

Modern Imaging Impacts Retinal Vasculitis Treatment Approach

JOHN HUANG MD, YALE UNIVERSITY

Thomas Albin, MD, moderator of the Uveitis Resource Center, recently spoke with John Huang MD, associate professor of ophthalmology specializing in vitreal retinal diseases and uveitis, at Yale University, about his approach to retinal vasculitis treatment. Their conversation follows:

DIAGNOSIS AND IMAGING

Thomas Albin, MD: What are your preferred imaging modalities for diagnosing retinal vasculitis?

John Huang MD: We commonly use fluorescein angiography to diagnose and follow these patients. It helps us see how much inflammation there is, whether vasculitis is present and if there is ischemia or complete vascular damage. We also use Optos wide-field, high resolution imaging to evaluate the extent of the disease and assess the risk of neovascularization and hemorrhage.

TA: Are there scenarios where you order an ICG angiography for vasculitis?

JH: When a patient has unexplained vision loss that is even more significant than what fluorescein shows us, ICG angiography may be helpful. For instance ICG angiography imaging and assessment may be helpful in the case of lupus patients, pre-eclampsia patients or patients who have extensive vascular inflammation that might involve cortical inflammation.

TA: In terms of your differential diagnosis, what infectious causes are considered and tested for?

JH: Syphilis is always at the top of our infectious differential, and we always rule out tuberculosis, as well. If we see retinal edema, we test for herpes. Also, here, in Connecticut, we see lyme-associated uveitis a lot, so we often send out for lyme titers; and when we see vascular inflammation we test for Bartonella disease.

NON-INFECTIOUS CAUSES

TA: Do you think it's helpful to separate retinal vasculitis into three groups: primarily arteritis vs phlebitis vs a combination of those two; and if so how does that shift your differential?

JH: When working with residents and fellows we always emphasize the importance of first identifying which vessels are affected. We ask is it a para-phlebitis, is it an arteritis or does it affect both the arteries and veins? Especially with respect to vascular inflammation, if it is predominately para-phlebitis my top line differential usually involves things such as sarcoid or even Eals disease.

With respect to pure arteritis, the things that we naturally think of are conditions such as lupus or Wegener's. Those conditions are more associated with systemic conditions. In that kind of situation, we usually manage the patient aggressively, and we initiate our work-up toward those conditions, as well.

TA: What about when a combination of arteries and veins are involved?

JH: In a patient with a combination artery and vein retinal vasculitis, we consider multiple sclerosis, Wegener's and sometimes lupus.

TA: There are patients whom we consider as having an intermediate uveitis who have capillitis or leakage around the vessels inferiorly. Are there any other conditions that come to mind with that type of finding?

JH: Most posterior uveitis will have some sort of leakage. We have patients with Behçet's disease who sometimes will have these mild signs, as well.

TA: Intermediate uveitis occurs often in children and has one of the best prognoses, but the treatment options have some serious risks. If you have a patient with no anterior chamber cell, no cystoid macular edema and pretty good vision, but they have capillitis how much do you tolerate before you treat?

JH: Making the decision to put children on immunosuppressive therapy or aggressive treatments, such as methotrexate or giving them sub-tenons injections -- with their associated risk of glaucoma and cataract -- is very difficult. My treatment tolerance is high for pediatric patients, especially if their vision is 20/20 and they have only a little vascular inflammation in the periphery. In those cases, I'm willing to just watch it because it's not going to cause any vision damage. On the other hand, if the vascular inflammation is much more focal -- towards the optic nerve or towards the fovea -- where there is more of a threat of vision loss, I monitor them more carefully, and my threshold for treating them is lower.

In children who have retinal vasculitis disease, such as Coats' disease, we commonly follow them every three to four months with wide field angiography to make sure that the disease is not progressing. For even younger children, who can be difficult to follow, we typically use the Retisert camera right in the office to evaluate peripheral retinal vascular inflammation.

TA: What is your standard treatment for non-infectious retinal vasculitis?

JH: We usually start patients on .5 mg to 1 mg per kg of an oral prednisone and taper from one to three months. If the patient has a mild recurrence, we continue to monitor them; if they have a significant recurrence I discuss long term, immuno-modulation therapy with them.

TA: When you taper oral prednisone and the vasculitis returns, what immuno-modulators or anti-metabolites do you use?

JH: Methotrexate is at the top of my list. It's an excellent drug and easy to use, but it doesn't work on everybody so we sometimes do combination therapy with a calcineurin inhibitor, such as cyclosporine. Occasionally, we switch from methotrexate to hypotthalmate, as well.

TA: Once you see that a retinal vasculitis patient does not have a problem with oral steroids, do you ever consider local steroid therapy?

JH: When a patient has vasculitis that is not associated with any systemic disease, local steroids are a reasonable option. This is especially true in patients with localized eye disease that is idiopathic. I am very comfortable doing sub-tenons Triamcinolone or intravitreal Ozurdex injections in these patients.

TA: Do you ever treat these patients with IV steroids or alkylating agents?

JH: When we see patients with retinal vasculitis who have an associated vision- or life-threatening systemic disease, we admit them for IV steroids -- 1 gram per day of Solu-Medrol (Methylprednisolone). The standard of care for those with Wegener's granulomatosis is an alkylating agent, and sometimes Lupus patients require an alkylating agent such as chlorambucil therapy for long-term control. ■