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Central Retinal Arterial Occlusion in Granulomatosis with Polyangiitis

Dear Editor,

Granulomatosis with polyangiitis (GPA), previously known as Wegener's granulomatosis (WG), is a systemic disorder that involves both granulomatosis and polyangiitis. It is a form of vasculitis that affects small- and medium-sized vessels in many organs. Damage to the lungs and kidneys can be fatal [1].

We encountered a case of central retinal artery occlusion (CRAO) with well-controlled GPA in other organs. The case is described herein with a review of the relevant literature.

A 57-year-old female visited the retina clinic with a 1-day complaint of visual disturbance in the right eye. The patient had been diagnosed with atrial fibrillation, hypertension, congestive heart failure, and fibromyalgia 5 years previous. She took an antiplatelet agent (Lixana 30 mg; Daichii-Sankyo, Tokyo, Japan) regularly, and her recent transthoracic echocardiogram showed no cardiac thrombosis. One year previously, the patient was diagnosed with GPA and was started on prednisolone 5 to 10 mg twice a day and azathioprine 100 mg daily. The best-corrected visual acuity was finger count 30 cm in the right eye and 0.63 in the left eye. Fundus examination of the right eye showed a whitish posterior pole with a cherry red spot (Fig. 1A). Indocyanine green angiography of the right eye showed a wide hypofluorescent area including the fovea and optic nerve head in the early phase, implying choroidal hypoperfusion (Fig. 1B). In fluorescein angiography of the right eye, the arm to retina time was delayed to 42.37 seconds, and the arteriovenous transit time was delayed to 181 seconds. Optical coherence tomography showed increased reflectivity in the inner retina (Fig. 1C). Fundus examination and angiography of the left eye showed normal findings. The patient was diagnosed with CRAO and was referred to the neurology department. Doppler sonography of the carotid artery and magnetic resonance angiography of the brain and carotid artery revealed no specific stenosis. After 1 month, the arm to retina time had reduced to 17 seconds, but best-corrected visual acuity remained at finger count 30 cm.

Up to 77% of patients with GPA develop ocular manifestations during the course of the disease. The ocular manifestations of GPA are vascular occlusion, macular edema, and inflammatory destruction of the retina, optic nerve, or corneoscleral tissue [2]. The mechanism of retinal vasculitis in GPA remains uncertain; however, anti-neutrophil cytoplasmic antibody-associated vasculitis is suspected. In this process, neutrophils are bound to the vascular endothelium, inducing vessel wall injury [3].

There are several reports about ocular vascular occlusions. Kinyoun et al. [4] reported WG cases associated with bilateral choroidal infarctions. Mirza et al. [5] also reported CRAO in WG. The visual symptom was the first sign in these reported cases. However, in the current case, CRAO developed while vasculitis was well controlled with medication after diagnosis, and there was no definite vascular lesion triggering the CRAO. This case was evaluated using multimodal technology including indocyanine green angiography. Systemic workup including echocardiography, carotid vessel Doppler sonography, and brain angiography was performed, and none of these imaging tools indicated the possibility of emboli as a cause of arterial occlusion. Therefore, the retinal and choroidal ischemia in the present case might have been induced solely by vasculitis in retinal and choroidal arteries. CRAO combined with choroidal

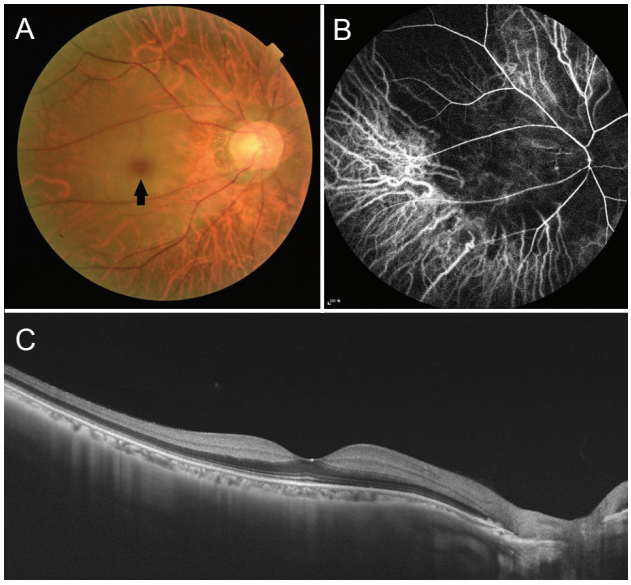

Fig. 1. (A) Fundus photograph of the right eye showing a whitish lesion with a cherry red spot (arrow) at the posterior pole. (B) Indocyanine green angiogram of the right eye showing a widefield watershed area perfusion defect. (C) Optical coherence tomogram showing hyper-reflective inner retina.

hypoperfusion suggests that GPA-induced vasculitis involved not only the retinal artery, but also choroidal vessels, or that the primary lesion involved a proximal vessel such as an ophthalmic artery, which can affect retinal and choroidal circulation simultaneously.

In conclusion, CRAO may develop even in well-controlled GPA without systemic vascular lesions.

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Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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