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Retinal Vasculitis: It's Significance

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Unlike Uveitis in the absence of retinal vasculitis, a situation in which approximately 30% of cases may turn out to be idiopathic, uveitis associated with retinal vasculitis is almost never idiopathic. Further, the disease causing the ocular inflammation is nearly invariably a systemic one. The systemic disease is often occult, defying even the best internist's efforts at uncovering the "cause" of the patient's uveitis and retinal vasculitis. But through time, extraocular manifestations of the patient's systemic disease eventually emerge making quite clear the nature of the underlying systemic disease. Polyarteritis nodosa, granulomatosis with polyangiitis, systemic lupus erythematosus, sarcoidosis, multiple sclerosis, Behcet's disease, and syphilis are but a few of the diseases that can behave in this way.

Furthermore, the significance of the de novo appearance of retinal vasculitis in the patient with a well-established systemic disease is also profound. For example, a patient with well characterized Behcet's disease with oral and genital ulcers, arthritis, and erythema nodosum may be well controlled and highly functional on twice daily Colchicine, an oral non-steroidal anti-inflammatory agent, and low dose prednisone. The appearance of retinal vasculitis, carries with it profound implications for such a patient. The aforementioned therapeutic "recipe" will no longer be sufficient for this patient. The appearance of the retinal vasculitis is a signal the underlying character of the patient's Behcet's disease has now changed, and unless the vigor of treatment is increased, not only is the patient likely to be bilaterally blind within four years, but the patient has approximately a 30% chance of developing vasculitis of the brain as well. Similar phenomena exist with respect to patients with system lupus erythematosus, rheumatoid arthritis, relapsing polychondritis, granulomatosis with polyangiitis, and polyarteritis nodosa.

The significance of the presence of retinal vasculitis, therefore is enormous, both from the standpoint of the likelihood of eventually discovering an underlying systemic disease causing the retinal vasculitis and also from the standpoint of the retinal vasculitis being a "barometer" of sub-clinical vasculitic lesions that may be lethal if the patient is not more vigorously treated.