Case study

Multiple myeloma relapse presenting as unilateral blindness.

Abstract

Multiple Myeloma is a chronic disease. While therapy is largely focused for control, relapses are un-evitable. Central nervous system relapse of myeloma is less common. Unilateral painless blindness is a rare entity and its occurrence in setting of multiple myeloma has not been We encountered a case of multiple myeloma on treatment, who developed unilateral blindness and later on confirmed to have relapse of disease.

Introduction

Multiple Myeloma is a systemic malignant condition of plasma cell lineage. About 3% of myeloma cases has extra medullary depositions of malignant plasma cells known as plasmacytomas.[1] While plasma cell deposits in upper half of skeletal system is common, involvement of central nervous system and orbits has been noted as well.[2,3,4] Unilateral optic nerve infiltration by plasma cells during treatment course of myeloma is an unusual and interesting finding.

Case Presentation:

A 57 years old female presented with loss of vision from right eye for 2 months. She was a diagnosed case of multiple myeloma [IgG, Kappa] for last 2 years. Her initial treatment was with inj Bortezomib, dexamethasone along with zolandronic acid. She achieved remission after 4 months and completed same chemotherapy for total 8 months. She was placed on thalidomide-dexamethasone regimen thereafter. She was on regular follow up and last assessment done 6 months back showed good control of disease. Her clinical examination now showed absence of light perception from right eye with no proptosis or ophthalmoplegia. Left ocular examination was normal. Detailed CNS examination showed no other significant finding. She underwent MRI brain which showed infiltrating mass compressing right optic nerve, distal to chaisma.[Fig1] Direct and indirect ophthalmoscopy was normal with normal fundus examination. Systemic

workup showed increase in plasma cells in bone marrow and increased beta 2 microglobulin, serum LDH and IgG kappa chain. A systemic relapse of disease was considered along with plasma cell deposits over right optic nerve. She was started on dexamethasone-cyclophosphamide regimen in view of CNS penetration of these drugs. Radiotherapy to plasma cell deposit was started. Inj bortezomib was added later in the course. She showed partial gain of vision [perception of light and finger counting at 1 meter present].

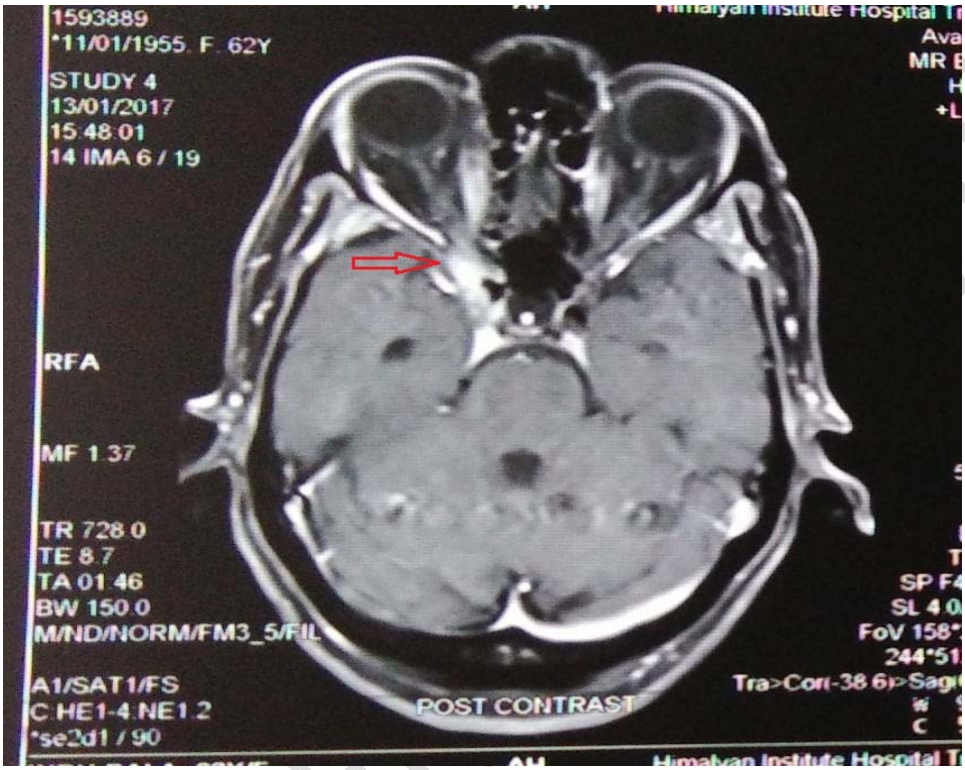

Fig I- MRI brain showing soft tissue density at right optic foramen.

Discussion

Multiple myeloma is a systemic disease of plasma cells. It is at one extreme of plasma cell dyscrasia, while milder localized forms can be detected as well. Being a systemic disease, its manifestation and presentation are variable. Usual presentation includes renal, bony and haematological parameters however infiltration of organs has been noted as well.[5,6] It is practically incurable disease and target of treatment is to control it. Usual course with treatment is also with relapses at variable time periods. Ocular manifestation of multiple myeloma at presentation of relapse are variable. It includes corneal deposits, macular exudative detachments,

or choroidal deposits. Usual presentation is with proptosis and hyperema. [7,8] Plasma cell infiltration of cranial nerves has been noted less frequently. Frequent involvement is noted for occulomotor, abducence and hypoglossal cranial nerves. [9,10] Optic nerve compression causing blindness has been noted as initial feature in literature.[11] However a systemic relapse presenting with unilateral blindness is an unusual finding.

Treatment of CNS involvement is not standardized and mostly derived from systemic therapy of myeloma. Drugs with good CNS penetration should work well. As plasma cell deposits are sensitive to radiotherapy, an immediate relief is expected from palliative radiotherapy. Index case presented to us with unilateral blindness for 2 months duration. Post radiotherapy, marginal improvement was noted only. A possibility of long standing compression causing permanent damage to optic nerve fibres is considered.

Conclusion:

Unilateral blindness due to malignant plasma cell deposits are uncommon clinical entity. As multiple myeloma is having vivid clinical presentation, a prompt suspicion should be kept for disease progression even for unusual complaints.

References

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