

---

# Essentials in Ophthalmology, Scleritis: Classification and Clinical Presentations of Scleritis

# 2

Dinesh Visva Gunasekeran, Mi Fang Helen,  
and Rupesh Agrawal

---

## Classification

The international classification of scleritis based on appearance of lesions and anatomical distribution of disease on initial examination was established by Watson PG and Hayreh in 1976 [1], and it is still in use today.

Scleritis is classified anatomically into anterior and posterior scleritis [1] and can also be classified into infectious and non-infectious aetiology [5]. Patients with anterior scleritis can be further subclassified according to morphology into diffuse scleritis, nodular scleritis, necrotising scleritis with inflammation (necrotising), and necrotising scleritis without inflammation (scleromalacia perforans) as in Fig. 2.1 [1]. These are further described under clinical assessment.

In the same landmark paper, Watson and Hayreh also propose a classification of episcleritis into simple episcleritis and nodular episcleritis [1]. However, episcleritis is not the subject of the current chapter as it usually does not give rise to scleritis. The only exception to this rule is herpes zoster which occasionally presents as a self-resolving episcleritis during the vesicular stage of disease and can recur at the same location as scleritis several months later [2].

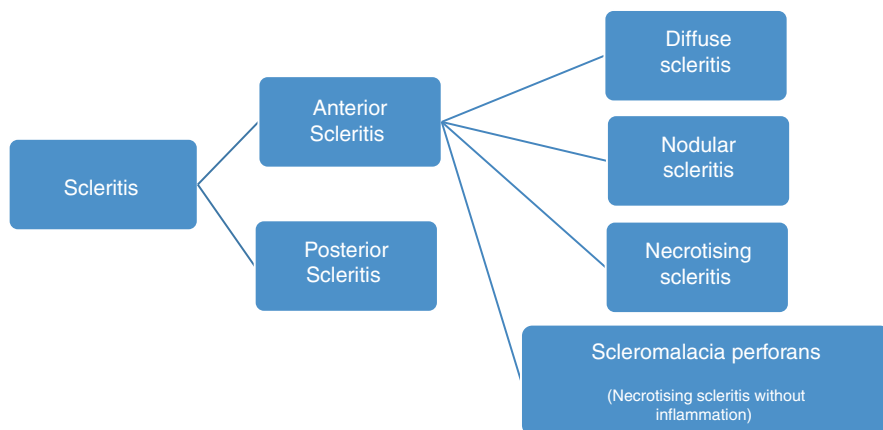
Necrotising scleritis was further divided into four morphologic groups based on histologic appearance by Riono et al. [8]. The four morphologic groups were zonal necrotising granulomatous scleral inflammation; nonzonal diffuse scleral inflammation, with or without granulomatous process; necrotising inflammation with

---

D.V. Gunasekeran, MBBS  
Ministry of Health Holdings, Singapore, Singapore  
Moorfields Eye Hospital (MEH), London, UK

National Healthcare Group Eye Institute, Tan Tock Seng Hospital, Singapore, Singapore

M.F. Helen, MBBS • R. Agrawal, FRCS, MD (✉)  
National Healthcare Group Eye Institute, Tan Tock Seng Hospital, Singapore, Singapore  
e-mail: rupeshttsh@gmail.com



**Fig. 2.1** Anatomical classification of scleritis. This classification was found to be consistent with the natural history of scleritis by Tuft and Watson [2]. Additionally, it was found to have a strong correlation with disease severity and thereby useful to direct therapeutic decisions for patients with scleritis, by Sainz-de-la-Maza et al. [3]. Necrotising scleritis has the worst phenotype, is the most difficult to treat, affects older patients, and is most often associated with systemic diseases

microabscesses, with or without evidence of micro-organisms in the section studied; and sarcoidal granulomatous inflammation [8].

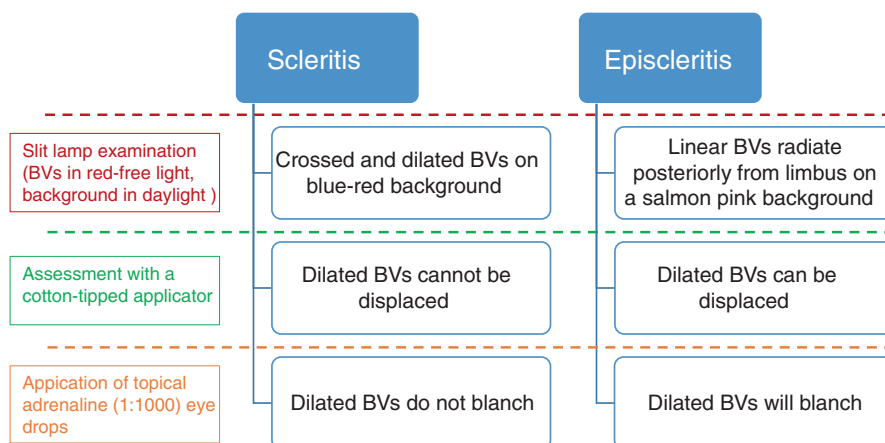
## Clinical Assessment

### Diagnosis of Scleritis

Patients with scleritis often present with the acutely red eye which is frequently quite painful. The location and character of ocular pain as well as the presence of any visual changes are pertinent details to elucidate on history taking. Ocular pain in scleritis tends to be quite severe, can be deep seated, and occasionally radiates in the distribution of the trigeminal nerve from the orbital margin to the ipsilateral temple or jaw, potentially leading to misdiagnoses such as migraine, sinusitis, or even cerebral tumour. Characteristically, it gets worse during the night, frequently waking the sufferer up. It is a pain that does not respond well to common analgesics [1–8].

Other helpful information includes any personal/occupational contact with irritants, as well as personal and family history of ocular/systemic disorders. These may include allergies and autoimmune disorders such as rheumatoid arthritis, gout, connective tissue/dermatological diseases, sarcoidosis, venereal disease, and tuberculosis [1–4].

Important differentials to consider in this presentation include anterior uveitis and acute angle-closure glaucoma (AACG). Another closely related differential which is often confused clinically is episcleritis, which often adopts a benign and



**Fig. 2.2** Clinical differentiation of scleritis from episcleritis

self-limiting course unlike scleritis. Clinical differentiation of scleritis from episcleritis is depicted in Fig. 2.2.

Scleritis can be identified based on the presence and appearance of dilated blood vessels on the episclera on macroscopic inspection with the slit lamp. In scleritis, crossed and dilated vessels are seen on a background of discoloured sclera, which may appear blue red or violaceous in colour. Furthermore, scleral oedema may be observed, appearing as external protrusion of the deep vascular network of the episclera [1–8].

Examination under red-free light is crucial for characterising vascular changes and under daylight for the colour of background scleral and episcleral tissue. In scleritis, the dilated blood vessels are fixed in the sclera and cannot be displaced by cotton-tipped applicator. The converse is true of the close differential episcleritis, whereby dilated episcleral blood vessels can be easily displaced. Episcleritis presents with linear dilated vessels seen to radiate posteriorly from the limbus on a background of salmon pink colour (background best assessed in daylight) and without any scleral oedema. The blood vessels which dilate in both episcleritis and scleritis are episcleral vessels. In episcleritis the superficial plexus is dilated, which show a criss-crossed pattern, easily distinctive since the deeper vessels are not dilated and there is no deep redness. In scleritis, the dilated deep episcleral plexus will not show the same pattern and will be a more diffuse redness frequently associated with oedema [1].

Topical phenylephrine 10% eye drops was used to differentiate between scleritis and episcleritis, whereby blanching of the dilated vessels is observed in episcleritis and not in scleritis. This distinction is pertinent given the more severe disease course in scleritis which impacts management. The use of topical phenylephrine 10% has been mostly abandoned in view of the risk of cardiovascular incidents, especially in older patients. The clinical findings are usually sufficient to make the diagnosis [1].

## Types of Scleritis

The various types of anterior scleritis are differentiated based on the distribution and appearance of scleral lesions. Scleral nodules in nodular scleritis would be observed as immobile, deep-red-coloured nodules that may be tender. Occasionally, there can be associated corneal infiltrate or episcleral congestion. This is easily differentiated from nodular episcleritis as the nodules can be observed as distinct and separate from the overlying episclera under slit lamp microscopic examination [1].

Diffuse scleritis would present with generalised scleral inflammation that may be so severe as to cause associated episcleral and conjunctival oedema that obscures the cornea and protrudes from between the eyelids. Application of topical adrenaline (1:1000) to the conjunctival sac would be useful in such cases to enable assessment of the underlying scleral tissue.

Necrotising scleritis with signs of adjacent inflammation is associated with the worst visual prognosis and increased risk of complications such as scleral thinning. These patients present initially with a localised patch of scleritis associated with severe acute congestion that progresses to yellowish/grey sclera and areas with complete loss of scleral tissue if not treated. Scleral oedema with an overlying/adjacent patch of avascular episcleral tissue in early stages of disease is strongly suggestive and should be treated aggressively to prevent scleral necrosis and eventual perforation.

Some cases of necrotising disease occur following a surgical trauma to the sclera (surgically induced necrotising scleritis – SINS), most frequently associated with limbal incisions in extra-capsular cataract extraction, but also pterygium, especially associated with mytomicin C application, squint surgery, and retinal detachment procedures [1].

Necrotising scleritis without signs of adjacent inflammation (scleromalacia perforans) presents with singular or multiple patches of yellowish/grey sclera without any reactive oedema. This is often followed by development of a surrounding circular crack like a demarcation line, with eventual separation of the affected sclera and overlying episclera like a sequestrum. This can progress to areas with complete loss of sclera. The number of blood vessels in the surrounding episcleral tissue may be diminished, giving the surrounding sclera a porcelain-like appearance when viewed from a distance. Patients most frequently have a history of long-standing rheumatoid arthritis.

Posterior scleritis is far less obvious clinically and presents with a paucity of external signs, giving rise to frequently missed diagnoses in patients that do not manifest concurrently with anterior scleritis. Isolated posterior scleritis is suspected in patients with severely tender proptosis, diplopia, and/or ophthalmoplegia associated with suggestive fundus changes. Fundus changes include papilloedema or exudative retinal detachment appearing as pale grey-white patches surrounded by a dark-grey demarcation line seen through the overlying detachment. Following treatment these changes often resolve, leaving behind a “high-water mark” appearance of a pale white patch with surrounding pigment migration [2].

Nida Sen et al. use the standard clinical images to develop scleritis grading system after using 10% phenylephrine eye drops [7]. The group graded the scleritis into grade 0 (none) with complete blanching on phenylephrine application, grade +0.5 (minimal/trace) with localised pink appearance of the sclera around minimally dilated deep episcleral vessels, grade +1 (mild) with diffuse pink appearance of the sclera around mildly dilated deep episcleral vessels, grade +2 (moderate) with purplish-pink appearance of the sclera with significantly tortuous and engorged deep episcleral vessels, grade +3 (severe) with diffuse redness of the sclera and details of the superficial and deep episcleral vessels cannot be visualised, and grade +4 (necrotising) with diffuse redness of the sclera with scleral thinning and uveal show [7].

## **Complete Clinical Assessment: Aetiology and Complications**

Nearly half of the patients with scleritis may have associated autoimmune connective tissue or vasculitic diseases that may be undiagnosed. Complete assessment of affected patients would include history taking (as described under diagnosis), dermatologic examination (of the skin, hairline, and nails), and rheumatological examination (of the joints) for relevant clues to exclude autoimmune associations. These are further discussed in a later chapter.

Although less common than autoimmune disease, infectious aetiology is just as important accounting for up to 10% of all patients with scleritis. These patients may have a history of relevant risk factors including trauma, immunosuppression, and ocular surgery, particularly pterygium excision surgery. Patients often present after a latent interval following the inciting event, with a painful red eye associated with epiphora. Pain is often severe and may be out of proportion to clinical signs. Clinical signs that may be observed include decreased visual acuity, significant anterior chamber flare, and evidence of satellite lesions or extension to surrounding structures. Involvement of extraocular muscles mimicking orbital inflammatory syndrome can also develop. Infectious scleritis is also further explored in a later chapter.

Finally, complete assessment of patients with scleritis should also include that for potential complications. This would include visual acuity testing, slit lamp examination for corneal involvement, anterior chamber inflammation, vitreous inflammation, and/or avascular patches (avascular patches are suggestive of vasculitic aetiology and imply a worse prognosis in either scleritis or episcleritis). Complications of scleritis that may be encountered include decreased visual acuity, anterior/posterior uveitis, keratitis, elevated intraocular pressure (IOP), retinal detachment, scleral thinning, and staphyloma (protrusion of the iris into thinned sclera in patients with elevated IOP).

It is also important to mention that there is a high risk of mortality in cases of necrotising scleritis associated with a systemic vasculitis. This makes the correct and quick diagnosis essential for the introduction of aggressive systemic therapy which can be life-saving.

**Compliance with Ethical Requirements** Authors declare that we have no conflict of interest.

No animal or human studies were carried out by the authors for this article.

---

## References

1. Watson PG, Hayreh SS. Scleritis and episcleritis. *Br J Ophthalmol*. 1976;60(3):163–91.
2. Tuft SJ, Watson PG. Progression of scleral disease. *Ophthalmology*. 1991;98:467–71.
3. Sainz-de-la-Maza M, Jabbur NS, Foster CS. Severity of scleritis and episcleritis. *Ophthalmology*. 1994;101:389–96.
4. Sainz de la Maza M, Molina N, Foster CS, et al. Clinical characteristics of a large cohort of patients with scleritis and episcleritis. *Ophthalmology*. 2012;119(1):43–50.
5. Ramenaden ER, Raiji VR. Clinical characteristics and visual outcomes in infectious scleritis: a review. *Clin Ophthalmol*. 2013;7:2113–22.
6. Homayounfar G, Borkar DS, Acharya NR, et al. Clinical characteristics of scleritis and episcleritis: results from the pacific ocular inflammation study. *Ocul Immunol Inflamm*. 2014;22(5):403–4.
7. Sen HN, Sangave AA, Goldstein DA, Suhler EB, Cunningham D, Vitale S, Nussenblatt RB. A standardized grading system for scleritis. *Ophthalmology*. 2011;118(4):768–71. doi:10.1016/j.optha.2010.08.027. Epub 2010 Nov 20. PubMed PMID: 21093921; PubMed Central PMCID: PMC3070789.
8. Riono WP, Hidayat AA, Rao NA. Scleritis: a clinicopathologic study of 55 cases. *Ophthalmology*. 1999;106(7):1328–33. PubMed PMID: 10406616.

Scleritis

Pavesio, C. (Ed.)

2017, X, 123 p. 37 illus., 34 illus. in color., Hardcover

ISBN: 978-3-319-49913-0