



THE OCULAR IMMUNOLOGY
AND UVEITIS FOUNDATION

Dedicated to Eye Disease Cure and Education

Retinal Vasculitis: It's Significance

Arash Maleki, MD; C. Stephen Foster, MD, FACS, FACR

Revised April 2026

Retinal vasculitis is a potentially blinding condition characterized by inflammation of the retinal blood vessels. The annual incidence is estimated at 1 to 2 cases per 100,000 in the United States. Retinal vasculitis may be found in infectious, noninfectious, neoplastic uveitis and can be associated with systemic inflammatory disease.

In contrast to uveitis without retinal vasculitis, in which approximately 30% of cases may be idiopathic, uveitis associated with retinal vasculitis is less likely to be idiopathic. Furthermore, the underlying cause of retinal vasculitis is more likely to be systemic; however, ocular findings may be the first manifestation of systemic diseases that can be life-threatening and Over time, extraocular manifestations emerge, making the nature of the underlying systemic disease more apparent. Polyarteritis nodosa (PAN), Granulomatosis with polyangiitis (GPA), Systemic lupus erythematosus (SLE), Sarcoidosis, Multiple sclerosis (MS), Relapsing polychondritis (RPC), and Behçet's disease, as well as infections such as Syphilis, Lyme disease, and Tuberculosis (TB), are examples of diseases that can behave in this way

Furthermore, the de novo onset of retinal vasculitis in a patient with a well-established systemic disease is clinically significant. For example, a patient with well-characterized Behçet's disease, presenting with oral and genital ulcers, arthritis, and erythema nodosum, may be well controlled on twice-daily colchicine and low-dose prednisone. The development of retinal vasculitis, however, carries significant clinical implications. This indicates that the aforementioned therapeutic regimen will no longer be sufficient for this patient. The appearance of retinal vasculitis signals that the underlying nature of the patient's Behçet's disease has changed. Unless treatment is intensified, the patient is at risk of bilateral blindness within four years and has an approximately 30% risk of developing central nervous system vasculitis. Similar patterns are observed in patients with SLE, Relapsing polychondritis, GPA, and PAN. This concept is also relevant to idiopathic forms of retinal vasculitis.

The significance of retinal vasculitis, therefore, is considerable, both in terms of the likelihood of identifying an underlying systemic disease and as a marker of subclinical vasculitic involvement that may be life-threatening if not treated more aggressively.

References

- 1) Maleki A, Cao JH, Silpa-Archa S, Foster CS. VISUAL OUTCOME AND POOR PROGNOSTIC FACTORS IN ISOLATED IDIOPATHIC RETINAL VASCULITIS. *Retina*. 2016;36(10):1979-85.
- 2) Maleki A, Ueberroth JA, Walsh M, Foster F, Chang PY, Anesi SD, Foster CS. Combination of Intravenous Methotrexate and Methylprednisolone Therapy in the Treatment of Severe Ocular Inflammatory Diseases. *Ocul Immunol Inflamm*. 2021;29(7-8):1559-1563.
- 3) Anesi SD, Chang PY, Maleki A, Manhapra A, Look-Why S, Asgari S, Walsh M, Drenen K, Foster CS. Effects of Subcutaneous Repository Corticotropin Gel Injection on Regulatory T Cell Population in Noninfectious Retinal Vasculitis. *Ocul Immunol Inflamm*. 2023;31(3):556-565.
- 4) Maleki A, Garcia CM, Asgari S, Manhapra A, Foster CS. Response to the Second TNF- α Inhibitor (Adalimumab or Infliximab) after Failing the First One in Refractory Idiopathic Inflammatory Retinal Vascular Leakage. *Ocul Immunol Inflamm*. 2022;30(5):1099-1108.
- 5) Anesi SD, Chang PY, Maleki A, Stephenson A, Montieth A, Filipowicz A, Syeda S, Asgari S, Walsh M, Metzinger JL, Foster CS. Treatment of Noninfectious Retinal Vasculitis Using Subcutaneous Repository Corticotropin Injection. *J Ophthalmic Vis Res*. 2021 Apr 29;16(2):219-233.