DISEASES OF THE EPISCLERA AND SCLERA

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GENERAL CONSIDERATIONS

- The sclera is inert and mainly supportive in function
- It is essentially collagenous
- Its structure is almost acellular and avascular
- Other than “inflammation” diseases are rare
- The pathophysiology of scleral disease is relatively simple
- The sclera is usually resistant to treatment (due to torpidity)
- Healing of the sclera is dependent on neighbouring tissues

REVIEW OF SCLERAL ANATOMY

- The sclera is also referred to as the *tunica sclera oculi*
- It has a brown lamina fusca in the suprachoroidal space
- It has a viscoelastic structure with white fibrous tissue made up of fine elastic fibers and connective tissue
- The sclera is a factor in intraocular circulation and intraocular pressure
- The fiber bundles of the sclera are regularly arranged thereby affording it its translucency
- The sclera makes up 5/6th of the outer coat of the eye, interrupted by the optic nerve and emissaria
- The emissaria may be pigmented or appear cystic
- The blood vessels in the sclera are scanty and capillaries are widespread

The circulation overlying the sclera includes 3 vascular layers:

- **Conjunctival plexus**, the most superficial plexus of fine vessels, which is movable over the underlying structures
- **Superficial episcleral plexus**, which consists of radially arranged vessels lying at the level of the Tenon capsule
- **Deep episcleral plexus**, also known as the scleral plexus, which lies deep to Tenon's capsule and directly over the sclera.

CONGENITAL ANOMALIES OF THE SCLERA

SCLERAL CYSTS

BLUE SCLERA

Found in:

- Young children
- Van der Hoeve and Kyne syndrome
- Osteogenesis imperfect
- Marfan's syndrome

OCHRONOSIS

Found in:

- Inborn errors of metabolism (phenylalanine and tyrosine)
- Alkaptonuria
- Pigmented sclera

MELANOSIS OCULI

Naevus of ota (compared with an acquired lesion)

Why are some sclera blue?

- The reason is the same as why superficial blood vessels in pale skinned people also appear blue rather than red
- Thinning and/or increased translucency leads to Rayleigh scattering of light

SCLERAL INFLAMMATION

Can either be:

EPISCLERITIS

- Simple (diffuse) found in 75% of cases
- Nodular found in 25% of cases

SCLERITIS

- Anterior
- Diffuse found in 35% of cases
- Nodular found in 45 % of cases
- Posterior found in 2% of cases
- Necrotizing
- With inflammation found in 10% of cases

Also:

- Scleromalacia perforans
- Sclerokeratitis

EPISCLERITIS

HISTORICAL NOTE

Episcleritis periodica fugaz was discovered by Fuchs in 1895.
Previously termed “subconjunctivitis” by von Graefe (1885) and “hot eye” by Hutchinson in 1885

WHAT IS EPISCLERITIS

Episcleritis is a diffuse congestion and oedema of an area of episcleral tissue and the conjunctiva over it, of a transient character and a strong tendency to recur with regular periodicity.

LESS COMMON SIGNS AND SYMPTOMS

- Pain and photophobia
- Spasms of iris sphincter and ciliary muscle (miosis and transient myopia)
- Angio-oedema of the lids
- Associated with migraine
- Family hx of atopy

OBJECTIVE

- Benign (no permanent damage)
- Self-limiting (does not progress to scleritis)
- Mild discomfort and tenderness
- Affects young adults
- Watering
- Dilated conjunctival and episcleral vessels
    - Sectoral or diffuse redness
- Rarely associated with systemic conditions

ASSESSMENT

Use of topical phenylephrine (2.5%) or 1:1000 epinephrine usually results in blanching of superficial blood vessels
One can manipulate the blood vessels

PLAN

- Depends on severity of condition
- Mostly self-limiting

- Without treatment:
- Improves 50% within 1st week
- Can resolve completely within 3 weeks

NODULAR EPISCLERITIS:

- Characterized by a nodule surrounded by injection
- Slit lamp examination shows that the sclera is intact
- Superficial scleral translucency can be a common sequel of the condition

MANAGEMENT

Mild

- No treatment
- Artificial tears
- Topical decongestants
- Aspirin/other non-steroidal anti-inflammatories
- Cold/warm compresses
- Mild topical steroids like FML

Severe

- Topical steroids (prednisolone 1%)
- NSAIDs
- Oral steroids (rarely needed)

SCLERITIS

Definition: Scleritis is inflammation of the sclera and is characterized by edema and cellular infiltration of the sclera and episclera. It is classified as either posterior or anterior.

- Granulomatous inflammation
- Mild to destructive disease
- Severe pain and photophobia
- Visual disturbance
- Deep red (+ associated episcleritis)
- Vaso-occlusion → necrosis

MOST COMMON SYSTEMIC CAUSES:

- Rheumatoid Arthritis
  - 1:200 develop scleritis
- Connective Tissue Disease
  - Wegener granulomatosis (form of vasculitis that affects the lungs, kidneys and other organs)
  - Systemic lupus erythematosus
  - Polyarteritis nodosa
- Herpes Zoster Ophthalmicus
- Miscellaneous
  - Surgically induced
  - Infectious

ANTERIOR SCLERITIS

Anterior scleritis is a sight-threatening eye disease, characterized by acute or chronic red eye and insidious, deep, boring pain that localizes to the sclera but is often referred to the temple and jaw.

Other complications of anterior scleritis include keratitis, cataract, uveitis and glaucoma.

ANTERIOR NON-NECROTIZING SCLERITIS

- Most common forms of anterior scleritis
- Diffuse
  - Similar presentation to episcleritis
  - More severe
  - More discomfort
  - There is a distortion of the vascular plexus and loss of the radial pattern
- Nodular
  - Resembles nodular episcleritis but more severe
  - Nodule involves the underlying sclera and cannot be moved

ANTERIOR NECROTIZING SCLERITIS

- With inflammation
  - Most severe form of scleritis
  - 75% develop visual impairment
  - 25% die from associated vascular disease within 5 years of onset
  - Complications include:
    - Corneal involvement
    - Cataract
    - Secondary glaucoma
  - Within 10 days it can result in ulceration and discharge of damaged collagen

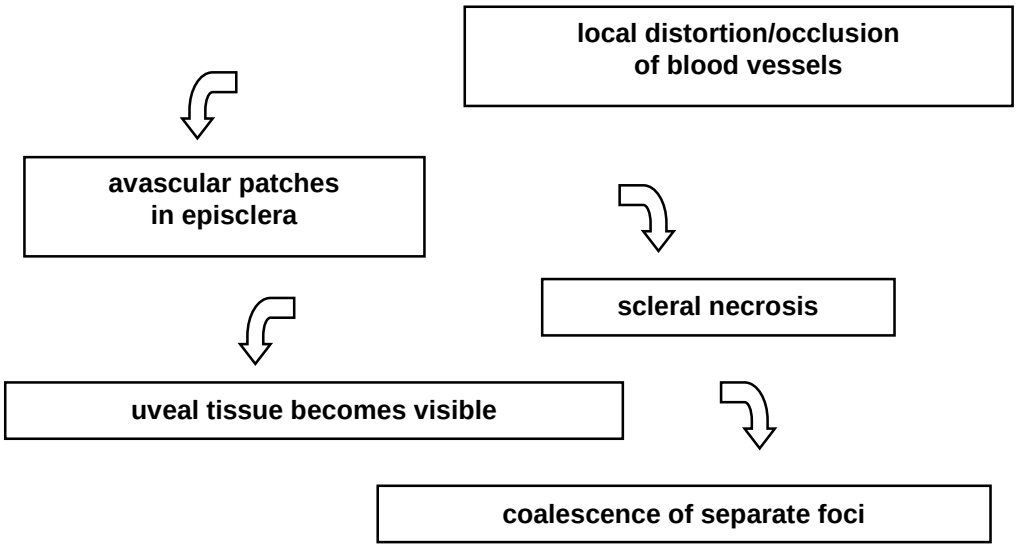

Figure 1: *Progression in anterior necrotizing scleritis*

- Without inflammation (scleromalacia perforans)
  - Predominantly in women with long standing rheumatoid arthritis (RA)
  - No or mild symptoms
  - Yellow necrotic patch spreads
  - Rarely perforates

SCLEROKERATITIS

- Complication of anterior scleritis

- Active limbitis
- Acute elevation of IOP
- White corneal opacity and lipid deposition results

SCLERITIS: COMPLICATIONS AND SEQUELAE

- Keratitis
- Uveitis
- Cataract
- Retinal oedema, exudates, detachment
- Optic neuropathy

SCLERITIS: ASSOCIATED SYSTEMIC DISEASES

- Collagen diseases
  - Rheumatoid arthritis, systemic lupus erythematosis, ankylosing spondylitis, etc.
- Granulomatous diseases
  - TB, syphilis, sarcoidosis, leprosy
- Metabolic disorders
  - Gout, Graves' disease
- Infections
  - Herpes (simplex/zoster), onchocerciasis
- Other
  - Trauma, iatrogenic

SCLERITIS: INDICATED LABORATORY TESTS ASSESSMENT

- WBC with differential
- ESR or equivalent
- Hemoglobin
- Electrolytes and protein
- Uric acid
- Serological tests for syphilis
- Imaging: chest, hands, feet, spine

SCLERITIS: MANAGEMENT

- Treatment of any underlying disease
- Topical steroids (not very effective):
  - Prednisolone 1%
  - Dexamethasone 0.1%
- Oral NSAIDs:
  - Ibuprofen 400 mg tid
  - Flurbiprofen 100 mg tid
  - Oxyphebutasone 600 mg/d
- Oral steroids:
  - Prednisone 60 mg/d increase by 20 mg
- Immunosuppressive agents:
  - Cyclophosphamide
- Subconjunctival steroids are contraindicated

SCLERITIS: MANAGEMENT FOR EXTREME THINNING OR PERFORATIONS

- Debridement of any infected tissue
- Lamellar or full thickness graft:

- Homologous donor sclera
- Other grafting materials:
  - Homologous donor cornea
  - Aortic tissue
  - Autologous periostium
  - Autologous fascia lata

DIFFERENTIAL DIAGNOSIS BETWEEN SCLERITIS AND EPISCLERITIS

Scleritis	Episcleritis
Deep vessels	Sclera not involved
No blanching	Blanching with phenylephrine
Females > males	Younger adults
Severe pain (radiates)	Discomfort rather than pain
Gradual onset (recurs)	Acute onset (recurs)
Sclera may be bluish	Movable vessels
Many patients have systemic disease	Usually idiopathic but may have collagen vascular, gout, infection

POSTERIOR SCLERITIS

BACKGROUND

- Underdiagnosed
- 30% related to systemic disease
- Pain, tenderness
- Disc oedema
- Macular folds (oedema)
- Proptosis
- EOM restrictions
- Exudative retinal detachment

DIAGNOSIS

- Refraction (hyperopic shift)
- Slit-lamp biomicroscopy
- Ophthalmoscopy
- Ocular motility
- B-scan ultrasound – shows a “lazy-T” sign, a thickened sclera and fluid in the sub-Tenon space
- CT scan

OTHER FUNDUS SIGNS

- Vitritis
- Choroidal detachment
- Choroidal folds
- Intraretinal deposits
- Subretinal exudates
- Retinal detachment

MANAGEMENT

- Elderly patients
  - Oral prednisolone
  - Immunosuppressive agents
- Young patients
  - Oral NSAIDs

INFECTIOUS SCLERITIS

BACKGROUND

- Rare
- Must be included in the differential diagnosis with scleral necrosis and abscess formation in the presence of a risk factor

RISK FACTORS

- Prior scleritis
- Recent or remote surgery involving the sclera
- Ocular trauma or irradiation

IDENTIFIED ORGANISMS

- Pseudomonas aeruginosa
- Staphylococcus Sp.
- Haemophilus pneumoniae
- Aspergillus fumigatus
- Acanthamoeba sp.
- Varicella zoster
- Mycobacterium tuberculosis (rare)

TUMOURS OF THE SCLERA

- Do not occur
- Sclera acts as a barrier for spread of intraocular tumours (note: scleral canals provide ports)
- Sclera and/or episclera may be involved by neoplastic changes in orbit, within and outside the globe.
- Dilation and tortuosity of deep episcleral vessels and new vessel formation suggests: intraocular malignant melanoma, myeloma or sarcoidosis