JAMA Ophthalmology Clinical Challenge

Bilateral Acute Optic Neuropathy in a Teenaged Girl

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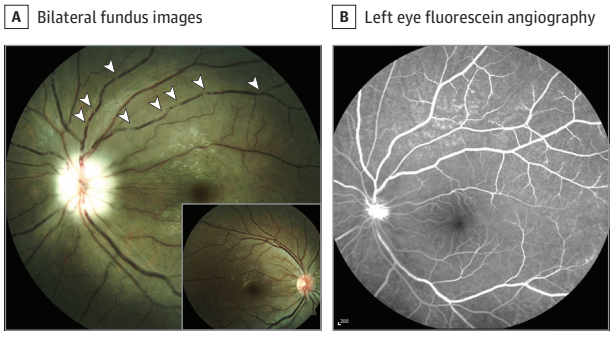

Figure 1. A, Fundus photographs of the left eye and the right eye (inset) showed optic disc pallor with blurred disc margins. The left eye had discontiguous blood flow in the superior retinal vessels (arrowheads). B, Fundus fluorescein angiography of the left eye showed microaneurysms at both the superotemporal and inferotemporal retina.

A 14-year old girl presented with sudden painless vision decline in the left eye for 5 days, without vision decline in the right eye. Her visual acuity was 20/20 OU 6 months earlier. She reported constant fatigue, occasional dizziness and headache, mild pain in the lower limbs, and intermittent claudication over the past 12 months, with no history of trauma, surgery, COVID-19 infection, influenza, or other infection.

Visual acuity was light perception OD and no light perception OS. Both pupils dilated, with the right eye reacting sluggishly to light and the left eye being nonreactive. Intraocular pressure was 14 mm Hg OD and 11 mm Hg OS. Posterior examination revealed optic disc edema in each eye, and the left eye had a swollen and pale optic disc with segmental blood flow in retinal vessels (**Figure 1**A). Fluorescein angiography of the left eye had prolonged arm-to-retina circulation time and multiple microaneurysms at the superotemporal and inferotemporal retina (Figure 1B). Visual evoked potentials were negative in both eyes. She had an increased white blood cell count (10 540/μL; to convert to $\times 10^9$ per liter, multiply by 0.001) and elevated erythrocyte sedimentation rate (73 mm/h) and had positive findings for cyclic citrullinated peptide antibody and antinuclear antibody. Other serology test results were negative, including hepatitis B and C virus, HIV, rapid plasma reagin, and anti–double-stranded DNA.

WHAT WOULD YOU DO NEXT?

- **A.** Perform diagnostic anterior chamber and/or vitreous tap
- **B.** Initiate intravitreal corticosteroids
- **C.** Obtain brain magnetic resonance imaging
- **D.** Arrange a genetic test

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Diagnosis

Takayasu arteritis

What to Do Next

C. Obtain brain magnetic resonance imaging

Discussion

For bilateral optic edema, ocular infection should be considered. The most clinically relevant slitlamp findings of the patient were optic disc edema in both eyes and abnormal pupil reaction to the light with no sign of infection. Her blood test results did not suggest systemic infection; therefore, a diagnostic intravitreal tap (choice A) would not be a preferred next step. Providing intravitreal corticosteroids (choice B) would not address the systemic symptoms, and no obvious retinal vascular inflammatory abnormalities were noted on fluorescein angiography. The patient had no family history of inherited genetic disorders to warrant genetic testing (choice D).

The patient reported fatigue, occasional dizziness and headache, mild pain in her lower limbs, and intermittent claudication and had findings of elevated erythrocyte sedimentation rate, cyclic citrullinated peptide antibody, and antinuclear antibody, suggesting the need for brain magnetic resonance imaging (MRI) (choice C). Brain MRI showed an abnormally high signal intensity in the left periventricular white matter (**Figure 2**) and was suspicious for a left middle cerebral artery infarct. Since MRI showed features consistent with a demyelinating disorder or local cerebral infarction in the left periventricular white matter, suggesting a systemic disease rather than optic neuritis, urgent referral to rheumatology was recommended for further investigation of systemic autoimmune diseases. The rheumatologist suspected Takayasu arteritis and arranged magnetic resonance angiography (MRA) and arterial angiography for the head and neck. Both MRA and arterial angiography showed left common carotid artery and internal carotid artery occlusion, with right innominate artery and right common carotid artery stenosis, distal arterial occlusion, and mul-

tiple collateral arteries to the brain parenchyma (Figure 2), confirming a diagnosis of Takayasu arteritis. Subsequently, 400 mg of prednisone was given intravenously for 3 days. After 1 day, visual acuity improved to 20/32 OD and remained substantially impaired at hand motions OS, without further improvement.

Takayasu arteritis is a rare autoimmune disease associated with inflammation of large vessels, such as the aorta and its main branches. It is known as pulseless disease and occurs commonly in young Asian women younger than 40 years.[1] Diagnosis of Takayasu arteritis may be challenging with lack of specific clinical symptoms and sensitive laboratory tests.[2] It is usually asymptomatic in the early stage, gradually proceeding to ischemic complications. Chronic ischemia can be associated with various clinical presentations, depending on the pattern of arterial involvement.[3,4]

Only approximately 9.6% cases present with vision loss as an initial clinical feature.[5] A comprehensive medical history and detailed physical examinations could provide valuable information to direct subsequent investigations. Diagnostic criteria for Takayasu arteritis consist of findings from imaging, laboratory tests, and physical examinations, eg, subclavian artery lesion, elevated erythrocyte sedimentation rate, and carotid artery tenderness.[6] On diagnosis, prompt and aggressive treatment using glucocorticoid therapy along with conventional immunosuppressive agents should be initiated, and surgical revascularization may be considered.[7,8] Long-term follow-up is recommended, especially for younger patients, due to a high relapse rate.[9]

Patient Outcome

After approximately 4 weeks of treatment, the patient's dosage of glucocorticoid had been reduced to daily oral methylprednisolone acetate, 20 mg, along with oral methotrexate, 10 mg once per week. Her visual acuity remains 20/32 OD and counting fingers OS, and her physical symptoms have resolved.

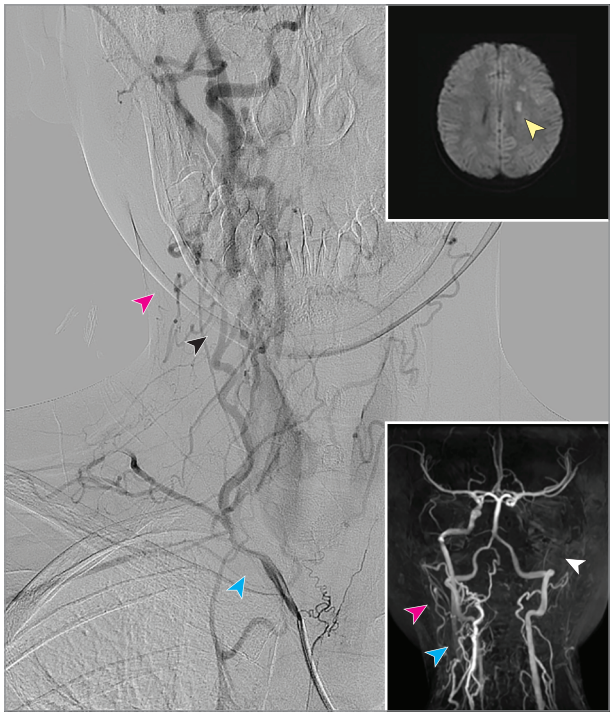

Figure 2. Diffusion-weighted magnetic resonance imaging (upper inset) suggested a suspicious demyelinating disorder or local cerebral infarction (arrowhead). Head and neck arterial angiography showed that a right innominate artery was stenosed (blue arrowhead), the right common carotid artery was narrowed with distal arterial occlusion (black arrowhead), and multiple collateral arteries had developed (red arrowhead). Magnetic resonance angiography (lower inset) showed that the left common carotid artery and the right distal common carotid artery were occluded (white arrowhead), the right distal common carotid artery was occluded (blue arrowhead), and multiple collateral arteries had developed (red arrowhead).

ARTICLE INFORMATION

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