Carotid artery dissection associated with Purtscher-like retinopathy

Purtscher retinopathy was first described in 1910 by Otmar Purtscher in a man who suffered a traumatic head injury.[1] Patients commonly present with a loss of visual acuity and a recent history of injury such as head trauma or chest compression.[2,3] In the case of a nontraumatic injury, the terminology commonly used is "Purtscher-like retinopathy." It is associated with various systemic diseases, including acute pancreatitis, pancreatic adenocarcinoma, thrombotic thrombocytopenic purpura, connective tissue disorders, and hemolytic uremic syndrome.[2] We report a case of Purtscher-like retinopathy associated with carotid artery dissection.

Report of a Case

A 40-year-old woman presented with sudden-onset vision loss in her right eye. Two days earlier she had an ischemic stroke of her right middle cerebral artery secondary to a right carotid artery dissection, which was confirmed by computed tomography angiography (Fig. 1A). Her medical history included uterine fibroids and iron-deficiency anemia. Her ocular history was unremarkable.

At presentation, her visual acuities measured 20/100 OD and 20/40 OS, improving to 20/60 OD and 20/20 OS with pinhole. Her intraocular pressures were within normal limits OU and she had a relative afferent pupillary defect OD. Her anterior segment examination was normal OU. Fundus examination revealed intraretinal hemorrhages, Roth spots, cotton-wool spots, Purtscher flecken OD (Fig. 1B) and a normal examination OS (Fig. 1C). Spectral domain optical coherence tomography revealed thickening and hyper-reflectivity of the inner retina, consistent with paracentral acute middle maculopathy (Fig. 2A). At 6-month follow-up her visual acuity improved to 20/25 OD with pinhole, and optical coherence tomography showed full resolution of the paracentral acute middle maculopathy lesions with mild parafoveal inner nuclear layer atrophy (Fig. 2B).

Comment

To our knowledge, this is the first reported case of Purtscher-like retinopathy secondary to carotid artery dissection. In young adults, carotid artery dissection is responsible for up to 20% of strokes.[4] Ipsilateral headache is the most frequent symptom in carotid dissection, occurring in 70% of patients and is the initial manifestation in half of these.[5] A painful third-order Horner syndrome occurs in about 60% of patients, characterized by ptosis, miosis, and the absence of anhidrosis.[6] Less common ophthalmologic features include transient monocular vision loss, nonarteritic ischemic optic neuropathy, posterior ischemic optic neuropathy, central retinal artery occlusion, ocular ischemic syndrome, and ocular motor nerve palsies.[6]

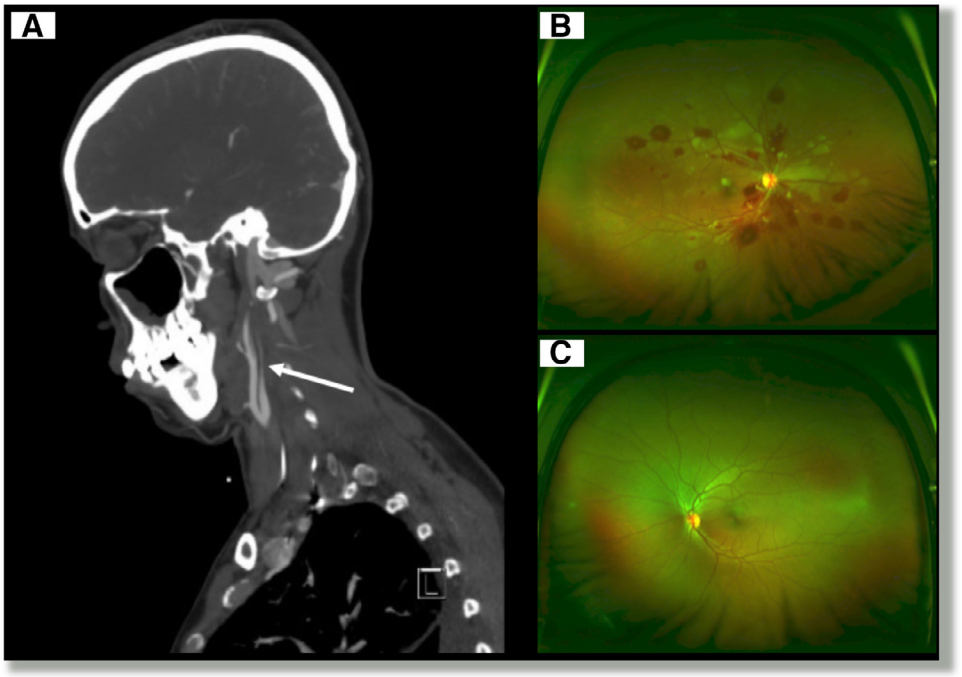

Fig. 1—(A) Computed tomography angiogram (CTA) reveals a right internal carotid artery (ICA) that is narrowed secondary to a right ICA dissection (arrow). The left ICA appears normal. (B) Fundus photograph OD showing characteristic polygonal areas of Purtscher flecken along with cotton-wool spots, Roth spots, and retinal hemorrhages. (C) Fundus photograph OS showing a normal retina.

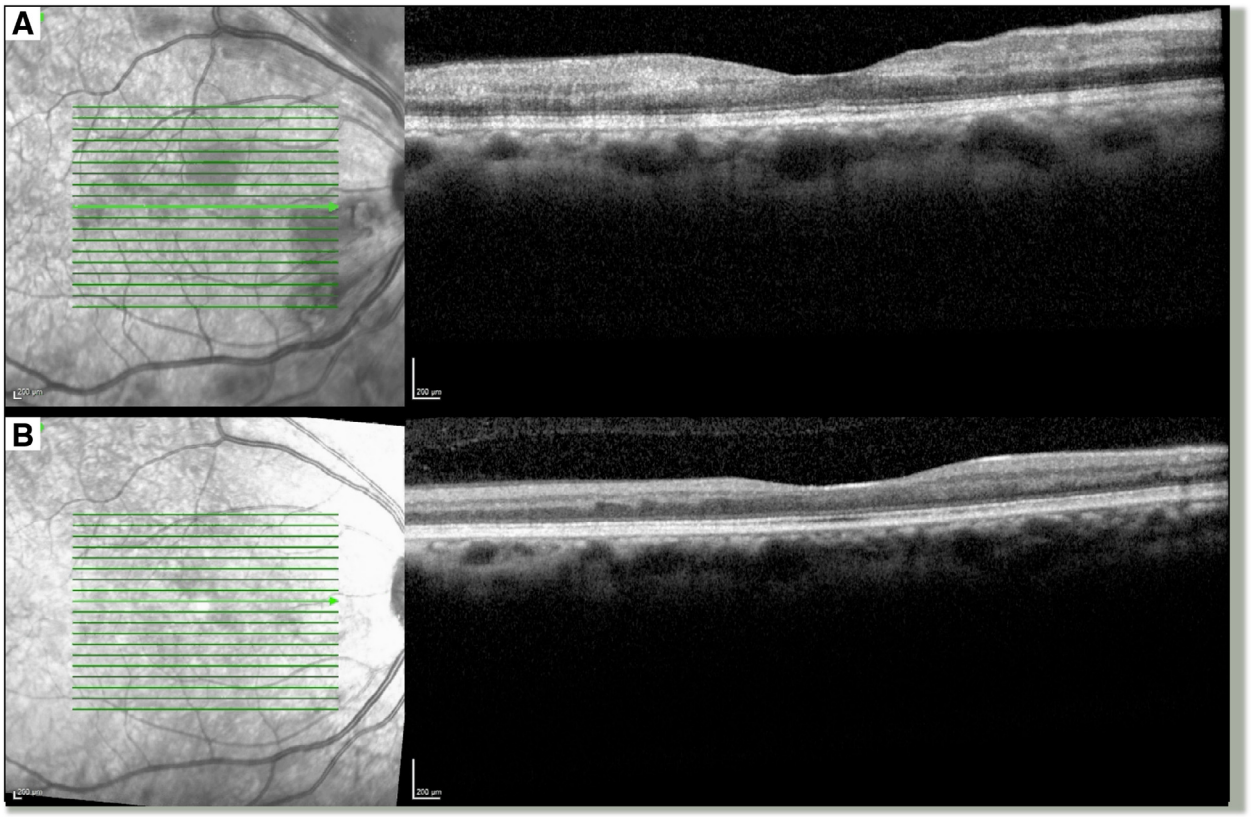

Fig. 2—(A) Ocular coherence tomography (OCT) OD revealed thickening and hyper-reflectivity of the middle macula. (B) OCT OD at 6-month follow-up showed residual parafoveal inner nuclear layer atrophy.

The visual acuity loss in Purtscher-like retinopathy is variable, it can be bilateral or unilateral, and it may occur hours to days after the onset of injury or associated illness. Fundus findings are characterized by cotton-wool spots, retinal hemorrhages, optic disc swelling, and Purtscher flecken. Purtscher flecken are a pathognomonic finding and appear as polygonal areas of retinal whitening with well-defined margins, between arterioles and venules.[3] Management usually involves observation and treatment of the underlying etiology. Other treatments have been reported with variable success, including corticosteroids, papaverine hydrochloride, hyperbaric oxygen, pentoxifylline, and low-molecular-weight heparin. Notwithstanding these anecdotal reports, the benefit over observation has still not been clearly established.[2,7] The visual prognosis can be varied depending on etiology. Agrawal and McKibbin reported that optic disc swelling or leakage and capillary nonperfusion on fluorescein angiography, as well as involvement of the outer retina, are associated with a poor long-term outcome.[3] Miguel et al. also described the male gender, the absence of macular edema, and the absence of a pseudo cherry red spot as good prognostic features.[2] Purtscher-like retinopathy associated with systemic diseases tended toward a worse visual outcome, whereas Purtscher retinopathy from traumatic etiologies tended to have better visual recovery.[2]

The pathogenesis of Purtscher and Purtscher-like retinopathy has been an area of considerable controversy given its lack of a precise etiology. The most accepted theory is from Agrawal and Mckibbin, who suggest that the pathogenesis involves embolic occlusion of the precapillary arterioles.[3] An alternate theory is the complement-mediated hypothesis, where complement C5a initiates leukocyte aggregation and embolization.[3] In our patient, it is postulated that microemboli from the carotid dissection were the source of occlusion. The embolic phenomenon observed in the eye is exceedingly rare after carotid artery dissection, as there is a reversal of flow within the ophthalmic artery to supply the brain during the dissection.[6] On the other hand, microemboli signals in the brain have been reported using transcranial doppler in 46% to 66% of patients with acute internal carotid dissection.[8] Our patient had a good visual recovery at 6 months, which supports the role of observation in these cases.

Supplementary Materials

Supplementary material associated with this article can be found in the online version at doi:10.1016/j.jcjo.2019.08.006.

Footnotes and Disclosure:
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