

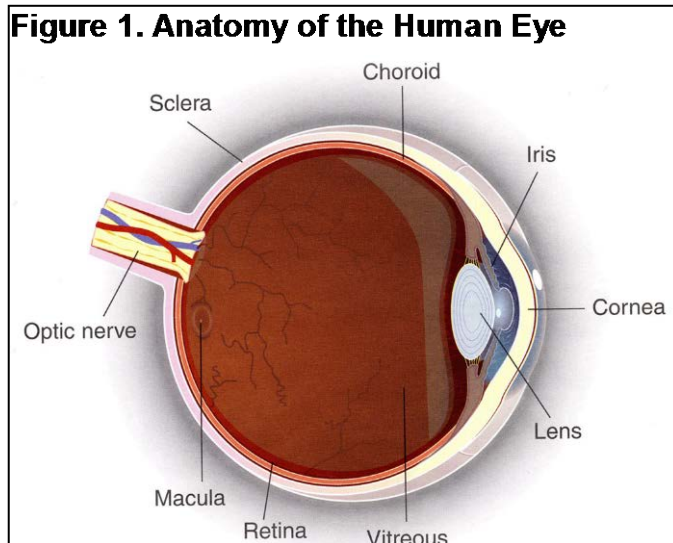
What You Should Know About Ocular Sarcoidosis

By David J. Browning, MD, PhD

Sarcoidosis is an inflammatory disease of unknown cause that involves many organ systems, but most commonly the lungs, the eyes, and the skin. Patients may range in age from childhood to the elderly, but the peak incidence occurs in the age range of 20 – 40 years. Women seem to be twice as frequently affected as men. African Americans have a disproportionate share of sarcoidosis, but no racial group is spared. Since the eyes are involved in 25 – 50% of all cases of sarcoidosis, the ophthalmic signs and treatments are important aspects of the care of patients with sarcoidosis.

What Happens to the Eyes in Sarcoidosis?

Any part of the eye can be involved in sarcoidosis. In 85% of cases, inflammation involves the front of the eye with inflammatory cells floating in the aqueous humor (iritis), inflammatory nodules developing in the iris and ciliary body (cyclitis), and scarring of iris to the lens. Figure 1 shows the anatomy of the eye to orient the reader.



In 25% of cases the tissues in the back of the eye are inflamed. Inflammatory clumps of white cells may float in the vitreous gel, inflammatory sheathing may occur around the retinal veins (perivenous sheathing), and inflammatory nodules may develop in the layer of vessels underlying the retina (called the choroid) or in the optic nerve, which carries the neural signal from the retina to the brain. Less commonly, inflammation involves

eyelid skin, the lacrimal gland (which makes tears), or the orbital soft tissues. Figure 2 shows a choroidal inflammatory nodule. Figure 3 shows perivenous inflammatory sheathing.

Approximately 20% of cases of sarcoidosis begin with an eye problem, thus in a case suggestive of sarcoidosis, investigations are often conducted to look for other organ involvement. The lungs and skin are carefully checked for inflammatory nodules and infiltrates.

Figure 2. Choroidal Inflammatory Nodule

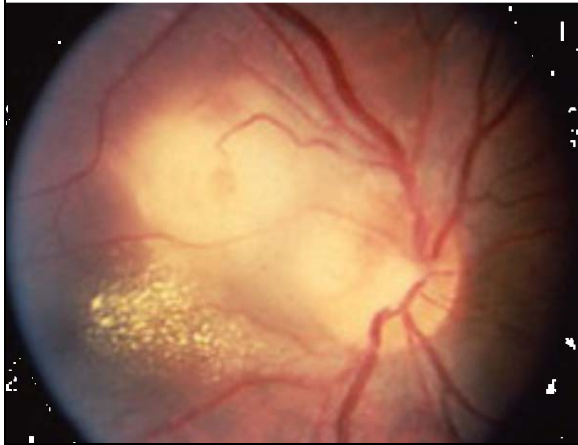


Figure 3. Perivenous Inflammatory Sheathing



Clinical Testing

Although no test is absolutely diagnostic for sarcoidosis, many tests contribute to an overall profile tending to confirm or refute a diagnosis of sarcoidosis. Blood tests include the angiotensin converting enzyme and serum lysozyme levels. Imaging studies include fluorescein angiography, ICG angiography, chest x-ray, CT scans, and gallium scans. The first two tests are done in the ophthalmologist's office and involve injection of dye into a vein in the patient's arm with a series of photographs taken of the back of the eye. The fluorescein angiogram helps most when the retina is involved by sarcoidosis. It can show retinal vascular leakage or occlusion. The ICG angiogram shows problems with a layer beneath the retina called the choroid. It can show inflammatory collections of cells called granulomas and choroidal nonperfusion.¹ If the cranial nerves are affected, a brain MRI scan can be useful. If these tests point toward an accessible inflammatory lesion, it may be biopsied, as this is the most reliable means to make a correct diagnosis. The conjunctiva (skin of the eyeball), any eyelid or facial skin lesions, or lesions of the lung are the most common sites for a biopsy.

Treatment of Sarcoidosis

Treatment of iritis usually begins with corticosteroid and pupillary dilating drops. The steroids reduce the inflammation and the pupillary dilation helps prevent iris scarring to the lens. If the posterior parts of the eye are inflamed, corticosteroids may be used by mouth or as injections in or around the eye. If steroids produce too many side effects or are unsuccessful in controlling the inflammation, then drugs called immunomodulators may be used, such as cyclosporine, azathiopine, or methotrexate. Periodic laboratory tests are needed if these drugs are used to ensure that toxicity to the bone marrow, kidneys, or liver does not occur. An internist or internal medicine subspecialist such as a rheumatologist is necessary to manage the systemic aspects of sarcoidosis.

Ophthalmic Monitoring of Treatment Side Effects

Even when sarcoidosis does not involve the eye, an ophthalmologist may be involved in care. Steroids can produce cataracts or glaucoma as side effects, and an ophthalmologist needs to monitor these problems. Sometimes drugs such as hydroxychloroquine or chloroquine are used to manage arthritis or skin lesions in sarcoidosis. Since these can cause retinal toxicity, an ophthalmologist needs to check the eyes at intervals.

Final Comments

Sarcoidosis is a chronic disease that can wax and wane over many years. The eyes are often involved, making ophthalmic attention an important part of a patient's overall care.

Once you have read this brochure, if you would like to find more in-depth information about sarcoidosis, an excellent resource is the National Library of Medicine website section called PubMed. It can be accessed via any search engine, or directly at this link, <http://www.ncbi.nlm.nih.gov/entrez/query.fcgi>. It includes an extensive database of reliable articles published in peer-reviewed medical journals from all over the world.

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References

1. Herbort CP, Neri P, El Asrar AA, Gupta V, Kestelyn P, Khairallah M, MantovaniA, Tugal-Tutkun I, Papadia M. Is ICGA still relevant in inflammatory eye disorders? Why this question has to be dealt with separately from other eye conditions. *Retina* 2012;32:1701-1703.