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CASE REPORT

Giant cell arteritis presenting with isolated cotton wool spots: a case report

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Abstract

This case aims to report a patient who presented with reduced vision in her left eye and was diagnosed with giant cell arteritis (GCA) associated with isolated cotton wool spots (CWS). An 82-year-old woman presented with reduced visual acuity of 20/200 in her left eye for a day. Fundus examination revealed only multiple peripapillary CWS in the left eye. She had an elevated erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP). A preliminary diagnosis of temporal arteritis, intravenous high-dose steroid therapy, was administered for 3 days. Then, the systemic symptoms resolved, and her ESR and CRP dropped. Temporal artery biopsy confirmed the diagnosis of GCA. The next 2 months, in the fundus examination, CWS resolved completely. The patient continued using systemic steroids and subcutaneous methotrexate with long-term gradual reduction. This extreme case should raise awareness for clinicians in the etiological investigation of CWS to identify sight-threatening GCA and promptly initiate appropriate treatment.

Keywords: Cotton wool spot; giant cell arteritis; ophthalmological emergency.

Giant cell arteritis (GCA), systemic granulomatous vasculitis that primarily affects large arteries, is one of the few true ophthalmic emergencies.[1] It commonly involves the ophthalmic artery, compromising blood supply to the eye, and as such has the potential to cause irreversible visual loss. The risk of vision loss can be reduced with an early clinical diagnosis of GCA and appropriate management.

The majority of clinical reviews on GCA to date have focused on the common ophthalmic manifestations, which primarily include arteritic anterior ischaemic optic neuropathy, central retinal artery occlusions, and cranial nerve palsies.[2]

By describing a patient who presented with unilateral, isolated, and nummular cotton wool spots (CWS), we hope to raise clinicians' awareness of the condition and the atypical ophthalmic clinical presentation of GCA.

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Case Report

An 82-year-old woman presented with reduced vision in her left eye for 1 day. The patient had a history of hypertension that was chronic but well-controlled and managed. She also mentioned that 5 days before applying, she experienced a 15-min temporary loss of vision as well as jaw pain while chewing. She denied experiencing joint pain, anorexia, or weight loss.

Ophthalmological examination revealed that visual acuity was 20/20 in the right eye and a reduced visual acuity of 20/200 with a relative afferent pupillary defect in the left eye. In the slit-lamp examination of the anterior segment, both eyes were pseudophakic. Color vision was 20/20 plate in the right eye and 1/21 plate in the left eye with the ishihara test. Dilated fundus examination revealed multiple peripapillary CWS in the left eye; the right eye was normal (Fig. 1). In the visual field, an inferior arcuate scotoma was observed in the right eye and a total scotoma in the left eye (Fig. 2). The right eye's visual field defect was evaluated as a frame effect. Spectral domain-optical coherence tomography of the patient was normal in the right eye, and hyperreflectivity which was compatible with the cotton wool spot was observed on the superficial retinal layers of the papillomacular bundle and disorganization in the ellipsoid zone of the left eye (Fig. 3). Physical examination also revealed prominence of the temporal arteries with tenderness bilaterally. She had an elevated erythrocyte sedimentation rate (ESR) (112 mm/h), C-reactive protein (CRP) (24.7 mg/L), and platelet count, which was normal (299,000 mcL). For a pre-

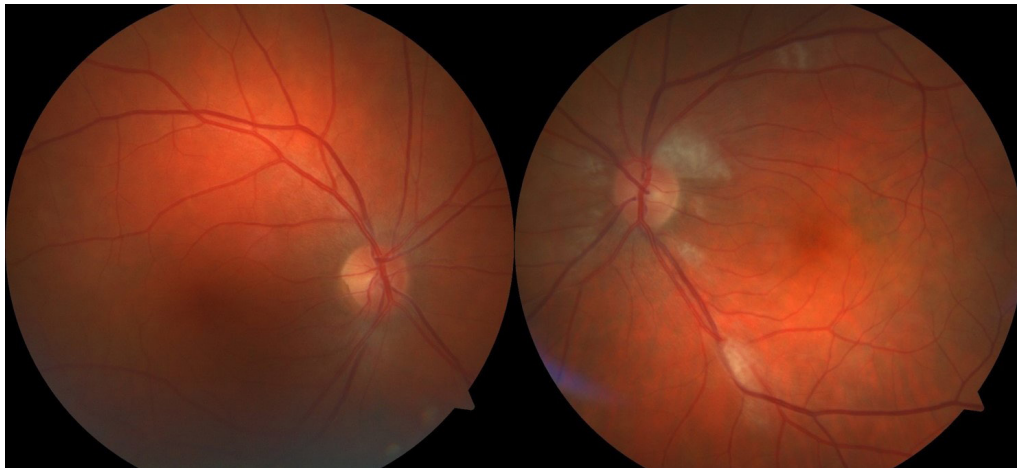

Fig. 1. In the fundus examination, peripapillary cotton wool spots were detected in the left eye and the right eye was normal

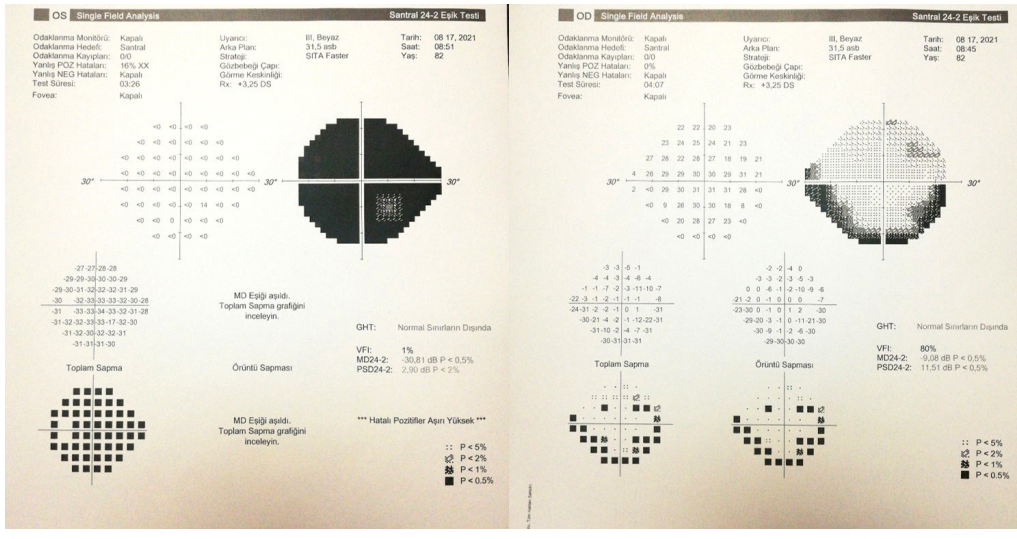

Fig. 2. In the visual field, an inferior arcuate scotoma was observed in the right and total scotoma in the left eye

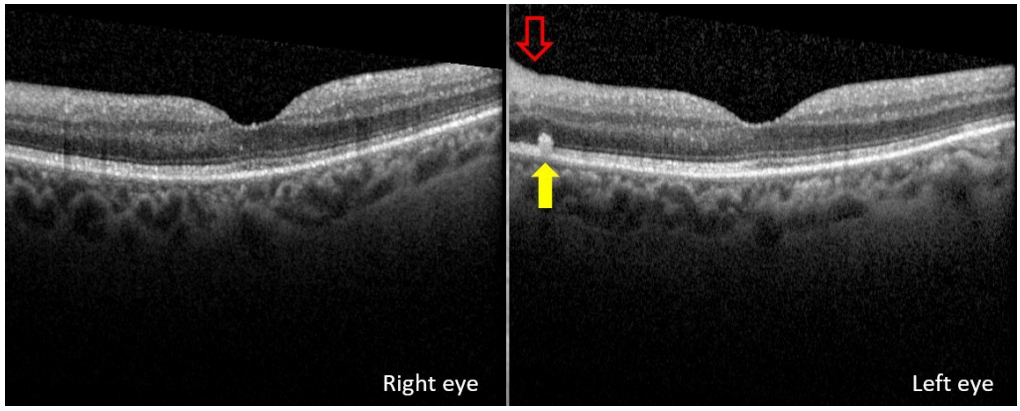

Fig. 3. Spectral domain-optical coherence tomography of the patient was normal in the right eye; and hyperreflectivity was observed on the superficial retinal layers of the papillomacular bundle (red arrow) and disorganization in the ellipsoid zone (yellow arrow) of the left eye

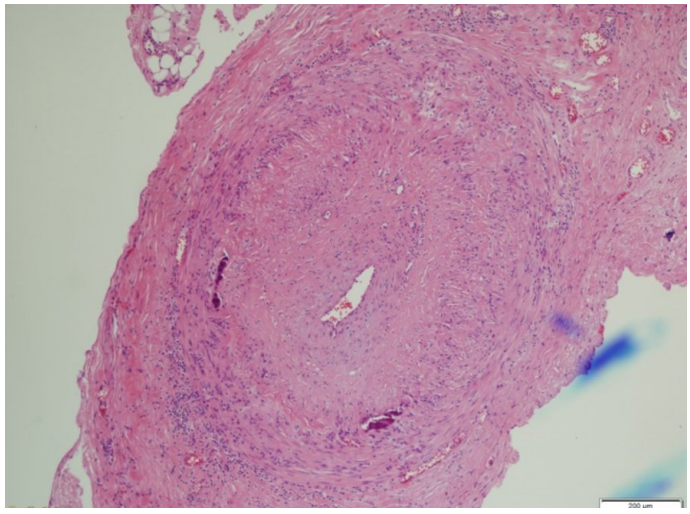

Fig. 4. Temporal artery biopsy demonstrated giant cells and lymphocytes near a fragmented internal elastic lamina

liminary diagnosis of temporal arteritis, intravenous high-dose steroid therapy was administered for 3 days. Despite high-dose intravenous treatment with steroids, her vision deteriorated in her left eye (20/400). However, the vision of the right eye was preserved (20/20). Three days later, the systemic symptoms resolved, and her ESR dropped to 43 mm/h and CRP to 4.8 mg/L. It was then followed up with an oral steroid dose of 1 mg/kg. Temporal artery biopsy demonstrated giant cells and lymphocytes in close proximity to a fragmented internal elastic lamina, confirming the diagnosis of GCA (Fig. 4). Over the next 2 months in the fundus examination, the CWS resolved completely and optic atrophy occurred in the left eye (Fig. 5). In the visual field, the right eye was normal and total scotoma continued in the left eye (Fig. 6). The patient continued to use systemic steroids and subcutaneous initial doses of 10 mg

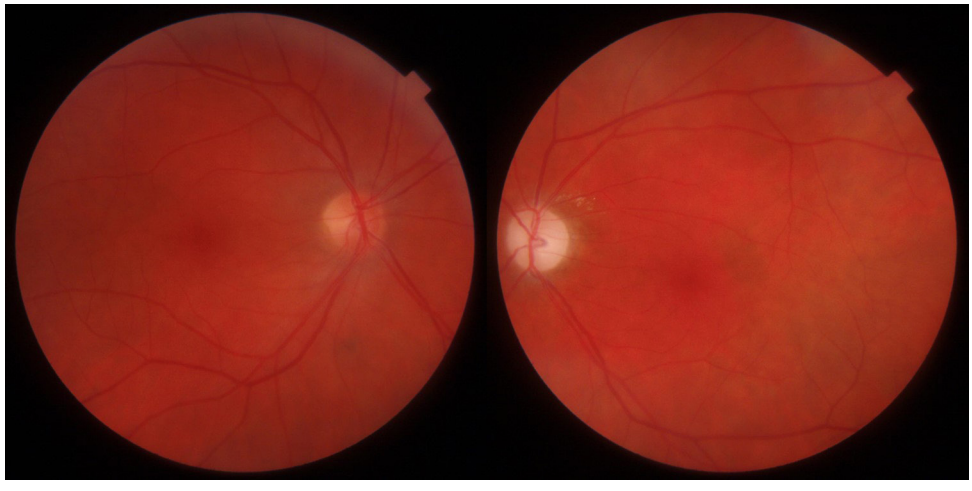

Fig. 5. In the fundus examination, the cotton-wool spots resolved completely, and optic atrophy occurred in the left eye

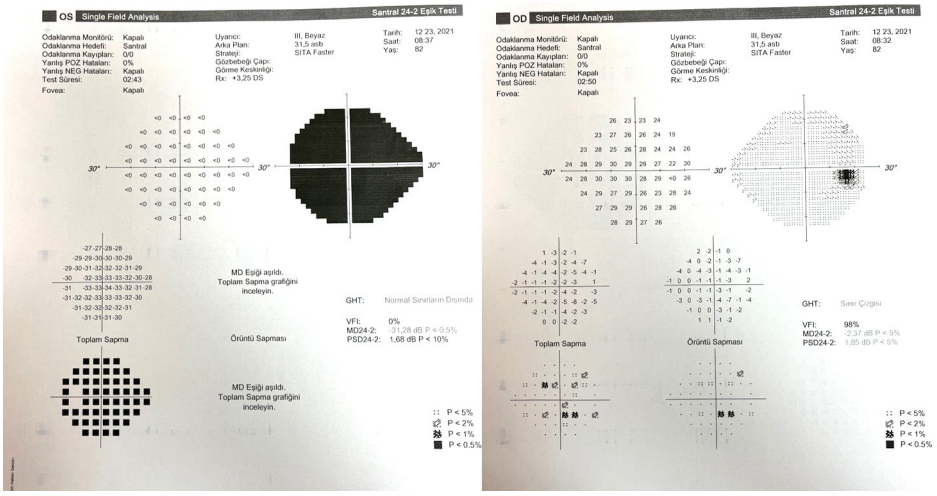

Fig. 6. In the visual field, normal was in the right eye and continue of total scotoma was observed in the left eye

methotrexate with long-term gradual reduction. No acute ophthalmologic signs of temporal arteritis were detected in the control visits.

Discussion

Ocular involvement occurs in up to 70% of patients with GCA. The most common ocular sign is anterior ischemic optic neuropathy, which presents with visual loss.[2] A pale swollen optic disc strongly suggests GCA-related anterior ischemic optic neuropathy, but the absence of pallor does not exclude this diagnosis.

CWS has been observed in approximately 30% of GCA patients who experience vision loss in the early stages of their disease.[3] Although CWS can occur in GCA, they are rarely isolated presenting signs. In a few studies, patients with temporal arteritis were found to have isolated CWS.[4-8] Melberg et al. reported seven cases of temporal arteritis with isolated CWS. Other than temporal arteritis, no other ocular, or systemic conditions to explain CWS were found in any of the study's patients.[7] Sanchez et al. described two patients who were presented with a single isolated CWS as the only clinical manifestation. They put forward the hypothesis that "CWS can be an early ophthalmoscopic finding in GCA and can precede an important visual loss."[8]

The etiology CWS of in GCA is likely multifactorial and includes microembolization of platelets and/or hypoperfusion of terminal portions of retinal vasculature involved by GCA. Our patient was unusual in manifesting CWS in the presence of clinically normal optic nerves. This was presumably caused by an unusual pattern of involvement, sparing the short posterior ciliary arteries while reducing flow in the retinal circulation. In our patient, the presence of a relative afferent pupillary defect and decreased color vision in the ophthalmologic examination; the presence of jaw claudication; and a high ESR were the keys to diagnosing GCA.

Conclusion

In routine clinical practice, the presence of CWS, regardless of the distribution or mechanism of their underlying formation including central retinal artery occlusion, and/or cilioretinal artery occlusion should notify the clinician of possible GCA diagnosis. Any patient whose vision is threatened due to suspected GCA should be given high-dose intravenous steroid treatments and referred for specialist management.

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Conflict of Interest: None declared.

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