Necrotic choroidal melanoma masquerading as scleritis

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We report a case of a patient who presented with symptoms of acute onset scleritis in the left eye but was found to have an underlying necrotic choroidal melanoma. Asymptomatic choroidal melanoma can undergo spontaneous necrosis and manifest as severe scleritis and should be considered as an important differential.

Key words: Choroidal melanoma, scleritis, tumour necrosis

Scleritis is defined as inflammation of outer eye wall. Majority of scleritis cases are related to systemic diseases or infectious diseases. Rarely, uveal melanoma can masquerade as scleritis and on the other hand, posterior scleritis can also mimic uveal melanoma. Differentiating these two diseases remains a diagnostic challenge. This case describes a case of necrotic choroidal melanoma which presented with signs and symptoms of scleritis. This case highlights diagnostic challenges, relevant investigation and management of masquerading choroidal melanoma.

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Case Report

A 55-year-old Caucasian male was referred with sudden onset severely painful red left eye. He did not have any significant past ocular history or family history of ocular disease or cancer. He has type 2 diabetes and he is a non-smoker. On examination, his visual acuities were 6/4 in right eye and 6/6 in left eye. Left inferior sclera was grossly injected with 1+ cells anterior chamber [Fig. 1a]. Recorded intraocular pressures in both eyes were normal. The initial suspicion was of left anterior non-necrotizing scleritis. Dilated fundus examination of the left eye revealed a large inferior choroidal mass with surrounding exudative retinal detachment and sub-retinal pigment dispersion [Fig. 1b]. A-scan [Fig. 2a] and B-scan [Fig. 2b] showed a dome-shaped choroidal lesion of variable internal reflectivity, areas of necrosis with associated retinal detachment, surrounding choroidal thickening suggestive of reactive choroidal inflammation and scleritis.

We clinically suspected this to be a necrotic choroidal melanoma masquerading as scleritis. The differential of scleritis with secondary posterior segment involvement was ruled out by performing a thorough vasculitis and scleritis screening blood tests which were all negative for other possible causes of scleritis. Choroidal metastasis was ruled out by performing a CT scan of chest, abdomen, and pelvis which was normal. Ultrasound abdomen did not show any liver metastasis. MRI orbit with contrast showed a multilobulated, locally invasive

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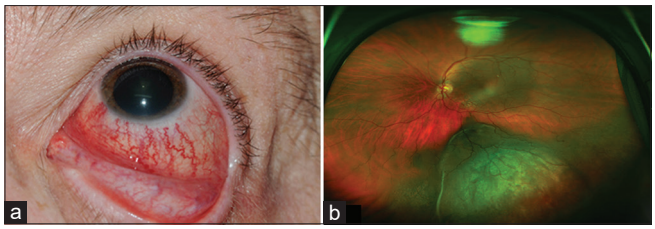

Figure 1: (a) Inferior scleritis at presentation (b) Fundus picture of left eye with suspicious inferior choroidal mass

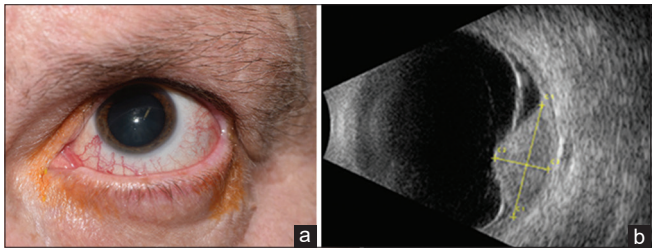

Figure 3: (a) Scleral hyperaemia had significantly improved following treatment with steroid. (b) Repeat B-scan showed an improvement of exudative retinal detachment and slightly enlarged choroidal mass

mass of high melanin content, possibly a melanoma, with a breach of sclera and associated inflammation in the left globe. There was no extraocular orbital disease. He was commenced on 2-hourly prednisolone acetate 1.0% eye drops and 3 grams of methylprednisolone infusion. Patient reported a significant symptomatic improvement [Fig. 3a] and the repeat B-scan within a week showed a more solid organized choroidal mass (T3c N0 M0 stage) with resolution of necrotic areas, improved exudative retinal detachment, and scleritis [Fig. 3b].

Patient then underwent uncomplicated left eye enucleation within 2 weeks of his initial presentation once the inflammation had settled. Histopathology results showed evidence of ciliary body and choroidal melanoma of mixed cell type (>10% epithelioid cells and <90% spindle cells) with extrascleral tumor extension which appears to arise from the infero-nasal vortex vein. This also confirmed low grade or resolving inflammation outer sclera adjacent to necrotic tumor [Fig. 4]. Following enucleation, patient has been symptoms free and is currently under regular ocular oncology clinic surveillance with no recurrence or metastasis at 1 year follow-up.

Discussion

The majority of ocular melanomas arise from uvea (83%).[1] Patients with uveal melanoma usually present of blurred vision, photopsia, floaters, visual field loss, or metamorphopsia. A significant proportion of patients (30%) are asymptomatic.[2] Our patient presented with symptoms of severe pain, which is classically associated with scleritis. Tumor necrosis is known to cause inflammation of the sclera and episclera, uveitis and raised intraocular pressure.[3] The cause of tumor necrosis in choroidal melanoma though is not fully understood. Thareja *et al.* hypothesized that the center of the tumor outgrows its blood supply in the watershed area. This leads to ischemic necrosis of the tumor, release of cytokines and tumor swelling with further necrosis, anterior displacement of the lens iris diaphragm, and secondary glaucoma which potentiates further tumor necrosis.[4] In our case, localized scleritis is most likely

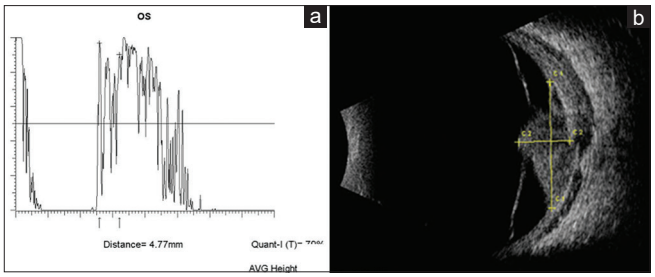

Figure 2: (a) A-scan showing mass of variable internal reflectivity. (b) B-scan showing a large suspicious choroidal mass with exudative retinal detachment, surrounding choroidal thickening and scleritis

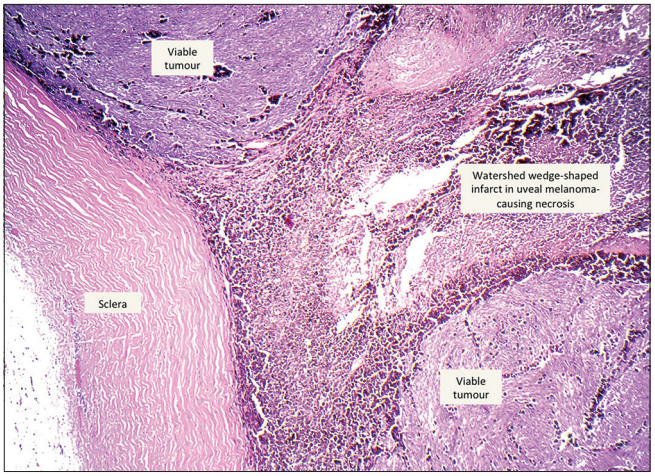

Figure 4: Necrotic tumor adjacent to sclera triggering local inflammation

due to rapidly growing large tumor which has outstripped its own blood supply. Alternatively, Brannan *et al.* suggested that unknown mechanism triggers tumor necrosis, release of cytotoxic products and the resulting vasculitis leads to infarction, swelling, and cellulitis of ocular and extraocular tissues.[5] Tumor necrosis may lead to spontaneous partial regression of choroidal melanoma which will eventually recur.[6]

Yap *et al.* reported a dramatic symptomatic improvement of masquerade scleritis and shrinkage of choroidal mass with systemic corticosteroid in the described two out of three cases of choroidal melanoma.[7] A trial of corticosteroid or non-steroidal anti-inflammatory drugs (NSAIDs) can differentiate masquerading scleritis of uveal melanoma from posterior scleritis with likelihood of resolution of mass following treatment with the latter.[8,9] Our patient was treated with topical and systemic steroid, which improved the secondary scleritis and exudative retinal detachment. Though it did not shrink the choroidal mass, the tumor appeared more solid and well organized, supporting the diagnosis of choroidal melanoma. Following counselling, our patient opted for enucleation not only due to the large melanoma size and evidence of possible scleral breach on MRI orbit, but also due to risk of scleritis flare up with radiotherapy.

Conclusion

Our case highlights that uveal melanoma can mimic other ocular diseases including scleritis. Radiological examinations as well as scleritis and vasculitis screening blood tests are required to

investigate for infective or systemic causes of scleritis. B-scan should always be performed in patients with scleritis as it may detect underlying intraocular malignancy. Atypical scleritis which is non-responding to treatment should warrant a biopsy.[10] Despite its diagnostic challenges, a step wise approach will help distinguish scleritis from masquerading scleritis of uveal melanoma and help direct appropriate treatment.

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Declaration of patient consent

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

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Conflicts of interest

There are no conflicts of interest.

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