Scleritis and episcleritis

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Definition

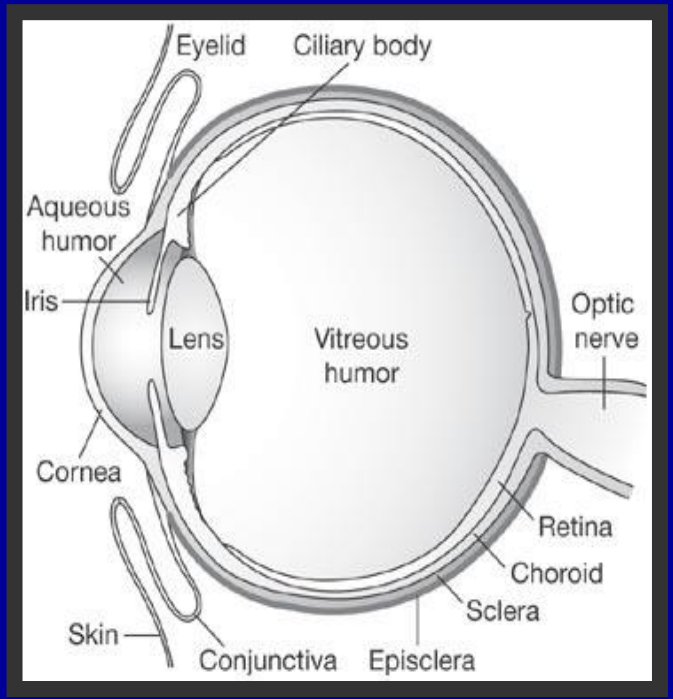
Scleritis is typically a severe painful inflammatory process centered in the sclera.

It may involve the cornea, adjacent episclera, and underlying uvea.

Relevant anatomy

The sclera is an incomplete shell comprising approximately 90% of the outer coat of the eye.

It begins at the limbus and terminates at the optic canal.



Relevant anatomy *(continued)*

The sclera has a rich sensory innervation from the ciliary nerves explaining a severe pain of scleritis.

Many short posterior ciliary nerves supply the posterior portions and two long posterior ciliary nerves supply the anterior region.

Relevant anatomy *(continued)*

Because the extraocular muscles are inserted into the sclera, the dull ache of scleral inflammation is made worse by ocular movement.

Relevant anatomy *(continued)*

Sclera is fused with the dura mater and arachnoid sheaths of the optic nerve explaining the optic nerve swelling seen in 43% of eyes with posterior scleritis.

Etiologies

- -idiopathic
- -immune diseases
- -vasculitis
- -Infections
- -masquerade syndromes (malignancy)

Pathophysiology

The incidence of systemic disease in patients with scleritis is reported as 39–50%.

There is no known HLA association.

It is useful to note that the association of posterior scleritis with anterior scleritis is much more likely to occur in a patient in whom there is an underlying systemic disease.

Pathophysiology *(continued)*

A large number of connective-tissue disorders are associated with scleral disease but the most common is rheumatoid arthritis.

Wegener granulomatosis is the most common vasculitis associated with scleritis.

Pathophysiology *(continued)*

Table 1 Frequency of systemic disease among patients with scleritis.[1–3,a]

Disease	Percentage of patients
Rheumatoid arthritis	10.3–18.6
Wegener's granulomatosis	3.8–8.1
Relapsing polychondritis	1.6–6.4
Systemic lupus erythematosus	1.0–4.1
Inflammatory bowel disease	2.1–4.1
Seronegative spondyloarthropathies—ankylosing spondylitis, reactive arthritis, psoriatic arthritis	0.3–3.5
Polyarteritis nodosa	0.4–1.1

Pathophysiology *(continued)*

Although most scleritis is immune-mediated, it can also be triggered by infection, ocular surgery, malignancy, or drugs.

Pathophysiology *(continued)*

Infectious scleritis can be viral, bacterial, fungal, and parasitic.

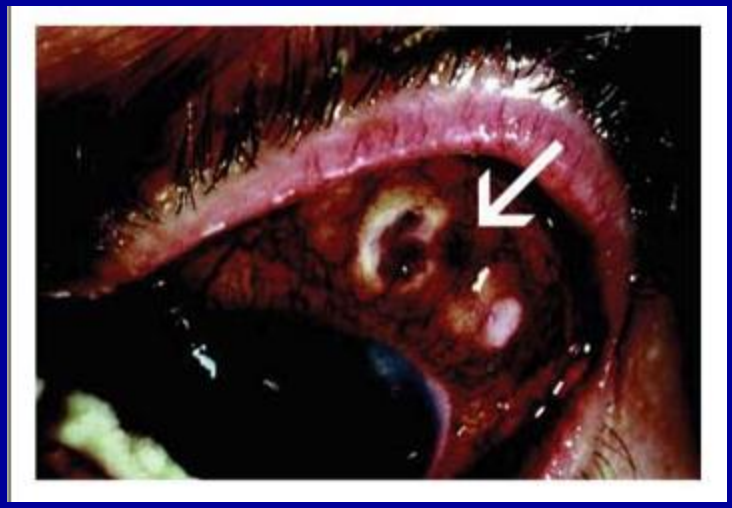
It is uncommon particularly in the absence of infectious keratitis.

The most common infection was herpes zoster ophthalmicus.

Pathophysiology *(continued)*

A common risk factor for infectious scleritis is a history of pterygium surgery with adjunctive mitomycin C administration or beta irradiation.

Once the sclera is invaded, the infection is extremely difficult to eradicate. Pseudomonas aeruginosa is the most common pathogen reported.



Surgically induced necrotizing scleritis (SINS)

It can occur after a variety of procedures, most commonly after cataract surgery particularly when a limbal incision is used with an extracapsular approach.

Other surgeries associated with SINS are pterygium removal and scleral buckle.

Of the patients who develop SINS, 75% of patients have undergone two or more surgical procedures prior to onset of the disease.

Surgically induced necrotizing scleritis (SINS)

Mean time to presentation from surgery has been reported as 9 months ranging from 2 weeks to 6 months.

The mean age of patients with SINS is significantly younger than the mean of all cataract patients.

Surgically induced necrotizing scleritis (SINS)

Patients who have SINS need careful systemic investigation as up to 90% of patients in one study were later diagnosed with autoimmune vasculitic disease which required immunosuppression therapy.

Clinical presentations

Patients with scleritis may present in one of two ways—they may already be known (up to 50%) to have an underlying related disorder, such as rheumatoid arthritis, or the scleritis may present de novo in the absence of any known underlying systemic disease.

Clinical presentations *(continued)*

Patients with scleritis are predominantly middle aged and Caucasian with a mean age at onset of 49 years.

Female patients comprise 71% of all patients with scleritis and 65% of patients with posterior scleritis.

Clinical presentations *(continued)*

The characteristic feature of scleritis is the severe pain that may involve the eye and orbit and radiates to involve the ear, scalp, face, and jaw.

It has a subacute onset and the intensity of the pain may increase over several weeks.

The pain is usually severe in nature and often resistant to mild analgesics.

The pain often awakens the patient from sleep in the early hours of the morning.

Clinical presentations *(continued)*

The patient may have photophobia and lacrimation and the globe is usually tender to touch, which may be extremely so.

Unilateral or bilateral inflammation can occur.

Classification

Classification of Scleral Inflammation

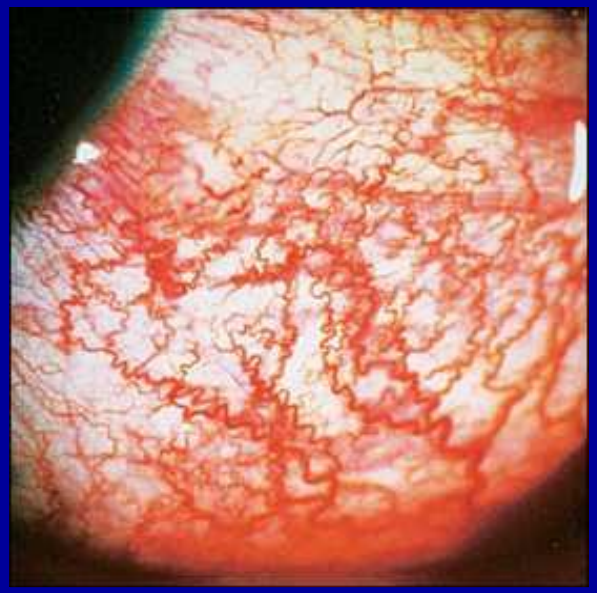
Type	Subtypes
Episcleritis	Diffuse
	Nodular
Anterior Scleritis	Diffuse
	Nodular
	Necrotizing
	With inflammation
	Without inflammation
	Scleromalacia perforans
Posterior Scleritis	Diffuse
	Nodular
	Necrotizing (at least on histopathology)

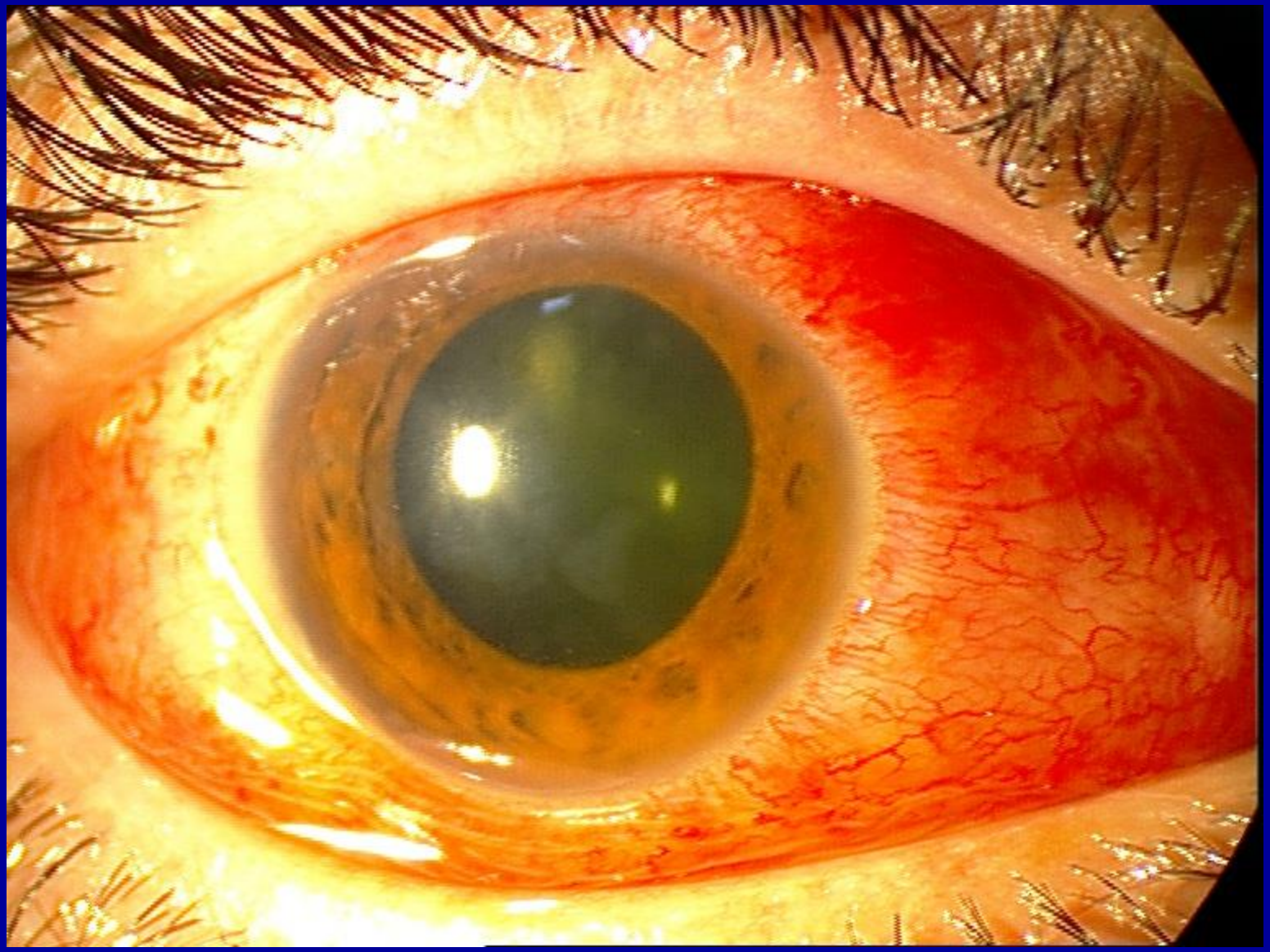
Classification *(continued)*

Scleritis may involve the anterior sclera, posterior sclera or both.

Anterior scleritis is the most common pattern of disease, and it may be diffuse, nodular, or necrotizing in type.

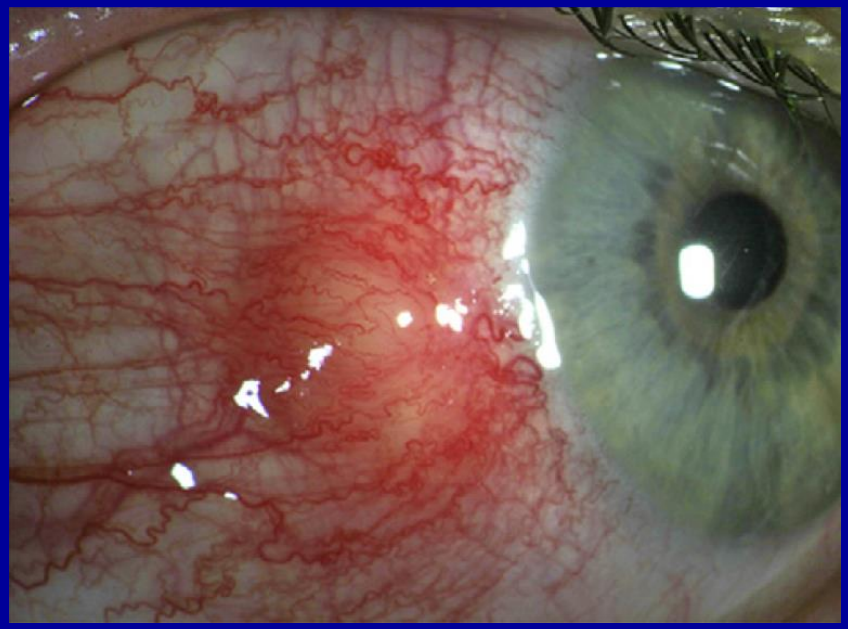
In diffuse form, it can be localized to a patch of the sclera or may involve the entire anterior sclera.





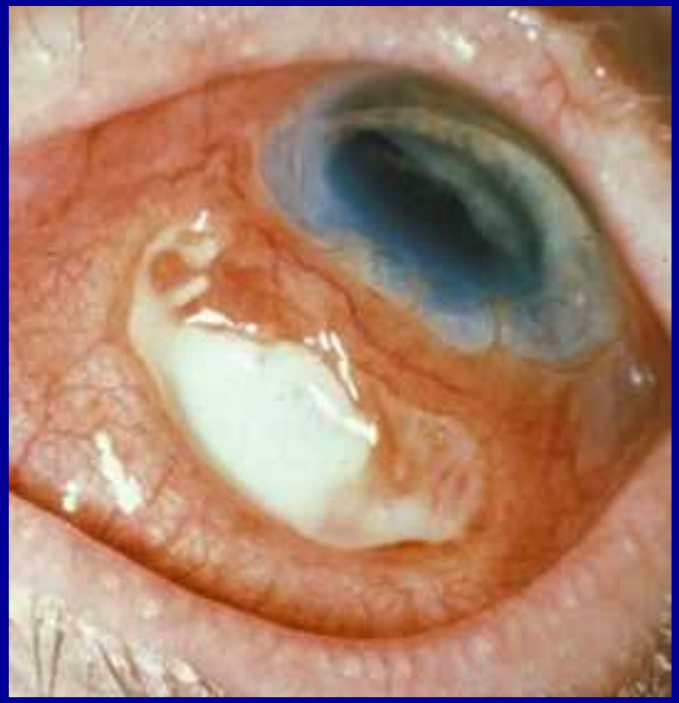
Classification *(continued)*

Nodular anterior scleritis is characterized by a more localized area of scleral edema such that distinct nodules result. They may be single or multiple and can become quite prominent and tender to palpation



Classification *(continued)*

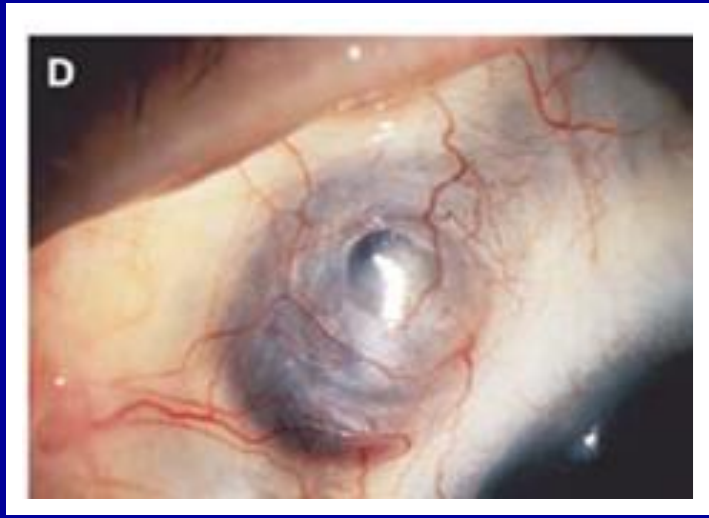
Necrotizing anterior scleritis is the most severe form of scleritis and is a serious threat to vision and the integrity of the eye.



Classification *(continued)*

Very rarely a necrotizing form of anterior scleritis occurs in the absence of pain and other clinical signs of inflammation in patients with longstanding rheumatoid arthritis and is termed **scleromalacia perforans**.

It is the result of an obliterative arteritis involving the deep episcleral vascular plexus.



Classification *(continued)*

scleromalacia perforans does not produce the acute clinical signs of necrotizing scleritis described above but is asymptomatic or presents with blurred vision from high astigmatism due to scleral thinning leading to loss of scleral rigidity. There is no corneal involvement except for limited peripheral corneal thinning.

Classification *(continued)*

Posterior scleritis is defined as involvement of the sclera posterior to the insertion of the rectus muscles and may be difficult to recognize and can be misdiagnosed with ocular tumor in the absence of good imaging as there may be little in the way of physical signs.

Clinical presentations *(continued)*

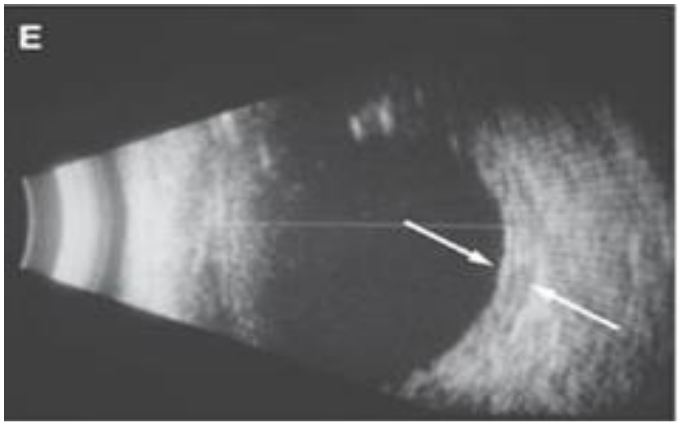
40% of patients with posterior scleritis have no evidence of anterior scleral inflammation, and 9% of patients have no abnormal clinical signs on examination

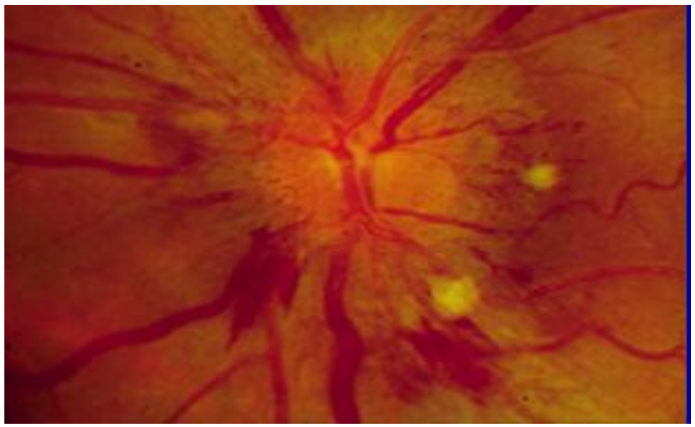
When only the posterior sclera is involved, clinical signs may be less obvious and ultrasonography or other imaging studies may be necessary to confirm the diagnosis.

Other findings can be proptosis, exudative retinal detachment, and disc swelling.

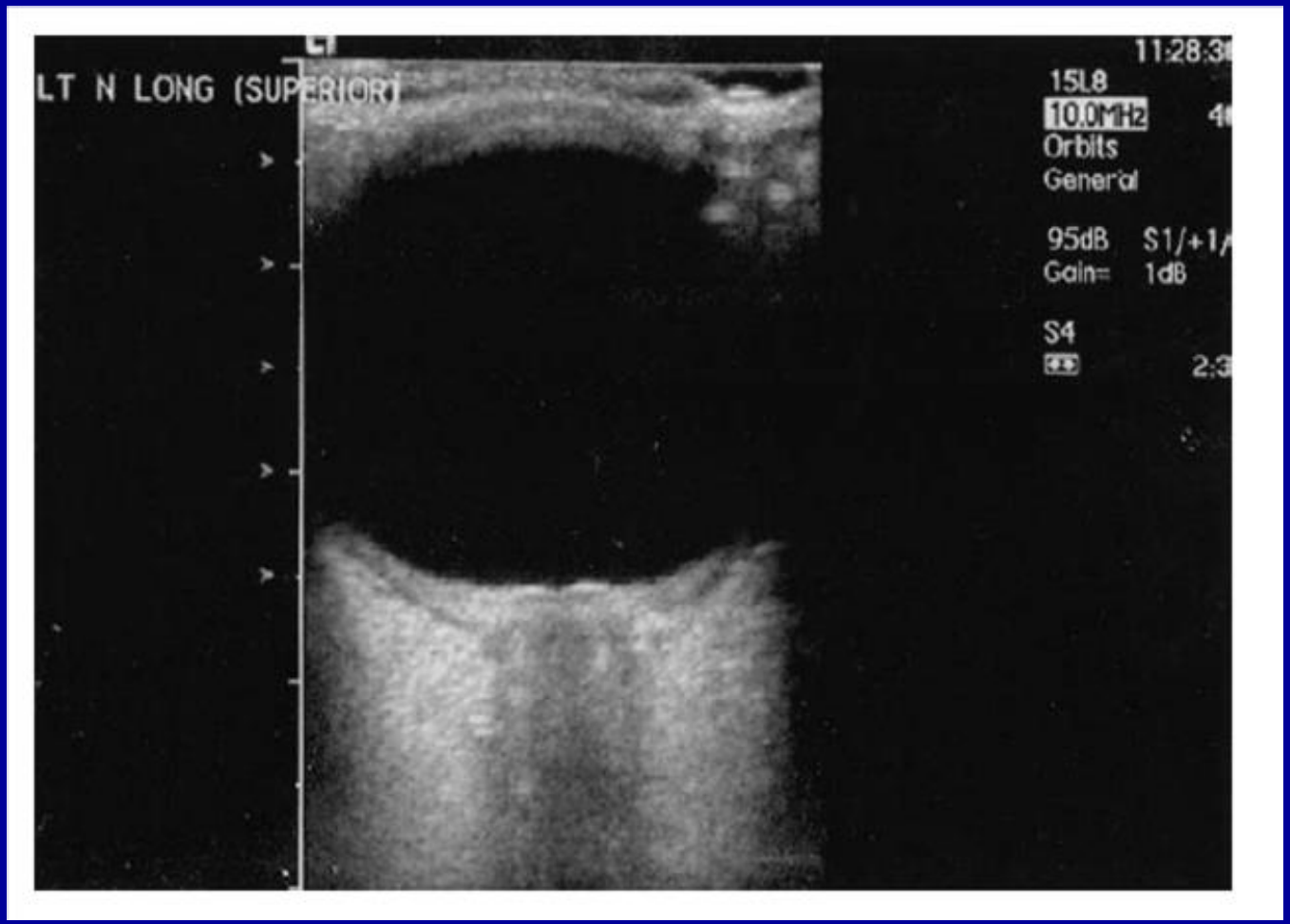
Classification *(continued)*

Modern B-scan ultrasonography
and high-definition orbital
magnetic resonance imaging
are used to detect posterior scleritis





Classification *(continued)*



Classification *(continued)*

In posterior scleritis, large serous retinal detachments can be associated with shallowing of the anterior chamber, and secondary angle-closure glaucoma from ciliary body rotation secondary to uveal effusion.

Classification *(continued)*

Necrotizing posterior scleritis has been reported on histopathological examination of enucleated eyes but cannot be recognized clinically at this time.

Complications

There is commonly spread of inflammation involving the adjacent structures including cornea, anterior chamber, ciliary body, and trabecular meshwork resulting in keratitis, anterior uveitis, and elevated intraocular pressure, which may lead to staphyloma formation.

Complications *(continued)*

Signs of corneal infiltrates, thinning, or stromal keratitis may be present with corneal ulceration.

Complications *(continued)*

All types of scleritis can be associated with uveitis which may be mild or severe.

Anterior uveitis occurs in up to 40% of eyes with scleritis.

It is more common with more severe scleritis, most often seen in association with necrotizing disease.

It is important to be sure that it does not signify an associated endophthalmitis when seen with scleral necrosis.

Complications *(continued)*

Glaucoma

- trabeculitis
- raised episcleral venous pressure
- Pupillary block due to PS formation
- Closed angle due to choroidal detachment
- Steroid respoder

Complications *(continued)*

Necrosis of the sclera can be subtle or profound, localized, or generalized, and progress rapidly to expose the choroid resulting in globe rupture.

Poor prognosis

Patients with posterior scleritis greater than the age of 50 years have been shown to have a greater risk of having an associated systemic disease, associated visual loss, and were more likely to require systemic immunosuppressive agents.

Episcleritis vs. Scleritis

Scleritis is often confused with episcleritis, which is inflammation confined to the superficial episcleral tissue and does not involve the deep episcleral tissue that overlies the sclera.

Episcleritis vs. Scleritis *(continued)*

Episcleritis is a mild nonvision-threatening form of ocular inflammation that is usually idiopathic in nature and is not usually associated with involvement of other ocular structures.

Episcleritis vs. Scleritis *(continued)*

In episcleritis the patient's main complaint is often redness, which may also be associated with a feeling of grittiness.

This is in contrast to scleral inflammation, where pain is much more prominent along with globe tenderness and redness that may involve the whole eye or just a small localized area.

Episcleritis vs. **Scleritis** ***(continued)***

Slit-lamp examination with red-free light and diffuse illumination accentuates visibility of blood vessels and areas of capillary nonperfusion.

Episcleritis vs. **Scleritis** ***(continued)***

In episcleritis, the conjunctival and superficial episcleral vascular plexuses are displaced outward and the underlying deep episcleral plexus is uninvolved.

In scleritis all vascular layers may be involved but the maximal involvement is in the deep episcleral plexus.

Episcleritis vs. Scleritis *(continued)*

Examination in natural daylight can be extremely useful that are often not appreciable using the slit-lamp.

The scleritis has a characteristic bluish-violet hue, while episcleritis has a distinct red hue.

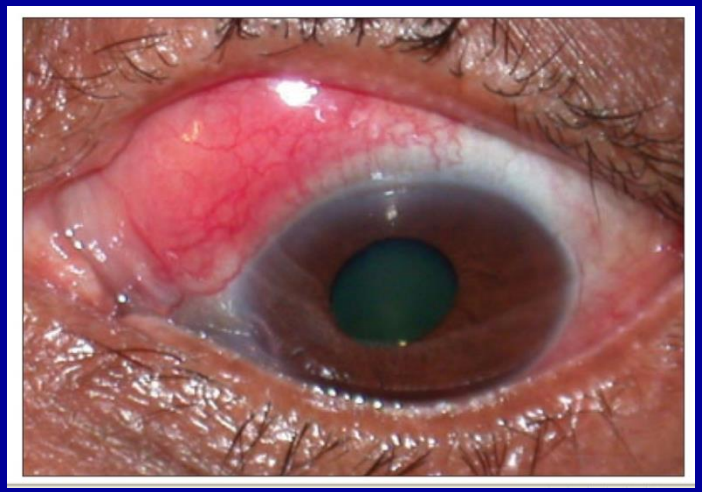
Episcleritis vs. **Scleritis** ***(continued)***

The conjunctival and superficial vessels can be blanched with 2.5–10% phenylephrine or 1:1,000 epinephrine while the deep vessels are hardly affected.

Ocular tumors vs. Scleritis

Intraocular tumors such as melanomas may mimic posterior scleritis but can also be associated with it.

Rarely conjunctival tumors and lymphoma can mimic scleritis and secondary malignant deposits can cause severe scleral inflammation.



Ocular tumors vs. Scleritis *(continued)*

Dorey et al described two patients in whom the initial presentation of lymphoma was misdiagnosed as scleritis.

The symptoms and signs did not respond to non-steroidal anti-inflammatory drugs or to steroid treatment.

Biopsy should be considered when the pain is atypical for scleritis and the mass is salmon pink, elevated, and solid.

Ocular tumors vs. Scleritis *(continued)*

Posterior scleritis may present as a mass lesion and some of these eyes have been enucleated erroneously.

The presence of high internal reflectivity and a retrobulbar echo-lucent area which represents oedema in the tenons capsule on B scan is in favor of posterior scleritis.

Angiography

The superficial episcleral capillary plexus is a radially arranged series of vessels, which anastomose at the limbus with the conjunctival vessels and with the deep plexus.

The deep episcleral capillary network is closely applied to the sclera.

Angiography *(continued)*

The longer transit time because of its protein binding properties, as well as leakage and staining patterns, make indocyanine green a potentially useful technique when investigating scleral inflammation.

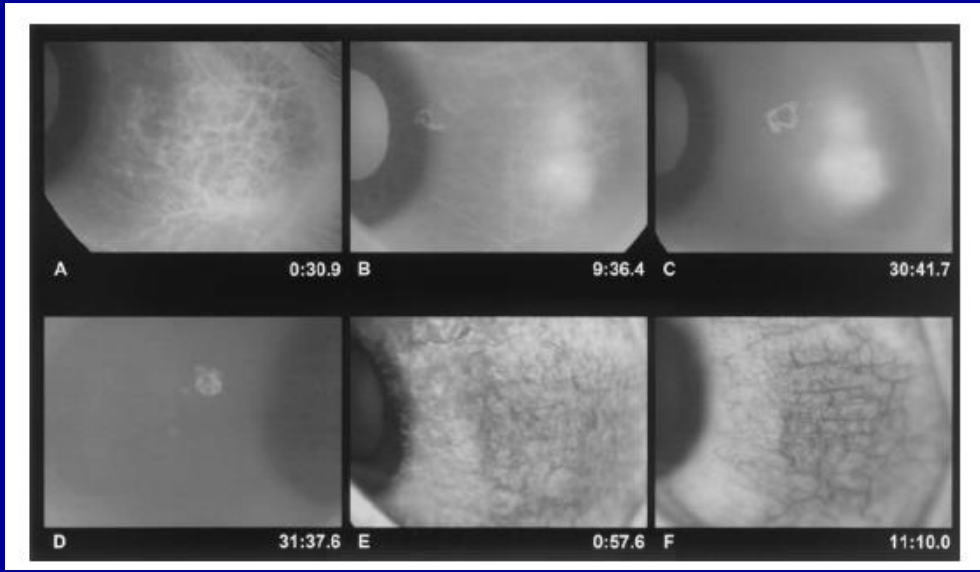
Leakage and staining with indocyanine green was also apparent in our patients with profound scleral inflammation.

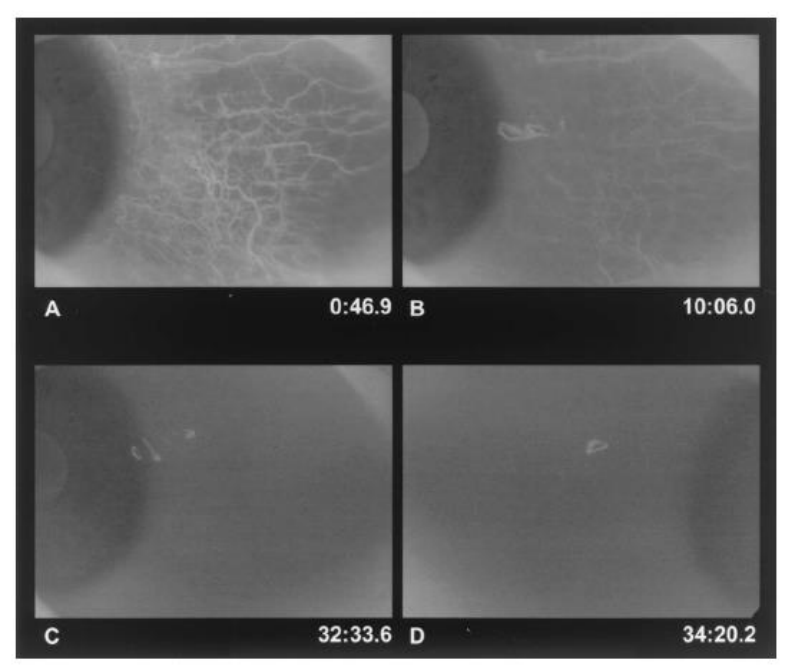
Angiography *(continued)*

Anterior segment indocyanine green angiography and fluorescein angiography were done in 14 patients with nodular scleritis and 10 patients with diffuse scleritis.

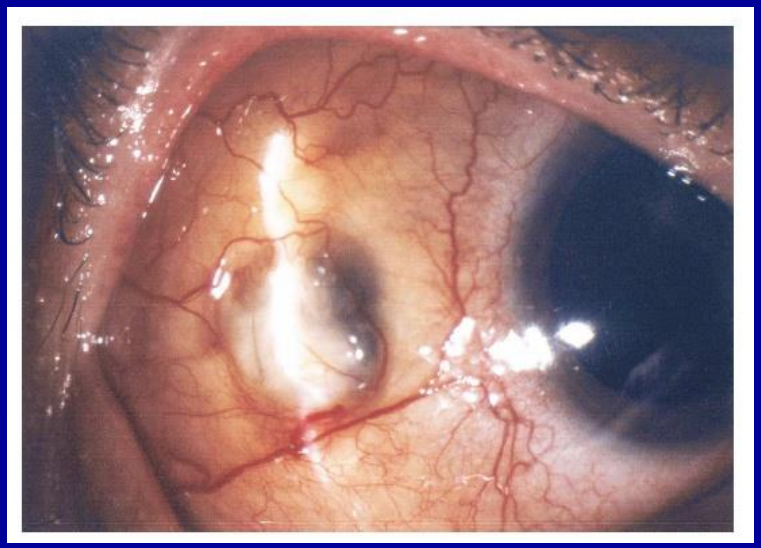
Although leakage of fluorescein and indocyanine green was observed in all patients, staining of the inflammatory area was apparent in only 3 patients with fluorescein and in all patients with indocyanine green angiography.

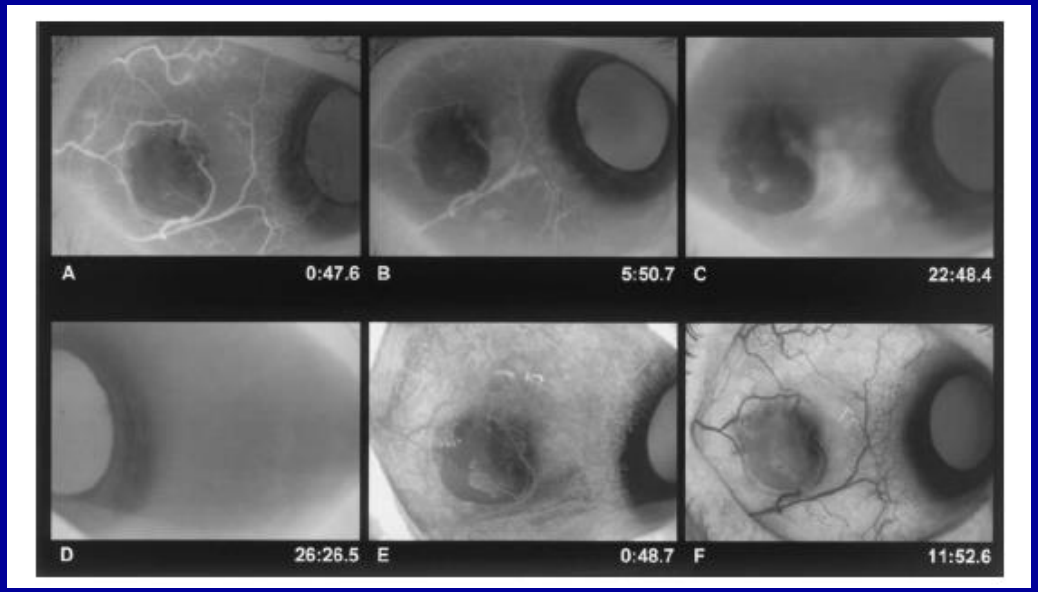
Angiography *(continued)*





Angiography *(continued)*





Pathologic findings

Most eyes with scleritis do not come to biopsy or to enucleation.

In one study of enucleated eyes, eyes with necrotizing scleritis showed vasculitis with fibrinoid necrosis and neutrophil invasion of the vessel wall in 75%, and vascular immunodeposits were found in 93% in the scleral tissue.

Pathologic findings *(continued)*

HLA-DR expression is dramatically increased. Other findings are increase in T cells of all types, macrophages, and neutrophils.

Granulomatous inflammation is observed in enucleated eyes with necrotizing disease suggesting an active cell-mediated immune response.

Antibody deposition was not seen nor was complement found, suggesting that T cells are the effector cell in scleritis rather than immune complex deposition.

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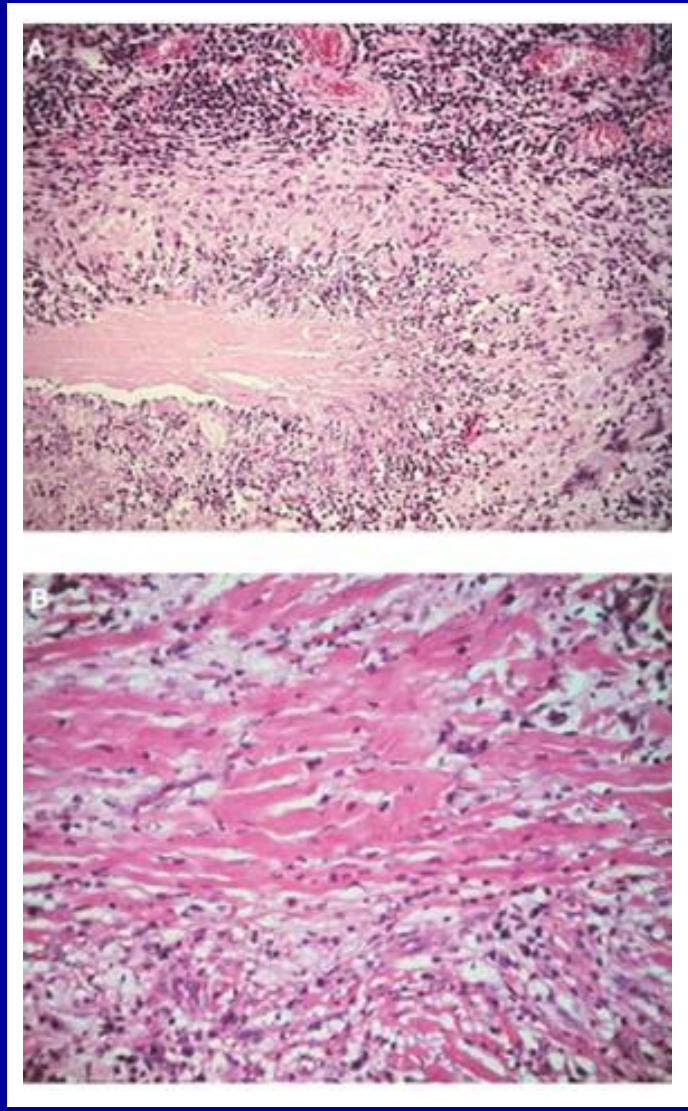
Pathologic findings *(continued)*

Zonal necrotizing granulomatous inflammation in a patient with rheumatoid arthritis

Diffuse nongranulomatous inflammation in a patient with ‘idiopathic scleritis’)



Treatment

The mainstay of therapy for non-infectious scleritis includes oral NSAIDs and oral prednisone.

Topical steroids remain useful when there is co-existing intraocular inflammation or mild disease.

Subconjunctival injection of triamcinolone may also be an effective treatment for non-infectious, non-necrotizing anterior scleritis.

Treatment

(continued)

Steroid-sparing therapy if the patient cannot be successfully tapered below 10 mg of prednisone without symptoms or clinical signs of active scleritis.

-Methotrexate
-Calcineurin inhibitors

Thanks for Your Attention