Case Report

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Nodular posterior scleritis – The great masquerader

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Abstract:

We aim to report the management of a patient who presented with a choroidal mass masquerading as an amelanotic choroidal melanoma. A 57-year-old male presented with defective vision in his right eye, which was associated with mild periocular pain. Fundus examination showed a large dome-shaped yellowish-orange subretinal mass in the macular region and exudative retinal detachment (RD). Magnetic resonance imaging (MRI) showed a 16 mm × 8 mm choroidal mass, which was hyperintense on T1-weighted images and hypointense on T2-weighted images. B-scan ultrasonography revealed a dome-shaped mass with homogeneous echogenicity, inferior RD, and fluid collection in the sub-Tenon space. There was no choroidal excavation. He was diagnosed as nodular posterior scleritis (NPS) with exudative RD in the right eye. The lesion regressed completely after treatment with oral steroids. Choroidal mass can pose a diagnostic dilemma to ophthalmologists. Atypical MRI features can further augment the confusion. Despite its low incidence, NPS should always be kept as a differential in the presence of an amelanotic choroidal mass.

Keywords:

Amelanotic melanoma, choroidal mass, diagnostic dilemma, magnetic resonance imaging, posterior scleritis, pseudomelanoma

Introduction

Choroidal mass has been reported to be the presenting feature in a plethora of ophthalmological conditions. The differential diagnosis includes various benign and malignant choroidal tumors, metastatic deposits as well as other inflammatory conditions.[1,2] Shields *et al*. reported that almost 15% of the patients referred to them as choroidal melanoma were ultimately diagnosed with conditions having mimicking features. They labeled these conditions as "pseudomelanomas."[2] The pertinent ancillary tests need to be interpreted precisely as a misdiagnosis can lead to unnecessary interventions and treatment delays.

We present the management of a patient who presented with a choroidal mass masquerading as an amelanotic choroidal melanoma.

Case Report

A 57-year-old male was referred to our hospital with the diagnosis of choroidal melanoma. He complained of defective vision in his right eye for the past 7 days, which was associated with mild periocular pain. He was on regular medications for diabetes mellitus, hypertension, portal hypertension, liver cirrhosis, and dyslipidemia. His best-corrected visual acuity (BCVA) was 20/1200 in the right eye and 20/20 in the left eye. The ocular adnexal examination and intraocular pressures were normal in both the eyes.

Anterior segment examination of the right eye showed conjunctival injection and mild anterior chamber reaction. Posterior examination revealed a healthy disc and a

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large dome-shaped yellowish-orange subretinal mass in the macular region. The mass extended beyond the vascular arcades and was associated with chorioretinal folds and exudative retinal detachment (RD). However, the mass did not have any orange lipofuscin deposits on its surface [Figure 1a]. The left eye examination was unremarkable.

The magnetic resonance imaging (MRI) images showed a 16 mm × 8 mm choroidal mass, which was hyperintense on T1-weighted images and hypointense on T2-weighted images. MRI brain was not suggestive of a primary central nervous system lymphoma [Figure 2]. Fundus fluorescein angiography (FFA) showed blocked hypofluorescence due to the presence of the choroidal mass, heterogeneous hyperfluorescence during the early phase with late staining in the area of the lesion, and a hot disc. There was no evidence of "double circulation" [Figure 3]. Optical coherence tomography (OCT) revealed an elevated retina with overlying subretinal fluid [Figure 4a]. B-scan ultrasonography revealed a dome-shaped mass with homogeneous dense echogenicity, RD, and fluid collection in the sub-Tenon space (T-sign). There was no evidence of choroidal excavation [Figure 5a].

Blood workup for autoimmune markers (complete blood count, rheumatoid factor, antinuclear antibodies [ANAs], antineutrophil cytoplasmic antibodies [ANCAs], and angiotensin converting enzyme [ACE]); infectious causes (hepatitis, human immunodeficiency virus, and syphilis serologies) was negative. The renal and liver function tests were normal. Mantoux test was normal, and the chest X-ray did not show any consolidation or lymphadenopathy. The erythrocyte sedimentation rate (ESR) and blood sugar levels were elevated.

He was diagnosed as idiopathic nodular posterior scleritis (NPS) with associated exudative RD in the right eye. He was advised three doses of intravenous methylprednisolone (IVMP) but could not receive the treatment due to his uncontrolled sugar levels. Hence, he was given 1 mg/kg oral steroids under strict sugar monitoring. Within the next 72 h, the mass became distinctively flattened [Figure 1b] along with a reduction in the scleral thickening on B-scan ultrasonography. His BCVA also improved to 20/60. Over the next 3 months, the mass lesion regressed completely and the exudative RD disappeared [Figures 1c-f, 4b-e and 5b-d]. Fundus examination now showed retinal pigment epithelium (RPE) degeneration at the macula [Figures 1f]. The oral steroids were then slowly tapered off. Recurrence was not noted till the last follow-up.

Discussion

NPS is an uncommon scleral inflammatory disease which can present as a choroidal mass, causing diagnostic ambiguity.[3] The differential diagnosis of an amelanotic subretinal mass includes amelanotic choroidal melanoma, choroidal hemangioma, choroidal metastasis, choroidal osteoma, and choroidal granuloma.[1,2] Shields *et al*. reported that 0.3%–1.5% of patients presenting with "pseudomelanomas" actually had posterior scleritis.[1,2] Such a misdiagnosis can lead to the application of aggressive treatment modalities such as radiotherapy, chemotherapy, or even enucleation. Hence, it is

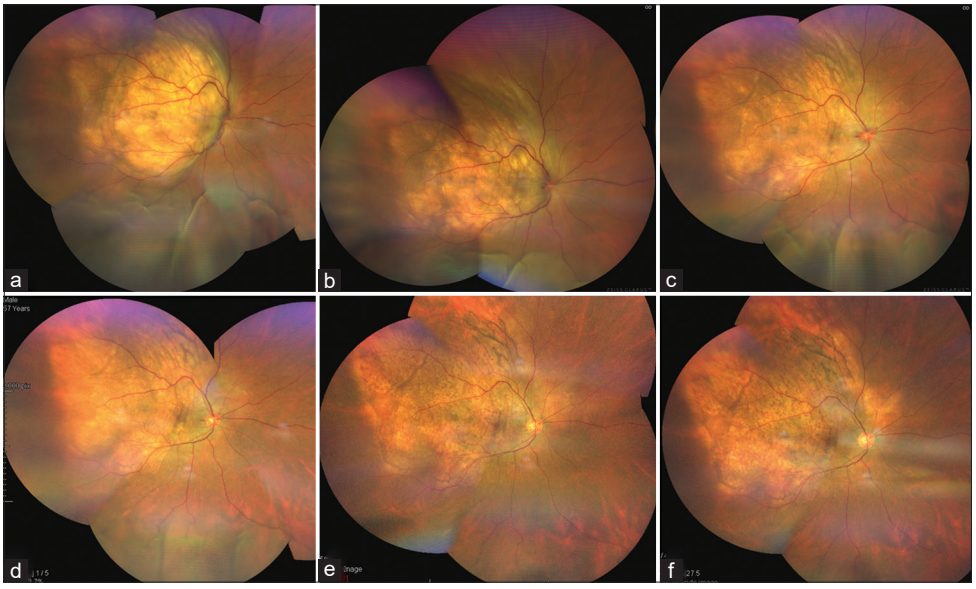

Figure 1: Montage of ultra-widefield image (Clarus, Zeiss) of the right eye showing (a) a large dome-shaped yellowish-orange subretinal mass in the posterior pole of macular region, associated with chorioretinal folds and inferior exudative retinal detachment at presentation; (b-e) reducing subretinal mass and exudative retinal detachment after 3 days, 1 week, 2 weeks, and 1 month after the start of the treatment; and (f) resolved mass with remnant retinal pigment epithelium degeneration at the macula while the mass and exudative retinal detachment have resolved, after 2 months after the start of the treatment

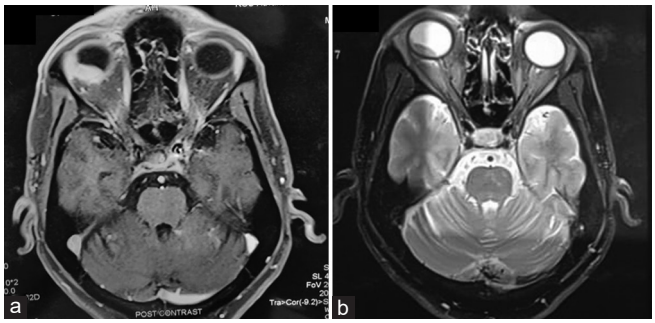

Figure 2: Magnetic resonance imaging at presentation showing a 16 mm × 8 mm choroidal mass which was (a) hyperintense on T1-weighted images and (b) hypointense on T2-weighted images

imperative to differentiate posterior scleritis from other pathologies that can present as a subretinal mass.

The presence of periocular pain, anterior scleritis, and other inflammatory signs favors the diagnosis of posterior scleritis.[3] Disc swelling, which is commonly seen in diffuse posterior scleritis, has rarely been reported in NPS. This may be due to the localized nature of the disease.[3] Thus, clinical examination in our patient suggested an inflammatory pathology. Currently, MRI is the most useful investigation to distinguish choroidal melanoma from other choroidal tumors. While a mass due to choroidal melanoma is characteristically hyperintense on T1-weighted images and hypointense on T2-weighed images, choroidal hemangioma is iso- to hyperintense on both T1- and T2-weighed images. However, these findings can be deceptive in the presence of inflammation. Inflammatory cells along with the free radicals can produce hyperintensity on T1-weighed images, thus mimicking the characteristic MRI findings of choroidal melanoma.[4] However, MRI can be helpful in ruling out intraocular lymphoma. The latter can present as a part of the systemic non-Hodgkin's B-cell lymphoma with choroidal infiltration. Thus, MRI findings in our patient were suggestive of either choroidal melanoma or an inflammatory pathology. The FFA findings are usually supportive but not definitive.[3] OCT in choroidal melanoma typically shows a gentle dome-shaped, smooth-surface topography. On the contrary, the presence of multiple nodular elevations of the RPE, called the "lumpy-bumpy" appearance, is characteristic of a choroidal metastasis or an inflammatory lesion.[5] Patients with choroidal metastasis can also present with pain, choroidal folds, and high reflectivity on ultrasound. In fact, it has been reported that it is not possible to find the primary tumor at the time of diagnosis of choroidal metastasis in almost one-fourth of cases.[6] Thus, OCT findings in our patient were suggestive of either choroidal metastasis or an inflammatory pathology. B-scan ultrasonography is considered to be the key noninvasive investigation as it provides the strongest evidence for the diagnosis of posterior scleritis. It is said

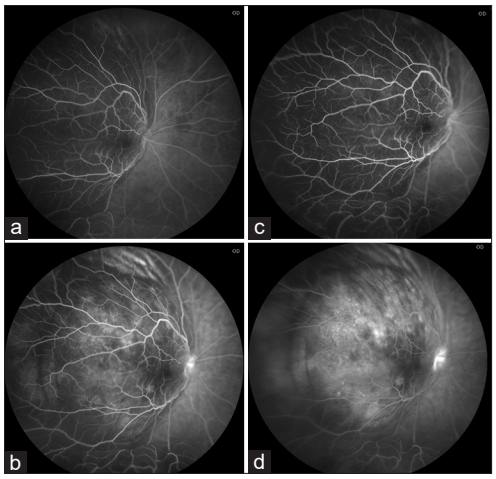

Figure 3: Fundus fluorescein angiography images showing (a) blocked hypofluorescence due to the choroidal mass; (b) heterogeneous hyperfluorescence in the area of the lesion during the early phase; and (c and d) late staining in the region of the lesion and a hot disc. There was no evidence of "double circulation"

to be just short of an invasive histological confirmation. While choroidal melanoma characteristically shows a collar-button configuration and choroidal excavation, choroidal osteoma typically shows a 100% spike with back-shadowing. On the contrary, the T-sign created due to a thickened sclera and the presence of retrobulbar edema is said to be the most pathognomonic sign of posterior scleritis.[3] Thus, B-scan findings in our patient were suggestive of posterior scleritis. To summarize, the fundoscopic and the MRI findings along with liver cirrhosis in this patient were suggestive of choroidal melanoma with probable liver metastasis. However, the presence of T-sign on B-scan ultrasonography, a "lumpy-bumpy" appearance on OCT, a hot disc on FFA, and an elevated ESR suggested an inflammatory cause. Hence, the patient was treated with oral steroids and he responded well.

McCluskey *et al*. reported that almost one-third of patients with posterior scleritis are associated with systemic diseases.[7] The various systemic diseases associated with NPS include rheumatoid arthritis (RA), temporal arteritis/giant cell arteritis (GCA), sarcoidosis, tuberculosis, ulcerative colitis, Wegener's arteritis, systemic lupus erythematosus (SLE), and birdshot retinopathy.[3,8,9] The absence of the typical clinical symptoms and a normal rheumatoid factor titers ruled out the presence of RA. The absence of any consolidation or lymphadenopathy on chest X-ray, a normal Mantoux reading, and a normal serum ACE level ruled out the possibility of both ocular sarcoidosis and tuberculosis. Although a temporal artery biopsy provides the most definitive diagnosis of GCA, it was not deemed necessary due to the absence of associated systemic features. Normal serum ANA and ANCA levels ruled out the possibility of both Wegener's arteritis and SLE.

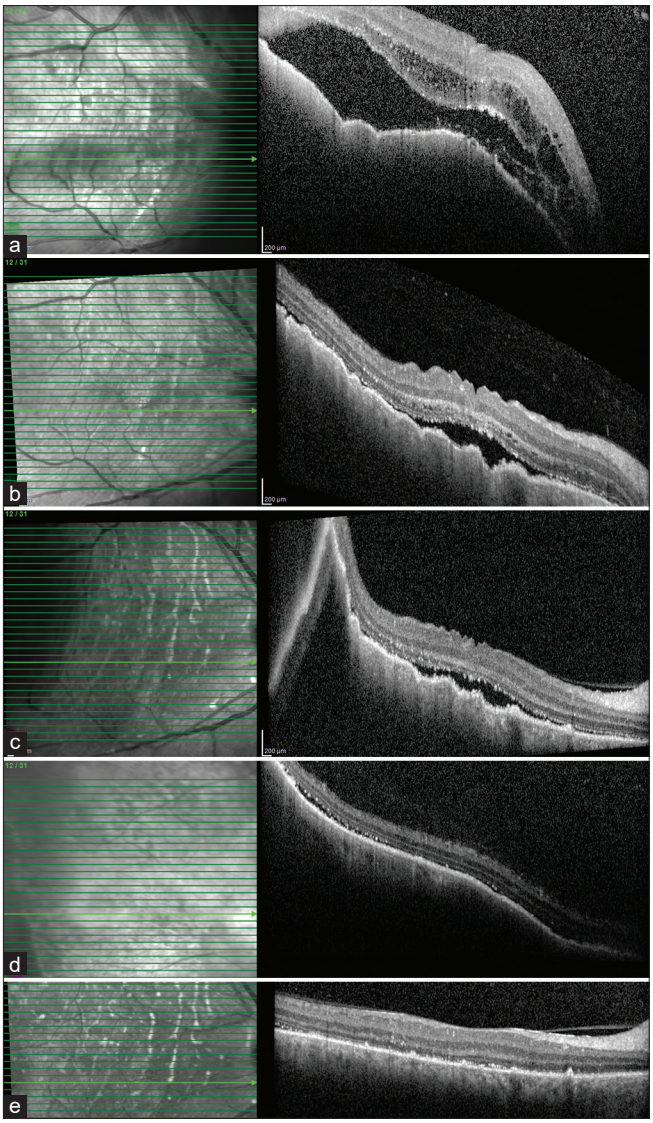

Figure 4: Optical coherence tomography line scans showing (a) multiple nodular elevations of retinal pigment epithelium with overlying subretinal fluid at presentation; (b-d) reducing subretinal mass and subretinal fluid after 1 week, 2 weeks, and 1 month after the start of the treatment; and (e) the mass and subretinal fluid have resolved after 2 months after the start of the treatment

To summarize, the presence of any associated systemic disease could not be established despite a thorough systemic workup. Hence, despite the presence of diabetes mellitus and liver cirrhosis, we concluded that NPS in the patient was most probably idiopathic. In fact, around 80% of the earlier reported cases of NPS were idiopathic in nature.[3,8,9]

Treatment of posterior scleritis includes nonsteroidal anti-inflammatory drugs, systemic or periocular steroids, and immunosuppressive agents.[3,8,9] Most authors advocate administering a pulse dose of IVMP; however, this patient could not receive it due to his uncontrolled sugar levels. Nevertheless, he responded well to oral steroids. Similarly, other authors have also

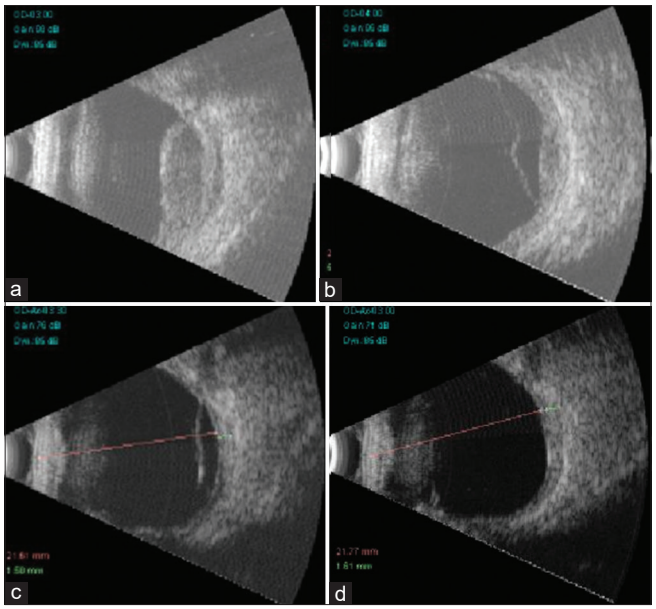

Figure 5: B-scan ultrasound showing (a) a dome-shaped mass with homogeneous dense echogenicity with inferior retinal detachment and T-sign but no choroidal excavation at presentation; (b and c) reducing height of the mass and fluid collection in the sub-Tenon space after 1 week and 3 weeks of the treatment; and (d) the mass and sub-Tenon fluid have resolved after 2 months of the treatment

reported that treatment leads to complete resolution of the nodule. However, Agrawal *et al*. showed that although the inflammatory signs reduced, the nodule persisted in almost all of their 11 cases. They suggested that a persistent but stable residual nodule does not merit any additional treatment. However, the patients should be kept under regular follow up as nodule thickness can fluctuate over time, which may warrant treatment.[3] Lim *et al*. have reported the beneficial role of intravitreal antivascular endothelial growth factor therapy as an adjunct for the treatment of posterior scleritis.[10] We would also have considered the same in case of a poor response to the primary treatment. However, our patient showed an early benefit and complete recovery with systemic treatment alone.

Choroidal mass can present a diagnostic conundrum for ophthalmologists. As seen in our case, atypical MRI features can further complicate the dilemma. Despite its low incidence, NPS should always be kept as a differential. Ancillary tests can help in differentiating it from other choroidal tumors. In case the diagnostic dilemma still persists, a therapeutic trial of systemic steroids can help resolve the issue. Unlike neoplastic lesions, posterior scleritis usually responds dramatically to steroids.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. The patient has given his consent for his images and other clinical information to be

reported in the journal. The patient understands that his name and initial will not be published and due efforts will be made to conceal identity.

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Nil.

Conflicts of interest

The authors declare that there are no conflicts of interests of this paper.

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