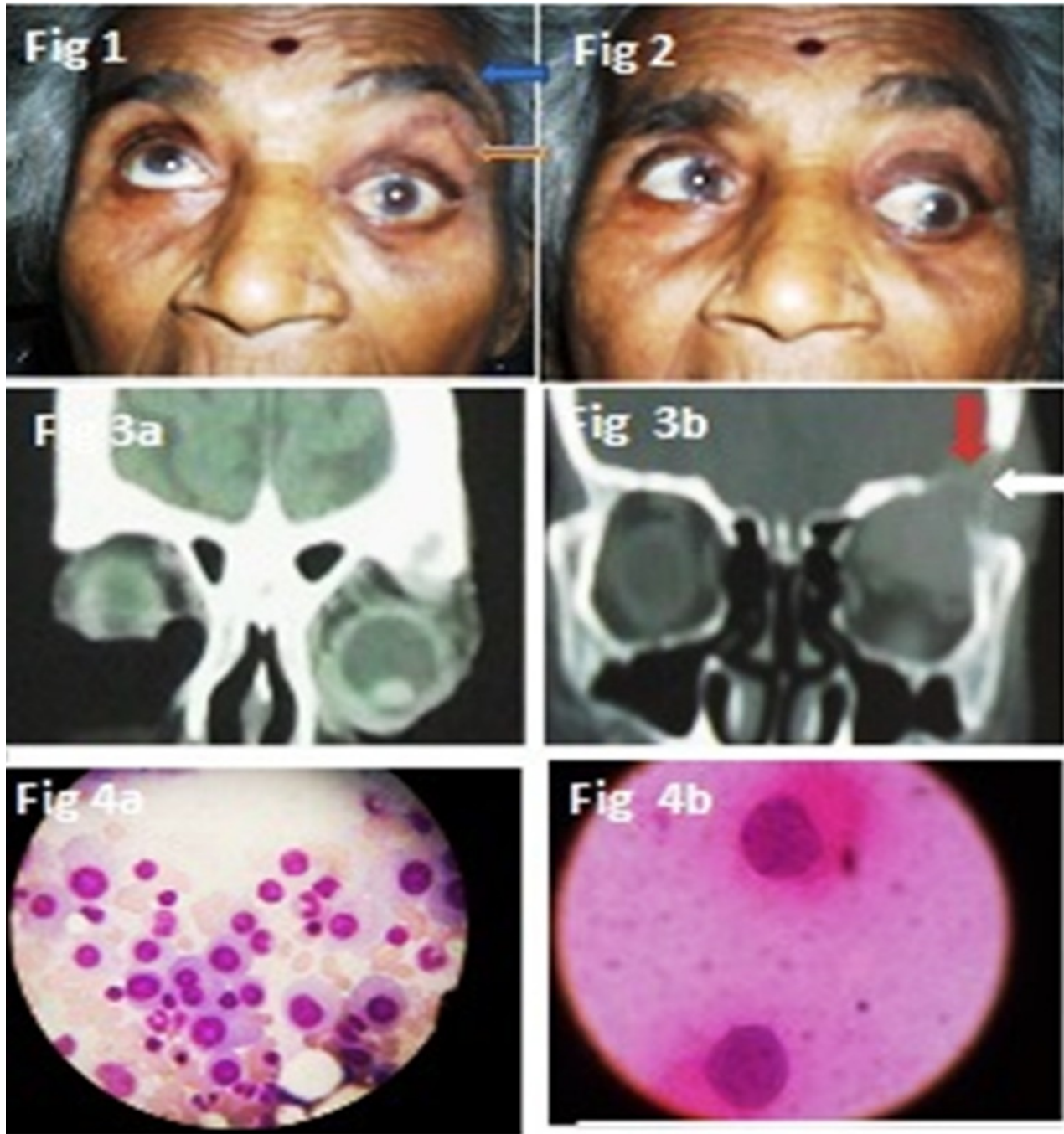


Fig. 1 & 2: Clinical picture showing gross restriction in elevation and abduction. Note the temporal fullness (Blue arrow) and the fullness of upper lid (Orange arrow)

Fig. 3a & 3b: CT scan of Orbit shows an isointense ill-defined mass in superior antero-lateral extraconal space extending to the medial side which was extending from the frontal and zygomatic bones. The bone window pictures show that the mass was extending into the temporal region (white arrow) and also into the brain (red arrow) with evidence of bone destruction.

Fig. 4: Fine Needle aspiration cytology shows cells with high nuclear to cytoplasmic ratio and a few binucleate plasma cells and plasmablasts.

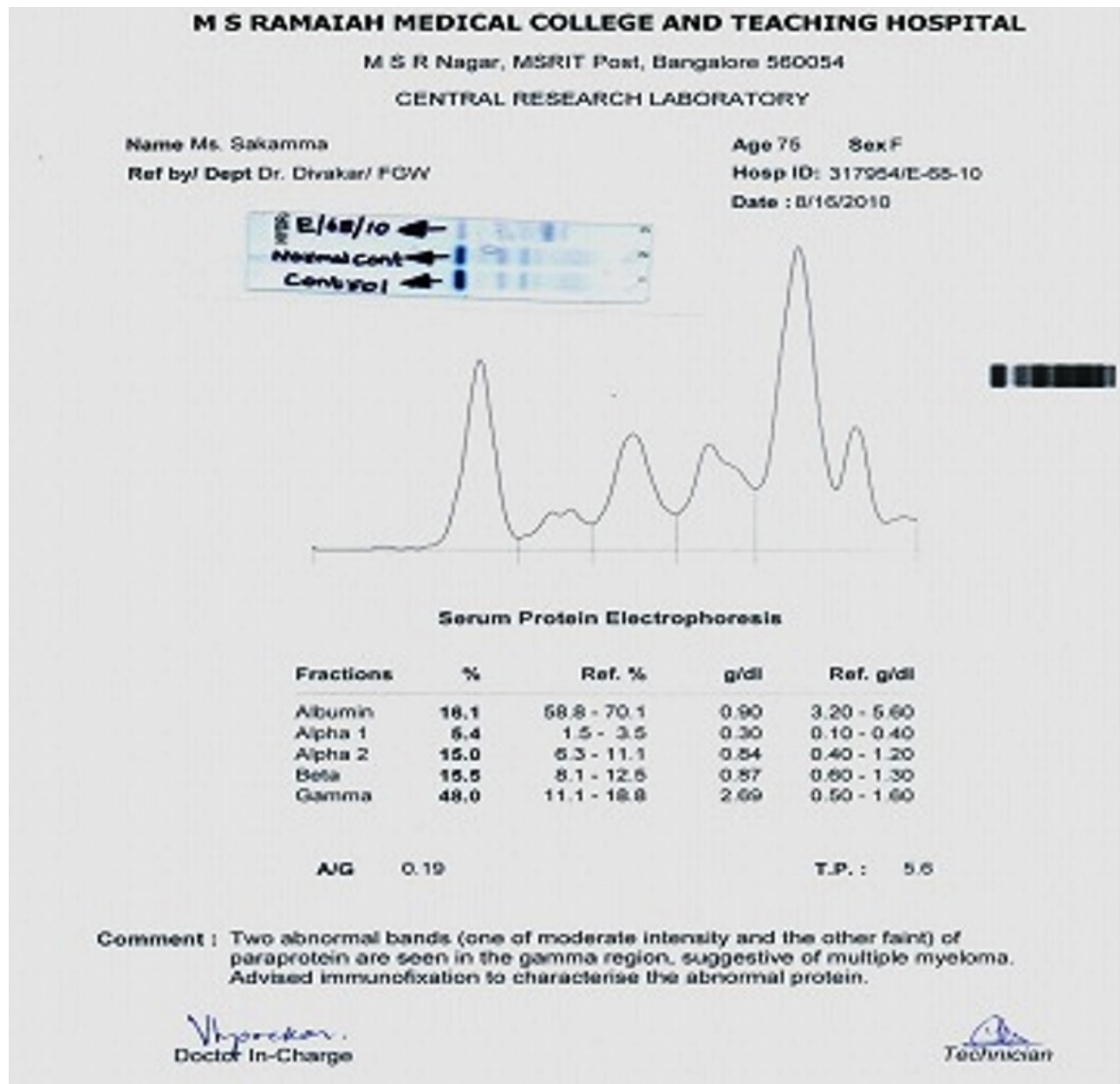


Fig. 5 : Electrophoresis Pattern

last 1 year for weakness. She also gave a history of receiving blood transfusion once for weakness. She was not a known diabetic or hypertensive. There was no Thyroid disease. On systemic examination patient had pallor, pedal edema and a mild splenomegaly.

On ocular examination nonpulsatile eccentric proptosis of left eye was noted with the globe displaced down by 10 mm, forwards by 6 mm and outwards by 2 mm. There was no change on Valsalva maneuver. Fullness was noted in the

supero-lateral part of the upper lid with a mild swelling of temporal region. There was a gross restriction of elevation and minimal restriction of abduction (Fig: 1 and 2)

The anterior segment examination was normal in the both eye except for the senile immature cataract in the left eye. The pupillary reactions of both eyes were normal. There were no signs of inflammation or visible mass. On palpation a mass was felt in the supero temporal region of the orbit. It was non tender, and firm in

consistency, its margins were ill defined and non pulsative. It was extending medially and also supero temporally onto the temporal fossa. It is nonpulsatile. Orbital margins were normal. Retropulsion was positive. The intraocular pressure was 10 millimeter of mercury in both eyes. Fundus examination was normal in both eyes except that the media was hazy in left eye due to cataract. The best corrected visual acuity was 6/9 in the right eye and 2/60 in left eye. Blood investigations revealed Hemoglobin of 6.2 gm %, total Count of 13,200 cell/cubic mm, differential count showed neutrophilia and ESR was 140 mm/hr. Blood Urea Nitrogen was 31 mg/dl, Serum creatinine was 2.6mg/dl and 24 hr urine protein 1290 mg. Abdominal ultrasound showed borderline splenomegaly, bilateral grade 2 Nephropathy, and minimal ascites. Breast and uterus examination was unremarkable and Pap smear was negative. Chest and spine radiology were normal. Computed tomography scan of head and orbit showed an isointense ill-defined mass in superior antero-lateral extraconal space extending to the medial side which was eroding the frontal and zygomatic bones.

At this point of time a differential diagnosis of secondaries from an unknown primary, lymphoma and neoplasia of the bone were considered. Since the CT was not suggestive of osteosarcoma or chondrosarcoma, plasmacytoma was considered and investigated further. Serum electrophoresis showed two abnormal bands of Para proteins in the gamma region and a diagnosis of multiple myeloma was made.

FNAC (Fig 4) showed cells with high nuclear - cytoplasmic ratio, eccentrically placed enlarged vesicular nuclei and prominent nuclei

and a few binucleate plasma cells and plasma blasts in proteinaceous background which was suggestive of multiple myeloma.

Patient was referred to a medical oncologist and chemotherapy was planned. Before the chemotherapy was started the condition worsened and the patient expired.

DISCUSSION

Multiple myeloma is a malignant plasma cell neoplasm characterized by monoclonal proliferation of a single clone of highly specialized B-lymphocyte engaged in the production of single immunoglobulin. It is a very rare cause of proptosis, where in unilateral proptosis is commoner than bilateral. A handful of cases of orbital myeloma are reported till date.^[4-8] In four series of orbital tumors all the incidence of orbital myeloma was 1 in 200 to 1 in 800. In 75 % of cases orbital involvement was the first presentation.^[9]

In most of the cases onset of symptoms is insidious with slowly progressing proptosis accompanied by pain, diplopia and visual impairment. Restriction of ocular motility particularly during abduction has also been described.^[10] Intracranial extension may lead to papilledema and cranial nerve palsies.^[11] Non-specific symptoms such as low grade fever, malaise and anorexia are common but can lead to an erroneous clinical impression in unsuspected cases. Orbital myeloma commonly presents as soft tissue intraorbital tumor which is an extension of bony deposit and is associated with a bony destruction. Rarely multiple myeloma involving base of the skull can present with proptosis with orbital invasion. Computed tomography is very helpful not only in

diagnosing the disease but also revealing amount of bone remodeling.^[12]

The diagnosis of multiple myeloma should be kept in mind in cases of proptosis. Early diagnosis and timely treatment would help to prolong the patient's life and to improve the quality of life.

CONCLUSIONS

In multiple myeloma, it is necessary to take a closer look at the ophthalmic manifestations as it may be the only clinical manifestation at early stage. Local orbital involvement might act as a reservoir for proliferation of myeloma cells and eventually can present with systemic relapse. All patients with multiple myeloma should thus undergo thorough ophthalmic examination at the time of initial diagnosis and during follow-up.

REFERENCES

1. Omoti AE, Omoti CE. Ophthalmic manifestations of multiple myeloma. *West Afr J Med* 2007;26(4):265-68.
2. Knapp AJ, Gartner S, Henkind P. Multiple myeloma and its ocular manifestations. *Surv Ophthalmol* 1987;31(5):343-51.
3. Fung S, Selva D, Leibovitch I, Husuan J, Cropton J. Ophthalmic manifestations of multiple myeloma. *Ophthalmologica* 2005;219:43-8.
4. Malik A, Narang S, Handa U, Sood S. 2009 Multiple myeloma presenting as bilateral orbital proptosis. *Ind J Ophthalmol* 2009;57:393-95
5. Fay AM, Lieb ML, Fountain K. Multiple myeloma involving the orbit. *Ophthalmic Plast Reconstr Surg* 1998;14:67-71.
6. Ajithkumar TV, Sivasankar C, Ramachandran K. Orbital multiple myeloma: Case report and review of computed tomography features. *Australasian Radiol* 2002;46:119-20.
7. Tuncbilek N, Karakas HM, Okten OO, Vural O. MRI in multiple myeloma with orbital and dural invasion. *Tumori* 2005;91:270-72.
8. Sharma A, Kaushal M, Chaturvedi NK, Yadav R. Cytodiagnosis of multiple myeloma presenting as orbital involvement: a case report. *CytoJournal* 2006;3:19.
9. Rodman HL, Font RL. Orbital involvement in multiple myeloma: a review of literature and report of three cases. *Arch Ophthalmol* 1972;87:30-35.
10. Knapp AJ, Gartner S, Henekind . Multiple myeloma and its ocular manifestations. *Surv Ophthalmol* 1987;31:343-51.
11. Clarke E. Plasma cell myeloma of the orbit. *Br J Ophthalmol* 1953;37:543-53.
12. Willinsky RA, Cooper PW, Kassel EE. CT of myeloma involving the skull base. *J Can Assoc Radiol* 1985;36:328-31.

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