A 35-Year-Old Male with Blurred Vision and Metamorphopsia

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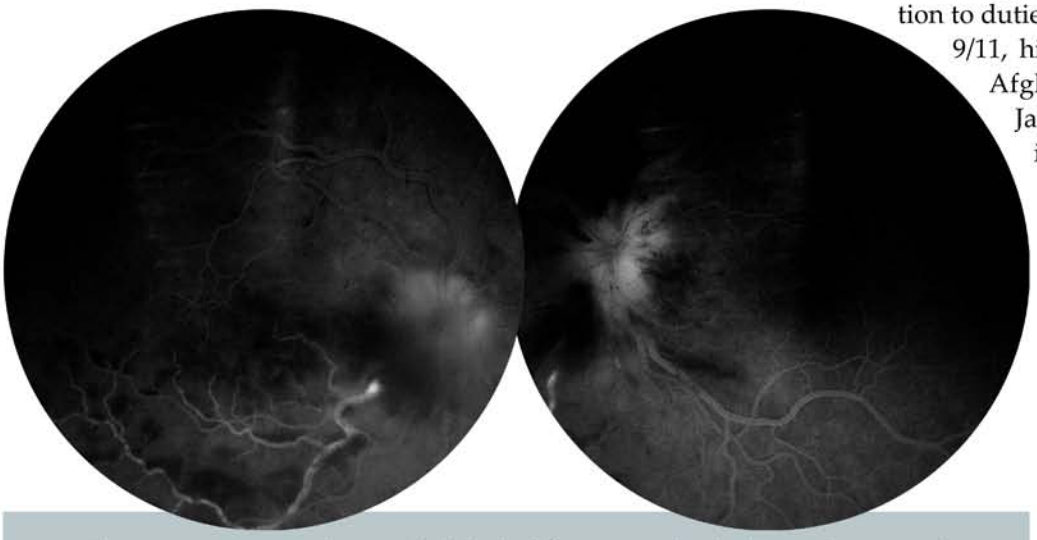
Introduction:

A 35-year-old, Caucasian male presented with a one month history of blurred vision in the right eye, and metamorphopsia in the left eye for three days. The patient had been experiencing severe headaches and night sweats for several months. A thorough medical evaluation including an infectious disease consult, MRI, and spinal tap were non-diagnostic. He was prescribed Neurontin for his headaches.

Upon presentation, his vision was hand motions in the right eye and 20/20 on the left. The anterior segments were normal, and there were 1+ vitreous cells bilaterally. The retina was remarkable for patchy hemorrhages, cotton-wool spots, and disc edema in both eyes.



Fluorescein angiography revealed blocked florescence by the hemorrhage noted clinically, disc staining, and some segmental vasculitis bilaterally.

Diagnosis:

This patient had a long career in the Marines. In addition to duties at ground zero in NYC after 9/11, his tours included missions to Afghanistan, Iran, Okinawa, Japan, Indonesia, and Djibouti in the horn of Africa.

No evidence of hematologic, pulmonary, or renal disease was found. Serology ruled out hepatitis, syphilis, toxoplasmosis, Cryptococcus, Lyme disease, and drug abuse. The sole laboratory abnormality was an elevated C-reactive protein of 46 (normal < 0.3). It was suspected that the patient's headaches, night sweats, and severe

posterior segment abnormalities were related. The diagnosis of Behcet's was entertained. Upon more thorough questioning, the patient over the previous two years experienced four scrotal ulcers and several oral ulcers, which resolved spontaneously. Since the diagnosis of Behcet's disease, the patient has been treated for a deep vein thrombosis of the leg and intracranial hypertension.

Discussion:

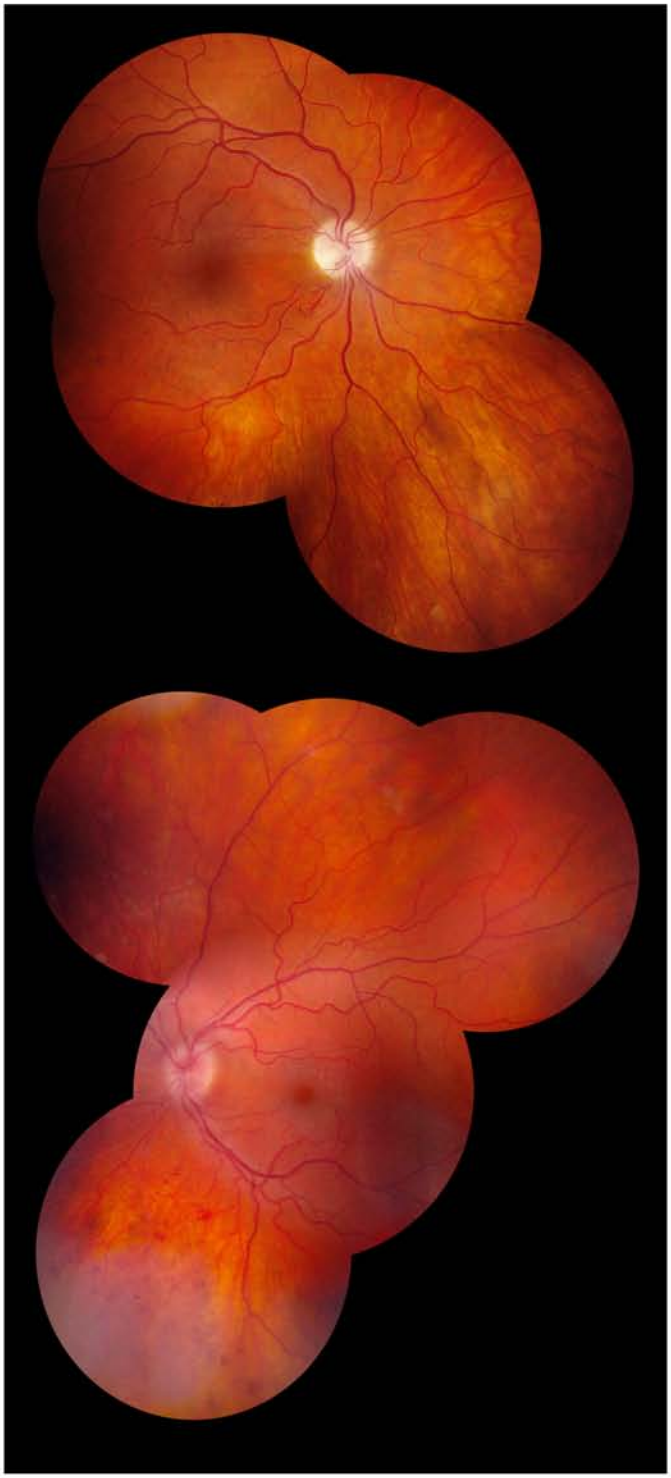
Although Hippocrates in the fifth century B.C. described a patient with classic Behcet's-like symptoms, this eponymous condition is named for a Turkish dermatologist who characterized the constellation of signs and symptoms in 1937. Behcet's is seen worldwide, but it is most common in the eastern Mediterranean and eastern rim of Asia. The highest prevalence is in Turkey with 100 per 100,000 affected. In the United States, the prevalence is 4 per 1,000,000 inhabitants. There is no apparent sex predilection. The mean age of patients is 25 to 35 years of age. The range is two months to 75 years. Familial cases have been reported, including a pair of monozygotic brothers. There is no spousal transmission, nor is there a consistent inheritance pattern.

Diagnostic criteria proposed by the International Study Group for Behcet's Disease include oral aphthous ulcers (at least three per year), plus two of the following: recurrent general ulcers, ocular inflammation, skin involvement, or a positive pathergy test. Acute phase reactants (sed rate, C-reactive protein) are often elevated. Recurrent painful oral ulcers are common, and they occur anywhere in the oral cavity. Genital ulcers, dermatographia, and erythema nodosum are typical. Vascular disorders are widespread and potentially life-threatening and include deep vein thrombosis, aneurysms, arteriolar occlusions, myocardial infarction, and arrhythmias.

In addition to headaches, neuro-Behcet's may lead to seizures, motor abnormalities, cranial motor palsy, pseudotumor, confusion, and hallucinations. Gastrointestinal ulcers may result in perforation. Pulmonary arteritis can lead to embolism and infarction. Ocular Behcet's occurs in 90% of men and 70% of women affected. It is bilateral in 80%. The second eye is usually affected within one year. Ocular Behcet's usually follows genital and oral lesions by a few years. A non-granulomatous iritis results in hypopyon in one-third of patients. Vitritis may be mild to severe. An obliterative necrotizing retinal vasculitis may affect the arteries as well as veins. Untreated progressive optic atrophy is common. Treatment with aggressive corticosteroids and cytotoxic agents is recommended.

Treatment:

Azathioprine, Imuran, and recently Remicade infusions. Two and a half years after presentation, our patient's vision remained light perception in the right eye and 20/25 in the left.



Conclusions:

Behcet's is a chronic inflammatory disorder characterized by exacerbations after long periods of remission. The etiology is unknown and there are no universally accepted diagnostics. The triad of retinal vasculitis, oral and/or genital ulcers, and skin lesions are the typical signs and should raise suspicions. Prompt and aggressive treatment combined with attentive surveillance and monitoring are critical to minimize morbidity and mortality.

References:

Colvard DM, Robertson, DM, and O'Duffy, JD. The Ocular Manifestations of Behcet's Disease. Arch Ophth 95:1813-1817, 1977

Chajek T and Fainaru, M. Behcet's Disease: Report of 41 Cases and a Review of the Literature. Medicine 54:179-196, 1975

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