JAMA Ophthalmology Clinical Challenge

Monocular Vision Loss in a Patient With Earache and Night Sweats

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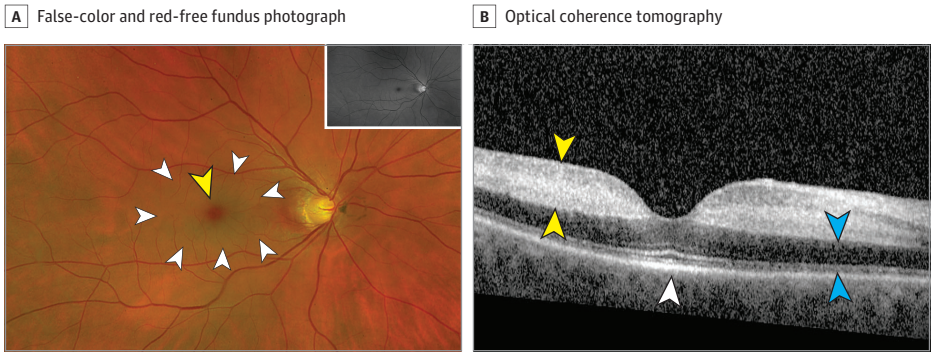

Figure 1. False-color and red-free (inset) fundus photograph of the right eye shows retinal pallor (white arrowheads) with a cherry red spot (yellow arrowhead) in the macula (A). Optical coherence tomography of the right macula shows hyperreflectivity and thickening of the inner retinal layers (yellow arrowheads) and hyporeflectivity of the outer retinal layers (blue arrowheads), except for increased signal intensity in the outer retinal layers of the fovea (white arrowhead) (B).

A 62-year-old patient presented to the ophthalmology department with sudden painless vision loss in her right eye that occurred 24 hours ago. She also reported a 4-week history of right ear pain and fullness, discomfort over her frontal and maxillary sinuses, and nasal congestion without discharge. Over the past 2 weeks, she had experienced drenching night sweats, cramps in her quadriceps, and weight loss of 3 kg. She denied respiratory symptoms, headaches, scalp tenderness, or jaw claudication. She took no regular medication and did not smoke.

At presentation, her visual acuities were perception of light OD and 20/20 OS. There was a right relative afferent pupillary defect. Dilated fundus examination showed a cherry red spot in the right macula with hyperreflectivity and thickening of the inner retinal layers on optical coherence tomography (**Figure 1**A and B). External eye examination results and ocular motility were normal. Examination by an otolaryngologist revealed normal tympanic membranes and a dry but otherwise normal nasal mucosa on flexible endoscopy. Blood tests showed leukocytosis (white blood cell count, 15 000 /μL; to convert to 10^9 per liter, multiply by 0.001) with neutrophilia (white blood cell count, 12 500 /μL), elevated C-reactive protein (19.4 mg/dL; to convert to milligrams per liter, multiply by 10), elevated plasma viscosity (2.06 mPas), and thrombocytosis (467 ×10^3/μL; to convert to ×10^9 per liter, multiply by 1). Kidney function was within normal limits. Chest radiograph results were unremarkable.

WHAT WOULD YOU DO NEXT?

A. Request a magnetic resonance image of orbits and sinuses

B. Commence high-dose corticosteroids

C. Commence broad-spectrum antibiotics

D. Request urgent stroke consult

Quiz at jamacmelookup.com

Diagnosis

Granulomatosis with polyangiitis

What to Do Next

B. Commence high-dose corticosteroids

Discussion

The presence of a central retinal artery occlusion (CRAO) in the context of sinonasal symptoms and elevated inflammatory markers in a middle-aged patient should raise suspicion of a vasculitic process. Granulomatosis with polyangiitis (GPA), previously known as Wegener granulomatosis, is a necrotizing vasculitis that typically affects patients older than 50 years and is characterized by the presence of antineutrophil cytoplasmic antibodies against proteinase 3.[1,2] The condition has a predilection for small- and medium-sized vessels of the upper and lower respiratory tract, frequently mimicking infective sinusitis, mastoiditis, or otitis.[2] Almost 50% of patients with GPA will develop ocular or orbital involvement.[3] Vision loss may occur due to compressive optic neuropathy from granulomatous inflammation at the orbital apex or, rarely, as a result of an occlusive retinal vasculitis or CRAO.[3,4] Magnetic resonance imaging of the orbits and sinuses (choice A) may show granulomatous orbital inflammation, sinonasal mucosa thickening, or osseous destruction in advanced disease.[5] However, these features are not specific for GPA and need to be correlated with clinical signs and laboratory tests.[3,5] Imaging or further blood tests (eg, neutrophil cytoplasmic antibodies) should not delay the commencement of high-dose corticosteroids (choice B) if a vasculitic process is suspected as immediate im-

munomodulatory treatment reduces morbidity, mortality, and relapses of GPA and other vasculitides.[6] The mainstay of treatment for organ-threatening GPA remains high-dose intravenous methylprednisolone, followed by oral corticosteroids and steroid-sparing agents such as cyclophosphamide or rituximab.[6-8]

The combination of elevated inflammatory markers, neutrophils, and upper respiratory tract symptoms may be suggestive of an infective process. Bacterial or fungal sinusitis can result in sight-threatening septic thrombophlebitis or cavernous sinus thrombosis, but vision loss due to an isolated CRAO is rare.[9] Importantly, this patient did not have proptosis or limited eye movements indicative of an orbital pathology. In this situation, starting broad-spectrum antibiotics (choice C) without confirming an orbital infection on computed tomography is not recommended.

Patients with a nonarteritic CRAO should have an urgent stroke consult (choice D) for consideration of intravenous tissue plasminogen activator, usually if presenting within 4.5 hours of vision loss.[10] This patient had no vascular risk factors and no visible embolus on fundus examination (Figure 1) and elevated inflammatory markers, thus making an inflammatory etiology more likely. Arteritic CRAO is most often due to giant cell arteritis but can be caused by other vasculitides.[10] In these circumstances, referral to a stroke specialist should not delay further inflammatory workup alongside administration of high-dose corticosteroids to reduce the risk of further sight loss.

Patient Outcome

This patient had a positive anti–proteinase 3 antineutrophil cytoplasmic antibodies and pulmonary nodules on computed tomography imaging of the chest (**Figure 2**), confirming a diagnosis of GPA. Treatment with intravenous methylprednisolone improved her constitutional and sinus symptoms and her visual acuity from perception of light to 20/125 OD. She subsequently developed a large pulmonary embolism, which was successfully thrombolyzed. Currently, she is receiving cyclophosphamide infusions for GPA and takes apixaban for ongoing anticoagulation.

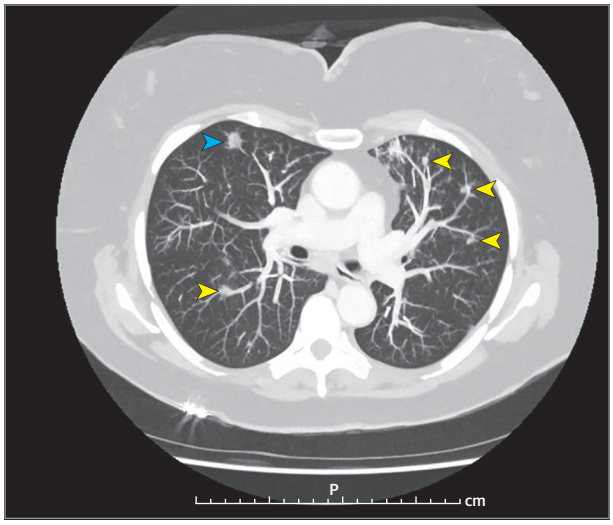

Figure 2. Postcontrast computed tomography image of the chest and lung window (8-mm maximum intensity projection slice) demonstrates multiple small bilateral lung nodules involving multiple lobes (yellow arrowheads). A larger, more focal nodule (blue arrowhead) is seen anteriorly within the right upper lobe.

ARTICLE INFORMATION

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