



## Case report

# A case of giant cell arteritis presenting with nodular posterior scleritis mimicking a choroidal mass

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## ABSTRACT

**Purpose:** Herein we present a case of giant cell arteritis presenting with nodular posterior scleritis and exudative retinal detachment mimicking a choroidal mass.

**Observations:** A 67-year-old man presented with sudden onset left-sided periorbital pain, blurry vision, and a choroidal lesion in the posterior pole. Despite treatment with high-dose oral prednisone for suspected nodular posterior scleritis mimicking a choroidal mass, the vision in his left eye did not recover, and he developed optic nerve pallor on exam. Further evaluation revealed an ESR of > 140 mm/hr (Upper limit of normal = 20 mm/hr), a CRP of 113 mg/L (Upper limit of normal = 9 mg/L), and a temporal artery biopsy consistent with GCA. The patient was started on methotrexate and the oral steroids were slowly tapered.

**Conclusions:** Given the potential for GCA to present with scleritis and the potential for nodular posterior scleritis to mimic a choroidal mass, presence of a painful choroidal lesion with optic nerve swelling should prompt an evaluation for GCA to prevent permanent and bilateral vision loss.

## 1. Introduction

Giant cell arteritis (GCA), also known as temporal arteritis, is a common systemic vasculitis of medium- and large-sized arteries in patients aged over 50 years with a wide spectrum of clinical manifestations. It is a true ophthalmic emergency—a delay in treatment may result in permanent and bilateral vision loss from ischemic complications. Ocular manifestations are often preceded by the most common systemic symptoms of the disease: new-onset headache, scalp tenderness, jaw claudication, and constitutional symptoms. Common ocular symptoms include visual loss, amaurosis fugax, diplopia, and eye pain. The most common ocular ischemic complication is arteritic anterior ischemic optic neuropathy with the classic sign of a pale and swollen optic disc. However, there are a variety of less common ocular manifestations of GCA, including anterior uveitis, ocular hypotony, scleritis, episcleritis, and, rarely, orbital pseudotumor.<sup>1</sup> We report a rare presentation of GCA with nodular posterior scleritis mimicking a choroidal mass.

## 2. Case report

A 67-year-old male presented to an outside ophthalmologist with sudden onset left-sided periorbital pain and blurry vision.

Ophthalmologic examination found a corrected visual acuity of 20/70 in the left eye and a choroidal lesion in the posterior pole. Given concern for a metastatic process to the left eye, he underwent an MRI of the brain and orbits and a CT of the chest, abdomen, and pelvis for systemic screening for malignancy. The MRI revealed a focal thickening in the posterior wall of the left globe with a posterior extraocular component. The CT found no masses.

Two weeks later, the patient presented to our clinic for further workup. By this time, vision in his left eye had further declined to 20/400 with continued periorbital pain. Dilation had been performed earlier that day and so evaluation for an afferent pupillary defect was not possible. Anterior segment evaluation of both eyes was unremarkable as was funduscopic evaluation of the right eye. Funduscopic examination of the left eye revealed an elevated amelanotic choroidal lesion in the macula measuring 10 mm × 7 mm, chorioretinal folds, exudative retinal detachment, and optic disc swelling (Fig. 1). Optical coherence tomography (OCT) revealed a dome-shaped choroidal “mass” with overlying subretinal fluid. Fluorescein angiography (FA) demonstrated punctate staining in the area of the lesion and leakage of the disc in the left eye. B-scan ultrasonography of the lesion revealed homogenous dense echogenicity with an apical height of 5.8 mm. Based on these findings, the patient was started on treatment with high-dose oral prednisone (60 mg daily) for suspected nodular posterior scleritis

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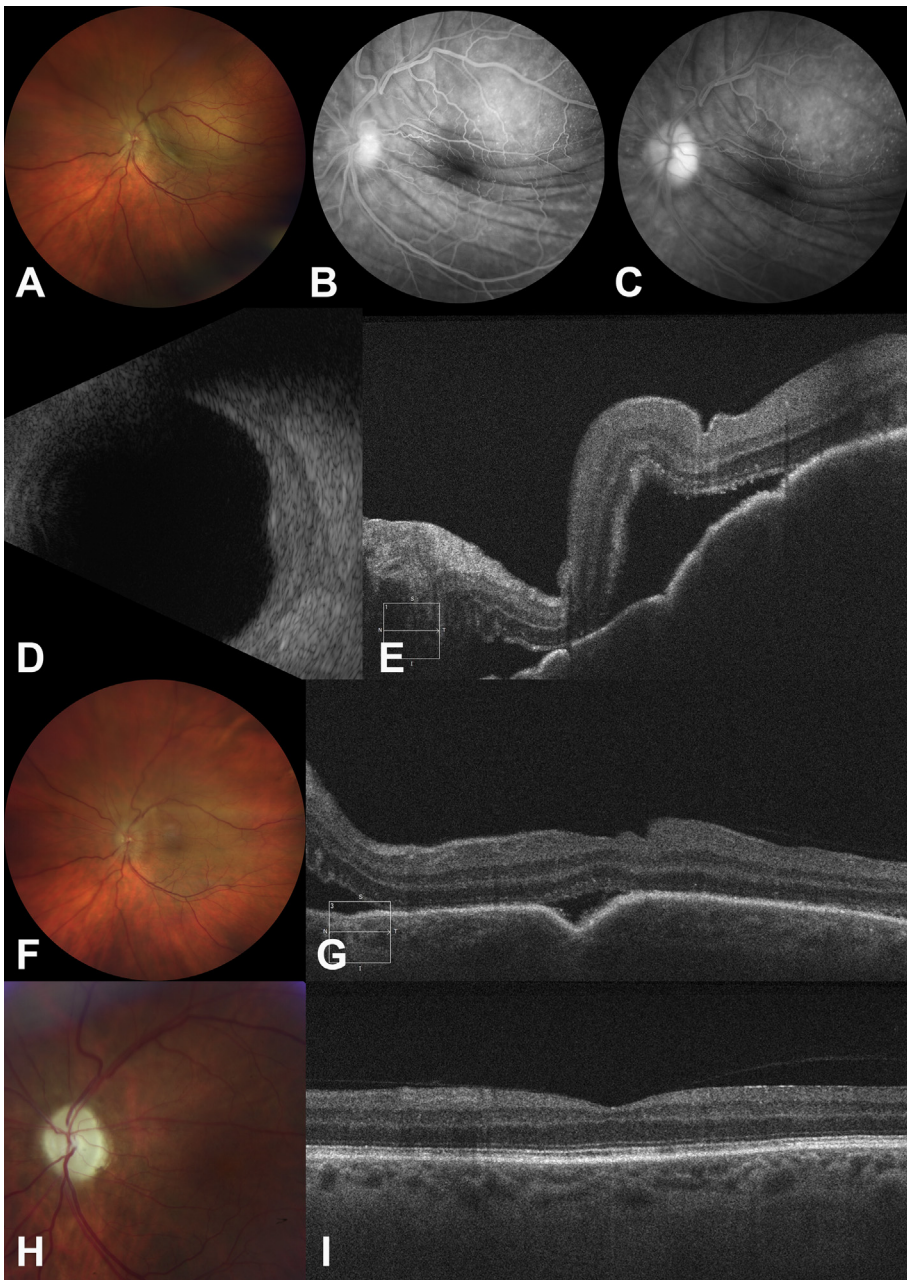
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**Fig. 1.** Baseline imaging of the left eye is represented in photos A-E. Fundus photo (A) showing an amelanotic macular choroidal lesion and optic nerve swelling. Mid-phase (B) and late (C) fluorescein angiographic images showing punctate staining of the lesion and leakage of the disc. B-scan ultrasonography (D) revealed diffuse choroidal thickening as well as an area of nodular thickening with homogenous dense echogenicity. Optical coherence tomography (OCT) through the lesion (E) revealed choroidal thickening with overlying subretinal fluid and retinal folds. After 3 weeks of oral steroids, the choroidal lesion had resolved (F) but optic nerve swelling was still evident. OCT at that time showed improvement of choroidal thickening and subretinal fluid (G). After another 3 weeks of oral steroids, optic nerve pallor was evident (H) on funduscopy while OCT revealed resolution of choroidal thickening and subretinal fluid.

mimicking a choroidal mass.

Laboratory workup for autoimmune markers (antinuclear antibodies, antineutrophil cytoplasmic antibodies, rheumatoid factor), infectious causes (syphilis, sarcoidosis, tuberculosis, Bartonella), angiotensin-converting enzyme and a chest X-Ray were all within the reference ranges. The patient's steroids were slowly tapered over several weeks to 20 mg daily with resolution of periorbital pain, choroidal thickening, exudative retinal detachment and optic nerve swelling. However, the vision in the left eye did not recover and stabilized at counting fingers (CF). Additionally, significant optic nerve pallor was noted at his 6-week follow-up while still on 20 mg of prednisone. Upon further questioning, the patient reported a slight headache with the taper of steroids. The degree of optic nerve pallor and headache raised concern for GCA, and the prednisone dosage was promptly increased to 60 mg daily. Further laboratory evaluation that day revealed an ESR of > 140 mm/hr (Upper limit of normal = 20 mm/hr) and a CRP of 113 mg/L (Upper limit of normal = 9 mg/L). A temporal artery biopsy was consistent with GCA. The patient was started on methotrexate and

the oral steroids were slowly tapered. No other systemic complications of GCA such as aortitis were found on further evaluation. Vision remained 20/20 in the right eye and CF in the left eye.

### 3. Discussion

Scleritis is an ocular inflammatory disease that is often associated with systemic autoimmune conditions (e.g. rheumatoid arthritis, relapsing polychondritis, psoriatic arthritis) some of which feature a prominent vasculitic component (e.g. granulomatosis with polyangiitis, systemic lupus erythematosus).<sup>2</sup> Given that GCA is a systemic immune-mediated vasculitis, which can involve any organ system,<sup>3</sup> involvement of scleral blood vessels with resulting scleritis may occur. In our review of the literature, we found only 2 reported cases of GCA in association with posterior scleritis.<sup>4,5</sup> This is, however, the first report of GCA presenting with nodular posterior scleritis mimicking a choroidal mass. GCA has, however, been reported to present mimicking a mass elsewhere including a submandibular mass<sup>6</sup> and testicular mass.<sup>7</sup>

Posterior scleritis classically presents with symptoms of decreased vision, ocular and periorbital pain, and headaches. Clinical signs include proptosis, ciliary and conjunctival injection, optic disc swelling, retinal striae, serous retinal detachment, and chorioretinal folds.<sup>4,5</sup> However, it can pose a diagnostic challenge due to its low incidence and variety of clinical presentations. In the case of nodular posterior scleritis, patients may present with a lesion large enough to mimic a choroidal mass. Although rare, reports in the literature serve as a reminder to consider nodular posterior scleritis if a patient presents with a painful choroidal mass.<sup>8–10</sup>

In our case, there was a delay in diagnosis of posterior scleritis due to reasonable concerns for a metastatic process by the referring physician. Additionally, the optic nerve swelling noted during our initial evaluation was incorrectly attributed to the posterior scleritis and while oral steroids were immediately started, an earlier diagnosis of GCA would have led to a more gradual taper of steroids. Initial fluorescein angiography did not reveal patchy choroidal filling which may have raised greater concerns for GCA earlier on. The diagnosis of GCA was ultimately suspected based on the significant and rapid onset of optic nerve pallor which is not characteristic of an inflammatory papillitis. Methotrexate has documented efficacy in the treatment of GCA and was our initial choice for steroid-sparing immunosuppression.<sup>11</sup> While tocilizumab is FDA-approved for the treatment of GCA, we reserve it for those who have failed a steroid taper while on methotrexate given tocilizumab's significant cost.

Given the potential for GCA to present with scleritis and the potential for nodular posterior scleritis to mimic a choroidal mass, presence of a painful choroidal lesion with optic nerve swelling should prompt an evaluation for GCA to prevent permanent and bilateral vision loss.

#### Patient Consent

Consent to publish the case report was not obtained. This report does not contain any personal information that could lead to the identification of the patient.

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#### Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

#### Declaration of competing interest

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