

Medical Mystery: Visual-Field Defect — The Answer

TO THE EDITOR: The medical mystery in the October 12 issue¹ involved a 31-year-old woman who presented with a sudden onset of an inferonasal visual-field deficit in the left eye. Two days earlier, she had traveled for 16 hours by air. On physical examination the day after the onset of

her visual symptoms, ophthalmoscopy revealed a band of opacification in the left macula that was diagnostic of a branch retinal artery occlusion, as shown on fluorescein angiography (Fig. 1A). The patient had digital erythema, cyanosis (Fig. 1B), and severe clubbing (Fig. 1C) of the hands and feet. Auscultation revealed a systolic ejection murmur over the right upper sternal border. The hematocrit was 57.4%.

The patient had Eisenmenger's syndrome associated with a ventriculoseptal defect and a patent ductus arteriosus. Anticoagulation was initiated with heparin, and she was discharged while receiving warfarin. In this case, the long airplane flight and secondary polycythemia with hypercoagulability probably led to clotting in the lower limbs, with embolism through the patent ductus arteriosus to the retina. The visual-field defect subsequently resolved.

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1. Mathura JR Jr, Jampol LM. Medical mystery — visual-field defect. *N Engl J Med* 2006;355:1592.

Editor's note: We received 986 responses to this medical mystery — 52% from physicians in practice, 26% from physicians in training, 15% from medical students, and 7% from other readers. Responses were received from 65 countries. Forty-four percent of the respondents correctly identified this condition as an intracardiac defect with a paradoxical embolism causing a branch retinal artery occlusion. Twenty percent of the respondents suggested polycythemia vera, 10% suggested left-sided valvular abnormalities as seen with infectious endocarditis or a rheumatologic condition (e.g., Libman-Sacks disease or the antiphospholipid antibody syndrome), and the remaining 26% suggested a variety of diagnoses, including the hyperviscosity syndrome, papilledema, Marfan's syndrome, and emboli such as cholesterol, fat, or air. The clubbing and polycythemia suggest a diagnosis of chronic hypoxemia. When these two findings are combined with the long plane flight, a cardiac murmur, and a branch retinal artery occlusion, a paradoxical embolic event must be strongly considered.

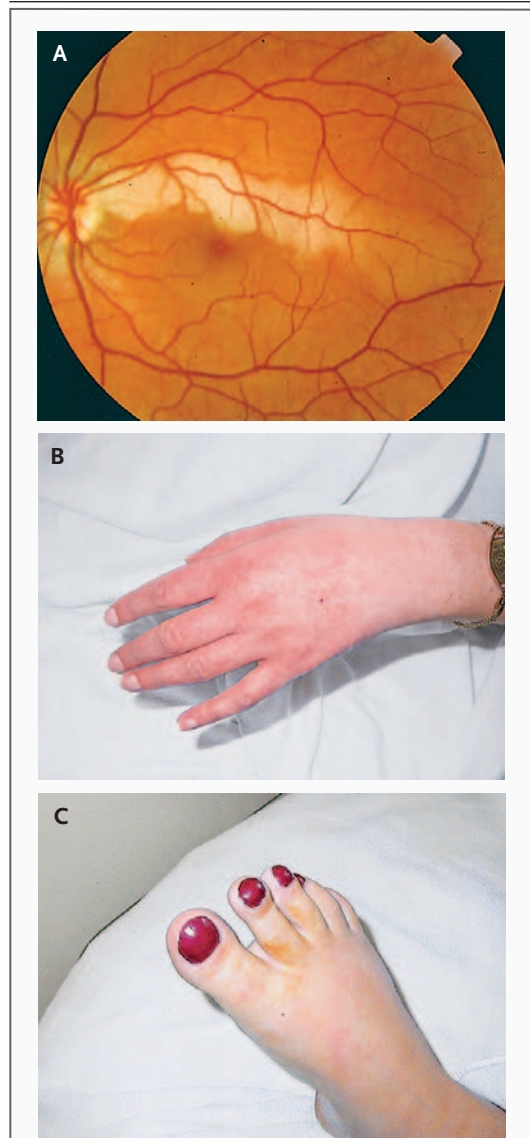


Figure 1. Eisenmenger's Syndrome Associated with a Ventriculoseptal Defect and a Patent Ductus Arteriosus.

Fluorescein angiography reveals a branch retinal artery occlusion (Panel A). Photographs show cyanosis of the hands (Panel B) and severe clubbing of the toes (Panel C).