Bilateral Posterior Uveitis

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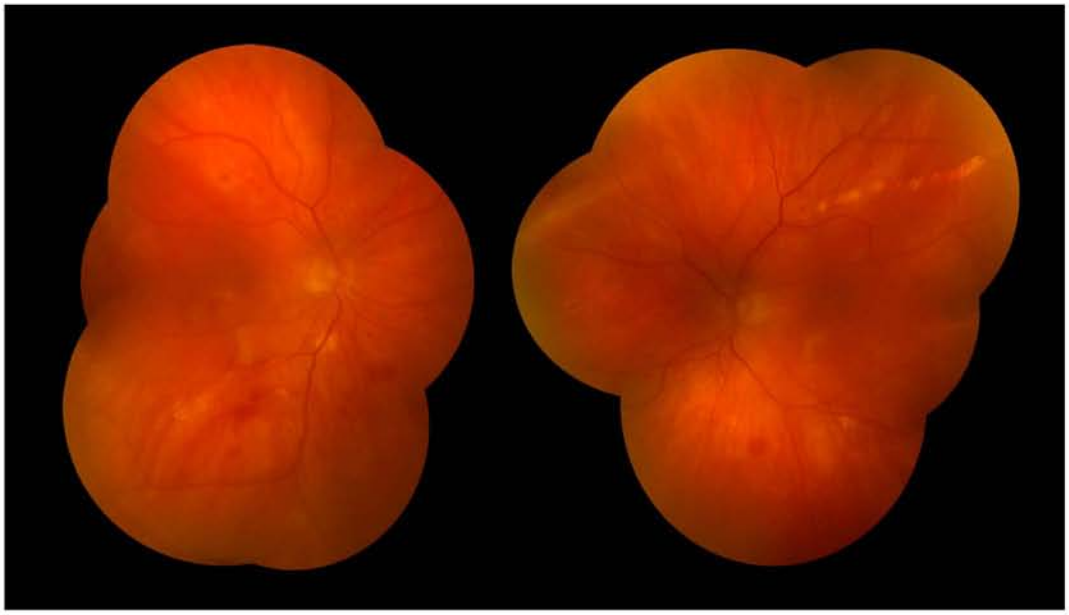
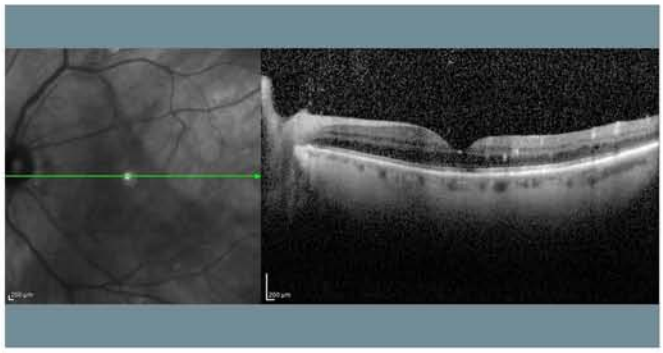
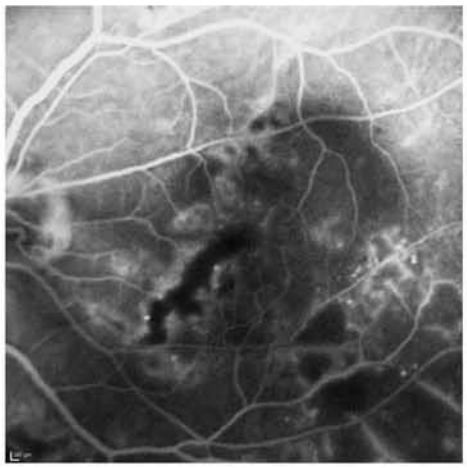
A 71 year old white woman presented to The Retina Institute with bilateral gradual decrease in vision for three months. Her ocular history was unremarkable. Her medical history was significant for type 2 diabetes mellitus and a recent hospitalization for a flu-like illness three months prior.

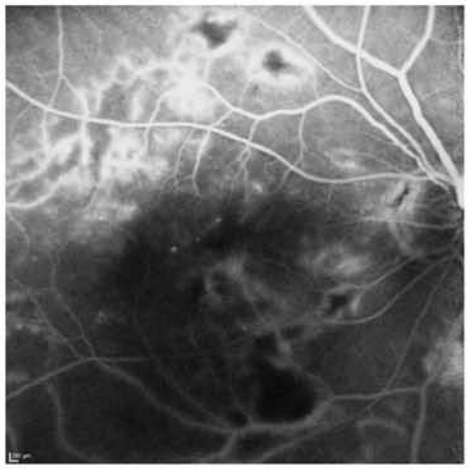


Visual acuity measured 20/50 in the right eye and 20/70 in the left. No relative afferent papillary defect was present. Intraocular pressures were 21 and 18 in the right and left eyes, respectively. Extraocular motility, confrontational visual field, and adnexal examination were within normal limits. Anterior segment revealed nuclear sclerosis without any anterior inflammation.



Posterior segment examination showed no evidence of vitritis or optic nerve edema. Microaneurysms and hard exudates were scattered throughout the posterior poles. Deep, circumscribed, creamy lesions were evident in both eyes, predominantly in a segmental distribution in the inferotemporal quadrant of the right eye and in a linear distribution along the superotemporal arcade of the left eye. OCT revealed preservation of inner and outer retinal architecture with and mild intraretinal exudates. Fluorescein angiography demonstrated late staining of the superotemporal linear lesions in the left eye and central hypofluorescence and peripheral hyperfluorescence of the other scattered lesions. Patches of capillary nonperfusion were also evident.





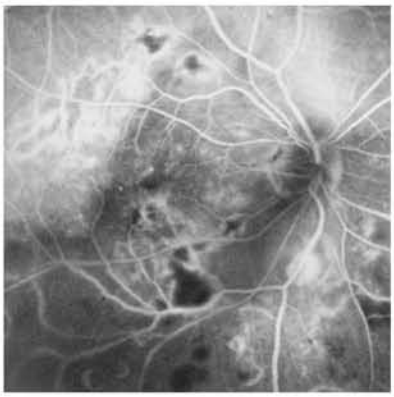
Upon further review of medical records, the patient one month inpatient hospitalization was complicated by two week ICU course for viral meningitis secondary to West Nile Virus. The clinical scenario along with posterior segment manifestations led to a diagnosis of West Nile Virus chorioretinitis.

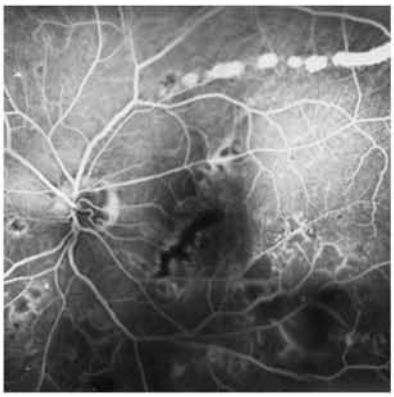
Discussion

◆ West Nile Virus (WNV) is a single stranded RNA virus from the flavivirus genus which also includes dengue, Japanese encephalitis virsus and Saint Louis encephalitis virus. After first isolated in 1937 in Uganda, outbreaks spread across North America in the 2000s and currently WNV is one of the most significant causes of viral encephalitis in the US. WNV follows a bird-mosquito-mammal transmission cycle with wild birds as the primary natural host and Culex species of mosquitos as the transmission vector to humans and horses. Other modes of transmission have been reported, including blood transfusion, organ transplantation, transplacental transmission, and breast feeding. WNV is spread both sporadically and in outbreaks, occurring mainly in late summer.

◆ The vast majority of WNV cases are asymptomatic. When symptomatic, a self-limited flu-like illness is present with high grade fever, headache, myalgias, arthraligas, malaise, nausea, vomiting, diarrhea, rash, weakness, pharyngitis, lymphadenopathy. WNV can enter the CNS through direct infection via blood-brain barrier breakdown, transport by infected immune cells, or retrograde transport along peripheral nerves, causing a severe, neurologic disease with flaccid paralysis, meningitis, encephalitis can occur, carrying up to 10% mortality.

◆ Ocular disease occurs in up to 80% of cases most commonly manifesting a bilateral multifocal chorioretinitis. Lesions develop early in the disease course as circular, deep, creamy white lesions with early hypofluorescence and late staining. Inactive chorioretinal lesions appear as round atrophic with or without central pigmentation, usually "target like appearance" on FA with central hypofluorescence and peripheral hyperfluorescence. Most lesions are in the posterior pole or midperiphery with a notable linear clustering of 1 or more clusters per eye. The linear clustering is in a radial or curvilinear pattern suggesting contiguous spread from CNS and optic nerve head. Indocyanine green angiography often shows more lesions than clinical exam and fluorescein angiography. Other manifestations include anterior uveitis, retinal vascilitis, optic neuritis, and cranial nerve palsy. The differential diagnosis includes causes of typical causes of noninfectious posterior uveitis (multifocal choroiditis, ocular histoplasmosis syndrome, sarcoid, birdshot chorioretinitis) and infectious choroiditis (syphilis, tuberculous, HSV1/2, VZV) as well as disease processes based on endemic risk factors (coccidiomycosis, Rift Valley fever, Behcet's disease).

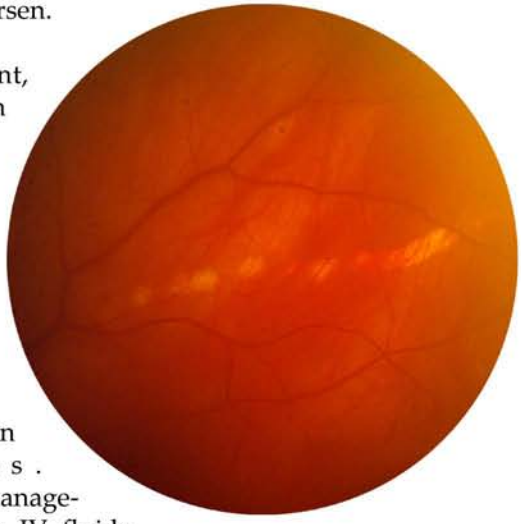




◆ The diagnosis of WNV is largely based on a syndrome with a typical constellation of symptoms. A history of mosquito bites is helpful. Laboratory confirmation includes MAC-ELISA (IgM Antibody Capture-ELISA) which identifies WNV-specific IgM antibody in serum and CSF. This is most accurate within 8-21 days after onset of symptoms and serum IgM can persist for 6 months or longer yielding higher false positives in asymptomatic endemic patients. PCR is highly specific for WNV infection, but not as sensitive as serological testing, thus often reserved for screening. CBC may reveal leukocytosis, lymphocytopenia, thrombocytopenia, mild anemia and CSF analysis may show lymphocytic predominant pleocytosis with elevated protein concentration.

◆ The overall systemic disease prognosis is good in most patients but neurologic disease carries 10% mortality and in comorbid and debilitated patients the mortality can be up to 20%. Ocular involvement usually has a self-limited course with visual acuity often returning

to baseline. Ocular complications include macular scarring, choroidal neovascularization, and sequealae of occlusive retinal vasculitis (peripheral neovascularization, vitreous hemorrhage, macular ischemia, optic atrophy). The prognosis has been demonstrated to be worse in patients older than 50 years old and with concurrent diabetic retinopathy, which can worsen.



◆ At present, no proven treatment exists. Prevention includes mosquito repellent in endemic regions and vaccine development in progress. Supportive management includes IV fluids, respiratory support, and prevention of secondary infections. Reports of ribavirin, INF-2b, anti-WNV Ig passive transfer have shown to be effective. Ophthalmic treatment includes topical steroids for anterior uveitis, and management of ocular complications, including peripheral laser for occlusive vasculitis, anti-VEGF therapy for CNV.

REFERENCES:

Foster CS, Vitale A. 2012. Diagnosis and Treatment of Uveitis Second Edition. Jaypee Brothers Medical Publishers, New Delhi, India.

Garg S, Lee JM. Systemic and Intraocular Manifestations of West Nile Virus Infection. Surv Ophthalmol 2005;50:1-13.

Khairallah M, Ben Yahia S, Ladjima L, et al. Chorioretinal involvement in patients with West Nile virus infection. Ophthalmology 2004;111:2065-70.

Nussenblatt RB, Whitcup SM. 2010. Uveitis: Fundamentals and Clinical Practice, Fourth Edition. Elsevier.

The Retina Institute participates in numerous national clinical trials. Visit the Studies section on our website at tri-stl.com for information regarding these trials and patient enrollment.