CASE REPORT

Multiple myeloma masquerading as panuveitis in a middle-aged woman

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SUMMARY

Panuveitis secondary to masquerade syndrome is uncommon. A middle-aged woman presented to the ophthalmology clinic with panuveitis associated with anaemia, joint pain, and renal impairment. An incidental finding of a lytic lesion over her left scapula following a chest x-ray prompted further skeletal survey and revealed further lytic lesions over the skull and pelvic bone. Bone marrow aspiration was performed and this confirmed the diagnosis of multiple myeloma. Her left eye vision and intraocular inflammation improved after commencement of chemotherapy. A detailed history is important to elucidate the aetiology of masquerade syndrome and to prevent any delayed diagnosis of underlying malignancy.

INTRODUCTION

Panuveitis is inflammation of the eye involving all the uveal structures, retina and vitreous. Causes of panuveitis can be infective, inflammatory or secondary to masquerade syndrome. Masquerade syndrome happens when there is intraocular invasion by neoplastic cells, manifesting as intraocular inflammation. It is frequently being misdiagnosed or overlooked, causing a delay in the diagnosis and treatment of the underlying malignancy. We report a rare case of multiple myeloma masquerading as panuveitis in a middle-aged woman.

CASE REPORT

A 40-year-old Malay woman presented to the ophthalmology clinic complaining of blurring of vision and redness of her left eye for two weeks. Her visual acuity was counting fingers in the left eye and 6/9 in the right eye. There was no relative afferent pupillary defect. The left eye conjunctiva was mildly injected. Cells 3+ and flare 3+ were present in the anterior chamber. There were peripheral anterior synechiae at the left eye. The intraocular pressure was normal. The posterior segment view was hazy with vitritis and choroiditis in the peripheral retina. A diagnosis of left eye panuveitis was made.

A detailed history revealed that she had normochromic normocytic anaemia about 10 months ago, with haemoglobin level of 7.0g/dL, for which she was admitted for a blood transfusion in the medical ward. The patient subsequently defaulted follow up. The patient also had a 10-month history of neck and back pain. In view of her diagnosis of left eye panuveitis with underlying systemic problems, she was admitted for further investigations and was started on topical corticosteroids in the left eye.

A chest X-ray during the current admission showed a lytic lesion over her left scapula (Figure 1). The patient also had impaired kidney function with a serum creatinine of 128µmol/L. Her erythrocyte sedimentation rate was high at 65 mm/hour. Infection screening for hepatitis B and C, HIV, VDRL, and Mantoux test were all negative. With the positive systemic findings of anaemia, joint pain, mild renal impairment, and lytic lesion over scapula, the panuveitis was now thought to be secondary to masquerade syndrome due to either multiple myeloma or metastatic malignancy. She was referred to the general physicians for further work-up.

A skeletal survey revealed further lytic lesions over the skull (Figure 2) and pelvic bone. The serum protein electrophoresis was also positive. Bone marrow aspiration was subsequently performed showing clonal plasma cells >10%, confirming a diagnosis of multiple myeloma.

During this admission, the left eye inflammation improved with topical corticosteroids. The left eye best corrected visual acuity remained at counting fingers. Optical coherence tomography was performed and showed relative thinning of the macula of the left eye. No sub-retinal fluid was noted. We were unable to perform fundus fluorescein angiogram as there was a risk of further deterioration of her kidney status.

She was commenced on chemotherapy for multiple myeloma. A few weeks later, her left eye vision improved to 6/60 with an improvement in the degree of intraocular inflammation.

DISCUSSION

The term masquerade syndrome is used for malignant diseases manifesting as intraocular inflammation. Masquerade syndrome is frequently misdiagnosed as chronic idiopathic uveitis. Any delay in diagnosis and treatment of the underlying malignancy may lead to serious systemic morbidity and mortality. A study in the United Kingdom showed the frequency of neoplastic masquerade syndrome in patients with uveitis was 2.5%.[1] Masquerade syndrome is a rare entity, and the commonest aetiology is intraocular lymphoma. Multiple myeloma masquerading as panuveitis is extremely rare.

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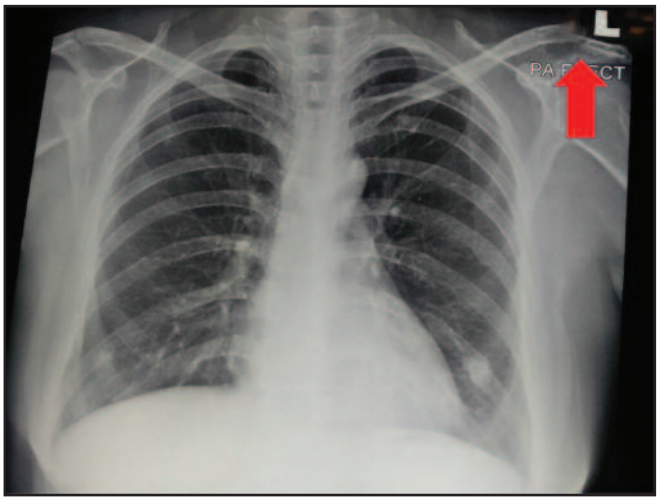

Fig. 1: An incidental finding on chest X-ray showing lytic lesion over the left scapula (arrow) which could easily be missed.

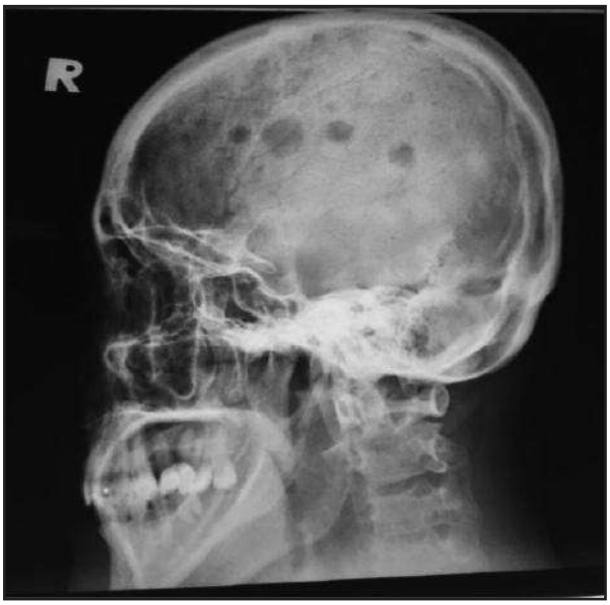

Fig. 2: Skull X-ray shows multiple lytic lesions, which are characteristics of multiple myeloma.

rare entity, and the commonest aetiology is intraocular lymphoma. Multiple myeloma masquerading as panuveitis is extremely rare.

From the literature, three cases were reported among the elderly. Guerriero et al., reported an elderly patient who presented with unilateral uveitis as a feature of multiple myeloma.[2] Vann et al., presented a case of choroidal melanoma masquerading as panuveitis in a patient with multiple myeloma.[3] Another case reported by Shakin et al., showed that there was iris involvement of neoplastic plasma cells manifesting as nongranulomatous uveitis in a 67-year-old elderly patient with multiple myeloma.[4] Although multiple myeloma causing masquerade syndrome appears to be associated with elderly patients, it can occur in younger patients; in our case, the patient is only 40 years old. Thus, clinicians must always exclude malignant myeloma as a cause of masquerade syndrome, however unlikely it may seem to be.

Multiple myeloma is a haematological malignancy where there is abnormal proliferation of plasma cells. The plasma cells proliferate in the bone marrow resulting in skeleton destruction and pathological fractures.[5] It is a disease of older adults, with a median age at diagnosis of 66 years. Only 10% of patients were younger than 50 years old. The common presenting symptoms were anaemia (73%), bone pain (58%), elevated creatinine (48%), hypercalcaemia (28%), and weight loss (24%).[5] Ophthalmological manifestations are rare in multiple myeloma. Diagnosis is by the presence of monoclonal protein in serum, clonal plasma cells of >10% in the bone marrow, and evidence of end organ damage due to plasma cell dyscrasia (lytic bone lesions, anaemia, and renal impairment). The treatment of multiple myeloma includes chemotherapy drugs (e.g., melphalan, cyclophosphamide, doxorubicin, thalidomide, and bortezomib), and corticosteroids.

This case highlights the fact that for cases of panuveitis, a detailed systemic enquiry is important to help elucidate the underlying aetiology. In our case, the history of normochromic normocytic anaemia, neck and joint pain for almost a year prompted us to consider masquerade syndrome and initiate a thorough systemic screen for an underlying malignancy. The lytic lesion and renal impairment provided us with the first clues that eventually pinpointed the aetiology of multiple myeloma.

In conclusion, diagnosis of masquerade syndrome in panuveitis is difficult but important. Besides ocular features, a thorough history may help raise suspicion of masquerade syndrome to an ophthalmologist and to differentiate masquerade syndrome from uveitis caused by infection or inflammation. Failing to take into considerations the systemic symptoms and signs of patients by the ophthalmology team would have led to the diagnosis of the underlying malignancy being delayed and further increasing the morbidity.

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