

BASIC MANAGEMENT OF ANTERIOR SCLERITIS

A discussion of the causes of and treatments for anterior scleritis.

BY NISHA ACHARYA, MD

Ophthalmologists who treat uveitis have a growing number of treatment options from which to choose. However, they must recognize the importance of selecting a treatment regimen that is most efficient while minimizing burden on the patient. In an effort to help make the treatment decision process less daunting, Thomas Albin, MD, moderator of the Uveitis Resource Center, interviewed clinicians to gain insights into their approaches to managing patients with uveitis. Videos of the interviews can be found on the Uveitis Resource Center (bit.ly/RT_URC).

In this issue, Nisha Acharya, MD, director of the uveitis service and the uveitis fellowship program at the F.I. Proctor Foundation at the University of California, San Francisco, recaps her talk with Dr. Albin about managing patients with anterior scleritis. She describes the typical presentation of the condition, what her patient workup consists of, and how she manages patients over time.



There are two types of scleritis, anterior and posterior. Anterior scleritis is the more common of the two, and, as such, it is a condition that many ophthalmologists encounter in practice. The classic sign is an extremely red eye. Patients will call the office and describe their eye as being really red, almost purple in color, and swollen. The classic symptom is severe, deep, aching pain—painful enough to disrupt sleep. The pain may worsen with eye movement but is for the most part constant. In this article, I explain how to diagnose and manage most cases of anterior scleritis.

SCLERITIS VS. EPISCLERITIS

Scleritis and episcleritis are sometimes confused; because both are parts of the spectrum of inflammation, severity, signs, and symptoms may overlap. Just like scleritis, episcleritis presents with a red eye and can be sectoral or diffuse. But symptoms are minimal with episcleritis. Patients either have no pain or minor pain that is sometimes described as a foreign body sensation. Additionally, the conjunctiva or the superficial episcleral vessels are involved with episcleritis, unlike in scleritis, where the deep episcleral vessels are inflamed. I find the clinical examination in natural light and with a slit lamp to be the best method of confirming a diagnosis. Phenylephrine 2.5% typically blanches the vessels in episcleritis, but generally does not blanch the vessels in scleritis.

When Scleritis is Necrotizing Scleritis

Necrotizing scleritis is the most severe form of anterior scleritis, and it requires aggressive treatment. Thus, early diagnosis of this form of the condition is of utmost importance. Typically what is seen on examination is a thin area of sclera caused by frank loss of tissue. The area may appear darker compared with the surrounding tissue because of the pigmented uveal tissue underneath it. If there is significant thinning, the area may look gray or even black because you are looking directly through to the uvea. These areas can also be avascular, in which case they may appear white in color because of poor blood flow, which is a risk factor for developing necrosis. These features—a dark or a white appearance—are both telltale signs of a necrotizing scleritis.

NEXT STEPS

Although a complete workup is not generally necessary for a diagnosis of episcleritis, we do a complete evaluation if we suspect that a patient has scleritis. I begin my examination with a head-to-toe review of systems, ask the patient detailed questions to understand what he or she is truly experiencing, and order laboratory tests.

About half of patients with scleritis will have either a known systemic autoimmune disease or some other associated condition—or you may be the first one to reveal that diagnosis to them. Roughly 40% of patients who have scleritis will have or will eventually develop rheumatoid arthritis (RA). Therefore, I

WATCH THE VIDEO

Dr. Acharya responds to questions posed by Thomas Albini, MD. The video can be viewed at bit.ly/acharyaURC.



always order a chest X-ray, urinalysis, and three laboratory blood tests: rheumatoid factor (RF); anti-cyclic citrullinated peptide antibody (anti-CCP antibody); and antineutrophil cytoplasmic antibody (ANCA).

A chest X-ray is helpful because granulomatosis with polyangiitis, formerly known as Wegener disease, can sometimes appear in the lungs, and elevated protein counts and red blood cell casts associated with this disease can be found with urinalysis. After RA, granulomatosis with polyangiitis is the most common condition associated with scleritis; other associated autoimmune disorders include inflammatory bowel disease, relapsing polychondritis, Churg-Strauss syndrome, and Cogan syndrome.

Nearly all patients with RA will have a positive RF, but it is important to note that the test yields a high false-positive rate. Alternatively, the anti-CCP antibody test has good specificity and is therefore less likely to produce a false-positive result. It is recommended to order both tests when evaluating for RA. ANCAs are antibodies present in many vasculitic conditions. When the ANCA test is ordered, most laboratories test for both perinuclear ANCA and cytoplasmic ANCA. In my opinion, checking ANCA is imperative. Some physicians also add human leukocyte antigen B27 and antinuclear antibody to the blood test panel, but I do not think either is indicated because ANCA is very nonspecific and B27-associated systemic diseases are not associated with scleritis. My practice is to also evaluate for syphilis and tuberculosis in all patients with ocular inflammation, including scleritis.

TREATMENT OPTIONS

Before initiating treatment of noninfectious anterior scleritis, I first assess the severity of the patient's condition.

Mild to Moderate Cases

For patients with mild to moderate noninfectious anterior scleritis and minimal pain, I prescribe an oral nonsteroidal antiinflammatory drug (NSAID). My drug of choice is 75 mg sustained-release indomethacin twice daily. Other options include naproxen 500 mg twice daily, ibuprofen 600 mg to 800 mg three to four times daily, and flurbiprofen 200 mg to 300 mg three times daily. Most patients with scleritis respond

well to oral NSAIDs, but, if after 2 weeks there is no response, then treatment should be changed.

When NSAIDs Are Not Effective

Systemic steroids are the next step in the treatment algorithm. Dosing typically begins with 1 mg/kg/day up to 60 mg. I maintain that dose until the patient returns for follow-up about 14 days later, at which time I begin tapering the dose by about 10 mg every 2 weeks. Depending on the level of inflammation, I may do a slower or faster taper and adjust as needed depending on the treatment response. It is necessary to observe patients closely, and the goal is to eventually wean them completely off of steroids. If tapering results in worsening of the scleritis, I then consider options for long-term management because we do not want to continue high doses of steroids.

Long-Term Management

Options for long-term management can include immunomodulatory therapies and biologics. I start with methotrexate because it works well for most patients with scleritis. However, some cases of scleritis, including necrotizing scleritis, may require use of a biologic. Anti-tumor necrosis factor drugs such as infliximab (Remicade, Janssen Biotech) and adalimumab (Humira, AbbVie) and the anti-CD20 antibody rituximab (Rituxan, Genentech) work well. For patients with severe disease who cannot be taken off steroids, I consider jumping directly to one of these biologics.

When Systemic Therapy Is Not an Option

For patients who choose not to be on systemic therapy or who cannot tolerate it, I prefer to use 0.1-mL subconjunctival injections of the 40-mg/mL dosage of triamcinolone. I do not inject triamcinolone in patients who have frank loss of tissue and necrotizing disease because of the potential to exacerbate the necrosis. However, for patients with nonnecrotizing disease, subconjunctival corticosteroid injection is an alternative to systemic therapy that can be effective for as long as 29 months.¹

CONCLUSION

Many patients with anterior uveitis are referred to us by optometrists and general ophthalmologists. Whatever their path to our doors, we owe these patients our best efforts to treat their conditions. We have a solid arsenal of therapeutic options from which to choose, but, in order to achieve successful resolution, compliance with the therapeutic regimen is essential. Inevitably, some cases will prove difficult. Such cases are best dealt with by uveitis specialists. ■

1. Albini T, Zamir E, Read RW. Evaluation of subconjunctival triamcinolone for nonnecrotizing anterior scleritis. Paper presented at: American Academy of Ophthalmology Annual Meeting; October 23-26, 2004; New Orleans, LA.

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