# Sclera

## Diseases related to the sclera; all are discussed in this lecture

1. Color Changes
2. Staphyloma and Ectasia
3. Trauma
4. Inflammation (Episcleritis and Scleritis)

## Basic Knowledge

### Function:

- The sclera and the cornea form the rigid outer covering of the eye.
- All six ocular muscles insert into the sclera.

**Morphology:** The sclera is fibrous, whitish opaque, and consists of nearly acellular connective tissue with a higher water content than the cornea.

- The sclera is thickest (1mm) anteriorly at the limbus of the cornea where it joins the corneal stroma and at its posterior pole.
- It is thinnest (0.3mm) at the equator and beneath the insertions of the rectus muscles.
- The site where the fibers of the optic nerve enter the sclera is known as the lamina cribrosa.
- In the angle of the anterior chamber, the sclera forms the trabecular network and the canal of Schlemm.
- The aqueous humor drains from there into the intrascleral and episcleral venous plexus through about 20 canaliculi.

### Neurovascular supply:

- Vortex veins and the short anterior and posterior ciliary arteries penetrate the sclera.
- The ciliary nerves course through the sclera from posterior to anterior.

### Examination Methods

- The anterior portion of the sclera about as far back as the equator can be examined directly with a **slit lamp**.
- Evaluation of the sclera posterior to the equator requires indirect methods such as **ultrasound**.

- **Transillumination** can provide evidence of possible abnormal changes in the posterior sclera. However, this method is not as precise as an ultrasound study.

## Color Changes

The sclera is normally dull white like porcelain. Altered color suggests one of the following changes:

**A.** **Conjunctival** and/or **ciliary injection** and inflammation will give the sclera a red appearance
**B.** A sclera that is **very thin** will appear **blue** because of the underlying choroid (this occurs in the newborn, in osteogenesis imperfecta, and following inflammation; see Fig. 6.**4**).
**C.** In **jaundice,** the sclera turns **yellow**.
**D.** In **ochronosis** (alkaptonuria), the sclera will take on **brownish** color. This should be distinguished from pigmented changes in the conjunctiva.

## Staphyloma and Ectasia

- **Staphyloma** refers to a bulging of the sclera in which the underlying uveal tissue in the bulge is also thinned or degenerated.

By far the most common form is ***posterior staphyloma*** *in severe myopia,* a bulging of the entire posterior pole of the eyeball (Fig. 6.**1**). Staphyloma can also occur secondary to scleritis (see Fig. 6.**4**).

- **Ectasia** is a thinning and bulging of the sclera without uveal involvement, as can occur secondary to inflammation.

- Both staphyloma and ectasia are secondary or incidental findings. No treatment is available.

## Trauma

- The sclera is frequently involved in penetrating trauma.

- Deep injuries that extend far posteriorly usually also involve the choroid and retina.

## Inflammations

- Inflammations are the most clinically significant scleral changes encountered in ophthalmologic practice.

- They more often involve the anterior sclera (episcleritis and anterior scleritis) than the posterior sclera (posterior scleritis).

**Classification:** Forms of scleral inflammation are differentiated as follows:

- **Location:** anterior or posterior, i.e., anterior or posterior to the equator of the globe.
- **Depth:**

– Superficial (episcleritis).

– Deep (scleritis).

- **Nature:**

– Diffuse (usually scleritis).

– Circumscribed or segmental (episcleritis).

– Nodular, with formation of small mobile nodules (scleritis and episcleritis)

Posterior staphyloma in a highly myopic eye.

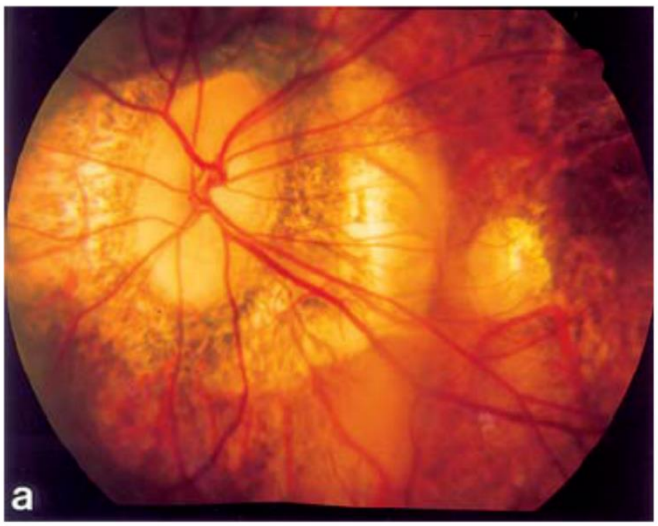
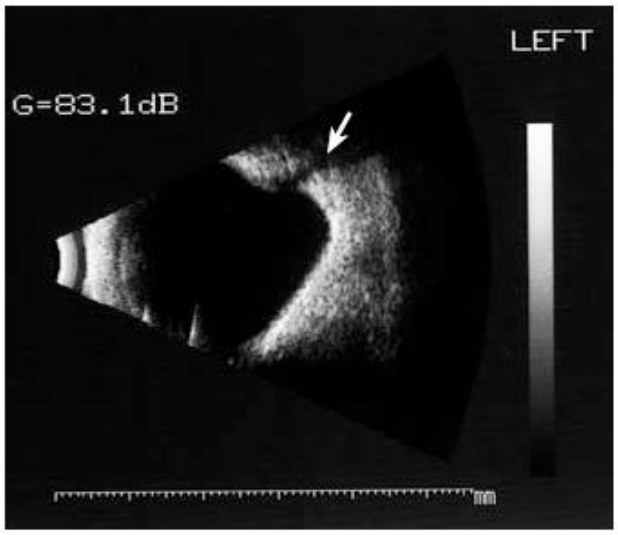



Fig. 6.1 **a** Ophthalmologic image of posterior staphyloma of the sclera.

**b** Ultrasound image showing




**b** Ultrasound image showing the posterior scleral bulge and oblique course of the optic nerve through the sclera.

## Episcleritis

**Definition**

- Circumscribed, usually segmental, and generally *nodular* inflammation of the episclera (connective tissue between sclera and conjunctiva).
- **Epidemiology:** Episcleritis is the most common form of scleral inflammation.
- **Etiology:** Episcleritis is rarely attributable to one of the systemic underlying disorders listed in Table 6.**1**, and is only occasionally due to bacterial or viral inflammation. Often episcleritis will have no readily discernible cause.
- **Symptoms:** Episcleritis can be unilateral or bilateral. It is usually associated with segmental reddening and slight tenderness to palpation.

**Findings:**

- The episcleral vessels lie within the fascial sheath of the eyeball (Tenon's capsule) and are arranged radially.
- In episcleritis, these vessels and the conjunctival vessels above them become hyperemic (Fig. 6.**2**).
- Tenon's capsule and the episclera are infiltrated with inflammatory cells, but the sclera itself is not swollen. The presence of small *mobile* nodules is typical of nodular episcleritis.

**Differential diagnosis:**

The disorder should be distinguished from **conjunctivitis** and **scleritis.**

- The conjunctival blood vessels are the most superficial; the episcleral vessels lie within Tenon's capsule and are arranged radially.
- When vasoconstrictive eyedrops are applied, the conjunctival injection will disappear but not the episcleral injection. This makes it possible to distinguish conjunctivitis from episcleritis.
- **Treatment and prognosis:** Episcleritis usually resolves spontaneously within one to two weeks, although the nodular form can persist for extended periods of time.

Severe symptoms are treated with topical steroids (eyedrops) or with a nonsteroidal anti-inflammatory agent.

## Scleritis

**Definition**

- Diffuse or localized inflammation of the sclera. Scleritis is classified according to location:

1. **Anterior** (inflammation anterior to the equator of the globe).
2. **Posterior** (inflammation posterior to the equator of the globe).

### Anterior scleritis is further classified according to its nature:

1. **Necrotizing anterior scleritis**
2. **Non-necrotizing anterior scleritis** (nodular or diffuse).with or without inflammation).

**Epidemiology:** Scleritis is far less frequent than episcleritis. Patients are generally older, and women are affected more often than men.

**Etiology:**

1. Approximately 50% of scleritis cases (which tend to have severe clinical courses) are attributable to systemic autoimmune or rheumatic disease (Table 6.**1**),
2. Or are the result of immunologic processes associated with infection. This applies especially to anterior scleritis. Posterior scleritis is not usually associated with any specific disorder. As with episcleritis, scleritis is only occasionally due to bacterial or viral inflammation.

Table 6.1 Systemic diseases that can cause scleritis

| Frequent causes | Rare causes |
|---|---|
| ❖ Rheumatoid arthritis | ❖ Tuberculosis |
| ❖ Polymyositis | ❖ Lues |
| ❖ Dermatomyositis | ❖ Borreliosis |
| ❖ Ankylosing spondylitis | ❖ Reiter's syndrome |
| ❖ Spondylarthritis | |
| ❖ Vasculitis | |
| ❖ Wegener's granulomatosis | |
| ❖ Herpes zoster ophthalmicus | |
| ❖ Syphilis | |
| ❖ Gout | |

- **Symptoms and findings:** All forms except for scleromalacia perforans are associated with *severe pain* and general reddening of the eye.
- **Anterior non-necrotizing scleritis (nodular form).** The nodules consist of edematous swollen sclera and are *not mobile* (in contrast to episcleritis).

**Segmental episcleritis.**

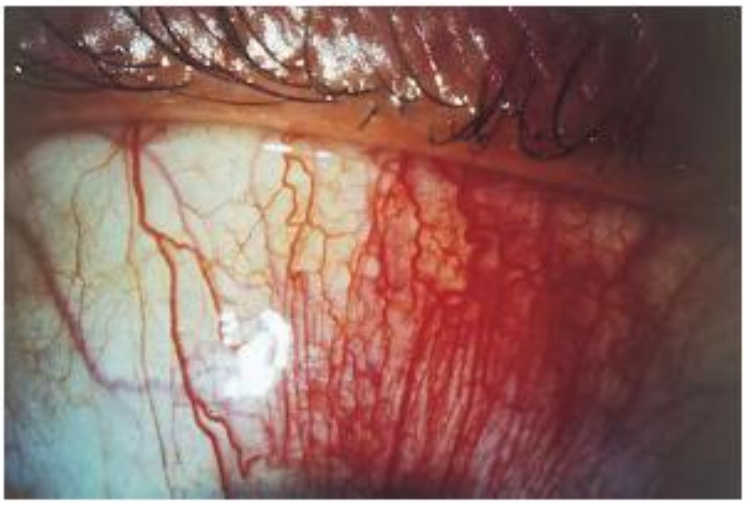


Fig. 6.2 Typical hyperemia and inflammation of the radial episcleral blood vessels.

Diffuse non-necrotizing scleritis.

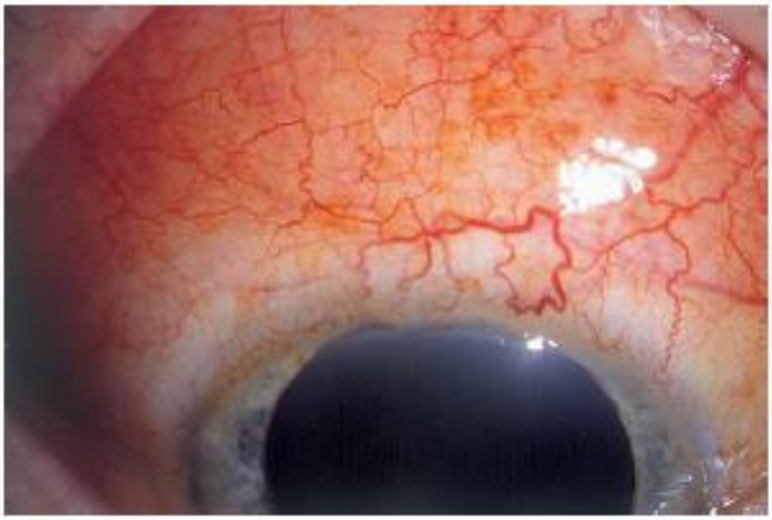


Fig. 6.**3** Typical signs include thickening and edema of the sclera and deep, diffuse reddening.

## Anterior scleritis

- **Anterior necrotizing scleritis (diffuse form)**

The inflammation is more severe than in the nodular form. It can be limited to a certain segment or may include the entire anterior sclera (Fig. 6.**3**).

- **Anterior necrotizing scleritis with inflammation.** Circumscribed reddening of the eyes is a typical sign. There may be deviation or injection of the blood vessels of the affected region, accompanied by avascular patches in the episcleral tissue.
- As the disorder progresses, the sclera thins so that the underlying choroid shows through (Fig. 6.**4**). The inflammation gradually spreads from its primary focus. Usually it is associated with uveitis.
- **Anterior necrotizing scleritis without inflammation (scleromalacia perforans).**
- This form of scleritis typically occurs in *female patients* with a long history of seropositive rheumatoid arthritis. The clinical course of the disorder is usually asymptomatic and begins with a yellow necrotic patch on the sclera.
- As the disorder progresses, the sclera also thins so that the underlying choroid shows through. This is the *only* form of scleritis that *may be painless.*

## Posterior scleritis

- Sometimes there will be no abnormal findings in the anterior eye, and pain will be the only symptom.
- Associated inflammation of the orbit may result in proptosis (exophthalmos) and impaired ocular motility due to myositis of the ocular muscles. Intraocular findings may include exudative retinal detachment and/or choroid detachment. Macular and optic disk edema are frequently present.

- The reddening in scleritis is due to injection of the deeper vascular plexus on the sclera and to injection of the episclera. Inspecting the eye in daylight will best reveal the layer of maximum injection.

Circumscribed scleral staphyloma secondary to scleritis.

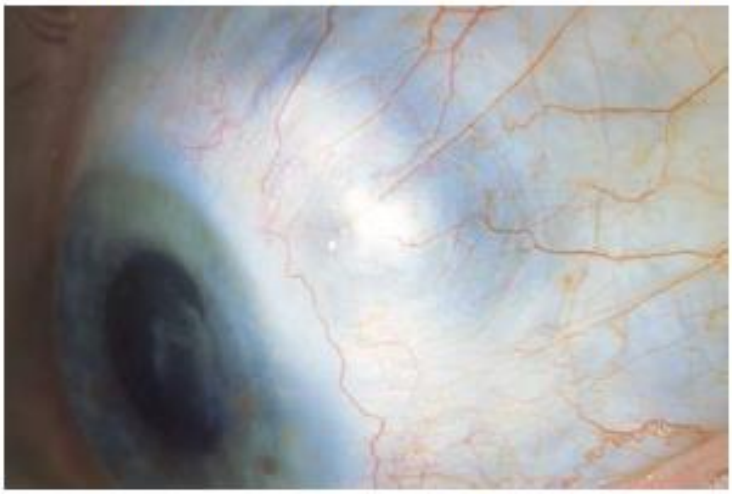


Fig. 6.4 The underlying choroid shows through at the bulge where the sclera is thinned, giving it a bluish tinge.

**Differential diagnosis:**

1. Conjunctivitis
2. episcleritis

**Treatment:**

- **Anterior non-necrotizing scleritis.** Topical or systemic *nonsteroidal* anti-inflammatory therapy.
- **Anterior necrotizing scleritis with inflammation.** Systemic *steroid* therapy is usually required to control pain. If corticosteroids do not help or are not tolerated, immunosuppressive agents may be used.
- **Anterior necrotizing scleritis without inflammation (scleromalacia perforans).**

- As no effective treatment is available, grafts of preserved sclera or lyophilized dura may be required to preserve the globe if the course of the disorder is fulminant.
- **Posterior scleritis.** Treatment is the same as for anterior necrotizing scleritis with inflammation.

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