Rare presentation of orbital metastasis in multiple myeloma

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Access this article online

Quick Response Code:

Website: www.saudijophthalmol.org

DOI: 10.4103/sjopt.sjopt_109_22

Abstract:

Orbital manifestations are rarely observed in multiple myeloma (MM) and when they occur, they are often the first manifestation. We report a rare occurrence of vision loss in a 51-year-old female from orbital metastases in MM without proptosis or diplopia. The ophthalmic presentation of MM is usually progressive proptosis, pain, diplopia, and visual loss. The presence of metastasis in MM indicates poor prognosis and orbital metastases have worse survival rates. In conclusion, in cases of profound vision loss with no obvious cause, neuroimaging should be performed to evaluate the orbital extent of the disease and exclude intracranial pathology.

Keywords:

Metastasis, multiple myeloma, ocular, ophthalmic manifestations, orbit

Introduction

Multiple myeloma (MM) is the second-most prevalent hematological malignancy after non-Hodgkin's lymphoma.[1,2] It accounts for at least 2% of all cancer-related deaths (approximately 3000 in the UK) and is more common in men aged over 75 years.[1,2]

MM is a malignancy characterized by abnormal plasma cell proliferation and is generally confined to the bone marrow.[3] Typically, there is an overproduction of one distinct immunoglobulin or fragment of immunoglobulin from a cell clone of the lymphocyte/plasma cells series.[4] However, 3% of cases may develop extramedullary involvement, as the formation of solid plasmacytomas forms outside the bone marrow.[5] Extramedullary involvement usually occurs in the upper skeletal system, but rarely orbital manifestations are observed and when they occur, they are often the first manifestation of MM.[6-8]

Orbital involvement occurs more frequently than intraocular involvement and is typically diagnosed through computed tomography.[5,9-11] Proptosis is the most common presentation of patients with orbital MM (81%), it is also an indicator of metastases and recurrence.[3,12] Other common ocular symptoms include diplopia (23%), reduced vision (23%), swelling (21%), and ptosis (13%).[13] Neuro-ophthalmic manifestations may be the result of either a hematological process, compressive effect or direct myelomatous infiltration of ocular motor nerves or the visual pathway.[14,15]

This report presents a case of orbital involvement observed during a new diagnosis of MM.

Case Report

A 51-year-old mixed-race female with a medical history of asthma, pulmonary embolism, hepatic adenoma, type 2 diabetes mellitus, and uterine fibroids developed a 1-week history of shortness of breath, cough, and lethargy. She denied any fever or history of trauma. She was admitted for left-sided parapneumonic effusion, alongside multiple lytic lesions observed in the vertebrae identified on her computerized tomography pulmonary angiogram (CTPA). Medication history included seretide, salbutamol, citalopram, clonidine, naproxen, and co-codamol. Familial and social history was noncontributory.

Four weeks later, while awaiting further diagnostic workup for the multiple lytic lesions found on CTPA, the patient presented to our clinic with a 3-day history of progressive painless right eye vision loss.

On initial presentation, the best-corrected visual acuity was counting fingers in the right eye and

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Submitted: 06-Jul-2022
Revised: 04-Jan-2024
Accepted: 14-Jan-2024
Published: 22-Feb-2024

How to cite this article: O'Donovan C, Wu AD, Lim WS. Rare presentation of orbital metastasis in multiple myeloma. Saudi J Ophthalmol 2024;38:185-7.

20/20 in the left eye. There was a right-sided relative afferent pupil defect (RAPD). Ocular motility was full in both eyes. There was no ptosis and no proptosis. Intraocular pressure was 15 mmHg in the right eye and 13 mmHg in the left eye. Anterior and posterior segment examination was otherwise normal, with no signs of intraocular inflammation. Optical coherence tomography of the macular showed no abnormality.

In view of the patient right-sided vision loss, urgent magnetic resonance imaging (MRI) brain and orbits were performed showing numerous soft-tissue lesions within the bones of the skull (calvarium, the anterior skull base, the left mandibular body and at the junction of the body, and right lateral mass of C2) with compression and displacement inferiorly of the right optic nerve at the level of the orbital apex at the optic canal [Figure 1a and b]. In view of the widespread abnormalities on body imaging, the bony lesions were likely to represent either metastatic disease or hematological malignancy. She was subsequently sent for an MRI spine which identified multiple metastases throughout the spine.

A full diagnostic workup was undertaken in view of the patient's sudden right-sided vision loss, including blood pressure, complete blood count including erythrocyte sedimentation rate (ESR), glucose level, C-reactive protein (CRP), liver function, urea and electrolytes, Vitamin D, calcium, phosphate, immunoglobulins, and kappa: lambda light chain, paraproteins, and SARS-CoV-2 (COVID-19) polymerase chain reaction test. These tests were requested by the respiratory doctor to look for MM because of the unexplained multiple lytic lesions found on CTPA before the patient vision loss. The following bloods were abnormal: hemoglobin (102); calcium (2.73); CRP (60); ESR (57), and kappa: lambda light chain reaction (70.08).

The clinical signs combined with the patient's scans and blood results suggested a diagnosis of MM with orbital metastasis, which was subsequently confirmed on bone marrow biopsy.

Eight days later the patient returned to the eye clinic with pain on eye movements and was found to have right hypertropia and right variable exotropia. She was discussed at a multidisciplinary team meeting and it was advised for her to start 10-week weaning course of oral prednisolone.

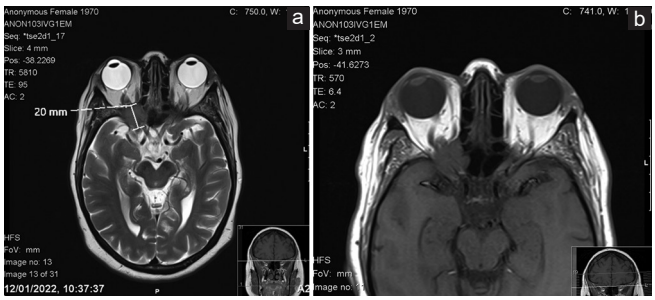

Figure 1: (a and b) Magnetic resonance imaging brain and orbits showing compression and displacement inferiorly of the right optic nerve at the level of the orbital apex at the optic canal. HFS: Hemifacial spasm, TR: Repetition Time, TE: Time to Echo, AC: Anterior commisure

She was also referred to a specialized hematology-oncology team for dexamethasone, thalidomide, cisplatin, adriamycin, cyclophosphamide, and etoposide (DT-PACE) chemotherapy and radiotherapy.

Discussion

MM is a progressive and incurable (although treatable) cancer, resulting in uncontrolled proliferation of neoplastic plasma cells in the bone marrow and excessive production of monoclonal immunoglobulins.[16] The international myeloma working group reported on diagnostic criteria for this disease: the presence of at least 10% plasma cells on examination of the bone marrow (or biopsy of tissue with monoclonal plasma cells), monoclonal protein (serum or urine), and evidence of end-stage organ damage.[2]

The onset of MM is insidious and the most common symptoms are pain, fatigue, and recurrent infections.[2] At least 90% of patients with MM have radiographically demonstrated "punched out" lytic lesions of bone or generalized osteoporosis.[8]

Plasmacytomas may also arise in the soft tissues primarily or in the surrounding bones with secondary orbital invasion.[8] In most cases, the proptosis is slowly progressive, 5 months on average, with accompanying pain, diplopia, and visual impairment.[12] There may also be restrictions in ocular motility, particularly abduction. Hence, this case represents an unusual presentation differing in the absence of common symptoms and rapid progression of vision loss.

The presence of metastasis in MM indicates poor prognosis and orbital metastases in particular have worse survival rates.[17] Chemotherapy and radiotherapy are essential in the management of MM in reducing tumor mass. Localized disease of MM is often solely treated with radiotherapy.[18] Surgery can also be performed for residual ophthalmic functional abnormalities.

In conclusion, we describe a rare occurrence of rapid vision loss from orbital metastases in MM without proptosis or diplopia as a presenting sign. This case highlights that in presentations of vision loss with a RAPD and otherwise normal ocular examination, neuroimaging should be performed.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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