Scleritis: Diagnosis and Management

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Plan of presentation

• Overview of episcleritis and scleritis
• Classification
• Clinical features
• Case examples: DD
• Investigations
• Management guidelines
Introduction

• Inflammation of the sclera with characteristic clinical picture
• Painful, indolent, locally destructive
• May be associated with serious systemic diseases
• Cornea, episclera and uvea
Extent of the problem

- Scleral disease is rare
- Necrotizing scleritis with RA:
  - 27% are dead in 5 years (Watson)
  - 45% are dead in 3 years (Mc Gavin et al)
  - 9% died in 11 years (de la Maza et al)
Associated with systemic disease:

40% to 57% of patients with scleritis
- 30% to 48% have an associated connective tissue or vasculitic disease
- 5% to 10% an infectious etiology
- 2% have atopy, rosacea, or gout
Practical management

• Is it episcleritis or scleritis or masquerade?
• Is it necrotising or non-necrotising
• Is it infectious or not?
• Which investigations are necessary?
• What is the management strategy?
Episcleral Inflammation

History of systemic diseases, pain, blanching with phenylephrine
Episcleral Inflammation

Symptoms: Irritation
Absence of pain
Blanching with phenylephrine
Scleral Inflammation

Examination findings
Episcleritis

- One third:
  - systemic vasculitic
  - connective tissue diseases
  - local ocular conditions
Episcleritis

• **One third:**
  - RA, IBD, SLE, myositis, RP, erythema nodosum, GPA, Cogan syndrome
  - Atopy, rosacea, gout, herpes zoster, herpes simplex, syphilis, psoriasis,
  - Drug reactions (pamidronate, alendronate, risedronate)
  - Pediatric patients: with rheumatoid arthritis (rare in age less than five)
Examination: scleritis

- Day light
- Violaceous hue/
diffuse bluish red
- Scleral thinning,
translucency
- Uveal exposure
Examination: scleritis

- Day light
- Violaceous hue/ diffuse bluish red
- Scleral thinning, translucency
- Uveal exposure
• Maximum congestion in deep episcleral network
• SLE: slit beam displaced forward
• Red free light: highlights blood vessels: capillary drop out
Classification

SCLERITIS

Non Infective  Infective

Anterior  Posterior

Diffuse  Nodular  Necrotizing

With Inflammation  Without Inflammation
Scleritis with peripheral ulcerative keratitis

- Increased chance of systemic disease association
Case D-P: 26/06/03

History:
- 37 year old HW
- Severe r/w/p OU since one year
- Underwent OU cataract surgery 2 years ago
- Systems: H/o dysuria for 10 years

Examination:
- BCVA: OU 20/20, N6
Investigations:

- CBP, ESR, XRC, Mantoux, ANA, RA, CRP, HIV, VDRL

Treatment:

- Oral steroids, methotrexate, cyclophosphomide, IV MP
- P and C-ANCA, ASO, Anti ds-DNA, SS-A, SS-B: all negative
Anterior Scleritis: RP

June’ 03

Sept’ 04

Oct ’ 05
Case D-P: Sept’ 04 (14 months)

- Triad of fleeting joint pains, cartilage involvement (nasal septum) and scleritis

Wegener’s granulomatosis?
Relapsing polychondritis?
Case D-P: Relapsing polychondritis

- Rare, systemic autoimmune disorder
- Diagnosis is based on clinical symptoms
- Three of the following:
  
  McAdam et al: 6 clinical criteria
  - Recurrent chondritis of both auricles
  - Non-erosive inflammatory polyarthritis
  - Chondritis of the nasal cartilage
  - Inflammation of ocular structures
  - Respiratory tract chondritis
  - Cochlear or vestibular damage
• 68 year old lady
• Presented on 26/4/2014
• Complaints of redness, pain and watering in right eye since 10 days
• Vision 20/200---20/50
History

✓ Granulomatosis with Polyangitis (Wegeners)
  ✓ Rheumatoid Arthritis (?)
  ✓ Pulmonary Kochs
  ✓ Hypertension

• Using oral steroids and ATT

• Already received recently 2 doses of Intravenous cyclophosphamide and methyl prednisolone; stopped later after the diagnosis of pulmonary Kochs
Investigations
At presentation

- CBP, LFT and RFT - WNL
- C-ANCA – positive
- RA factor – raised
- Sputum - positive for AFB
- ESR – 70mm in 1st hour
- ANA – negative
- CRP – raised
- S. Anti PR3 – positive
26-04-14: Active necrotising scleritis
Received 2 doses of pulse IVMP and Cyp elsewhere
Topical steroids

02-05-14:
Topical, oral steroids & MTX
Pulse IVMP & CyP

09-05-14:
Status improved
Monthly follow up with same treatment

08-09-14:
Steroids reduced
Azathioprine started

16-01-15:
Steroids continued at 5mg dose
Azoran stopped
C-ANCA - negative

15-02-16:
Healed scleritis with limbal staphyloma
On topical NSAIDS, oral steroids (6 mg) & VCZ

Jan 16:
Cavitatory pulmonary aspergillosis
PCR positive
Started on Voriconazole & Augmentin

16-01-15:
Steroids continued at 5mg dose
Azoran stopped
C-ANCA - negative

Jan 16:
Cavitatory pulmonary aspergillosis
PCR positive
Started on Voriconazole & Augmentin
26-04-14:

08-09-14:

Course of disease

15-02-16:
Infectious Scleritis

**Exogenous**
Surgery
Trauma
Extension from adjacent infections

**Endogenous:** systemic infection

Varying incidence of Infectious scleritis between studies 5 – 18%
# Infectious Scleritis - Organisms

## Bacteria
- Staphylococcus spp
- Streptococcus spp
- Pseudomonas aeruginosa
- Corynebacterium
- Nocardia spp
- E coli
- Mycobacterium
- Treponema paalidum
- Borrelia

## Fungii
- Aspergillus spp
- Paecilomyces spp
- Sporothrix Schenckii
- Cephalosporium
- Cladosporium Fusarium
- Rhizopus

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Infectious scleritis

Viruses
- Ebstein barr
- Coxsackie B5
- Varicella Zoster
- Herpes Simplex

Amoeba
- Acanthamoeba

Parasite
- Toxoplasma
- Toxocara


Case 7: Sclerokeratouveitis

- 35-year-old man
- C/o redness, decreased vision and pain for 1 month
- Diagnosis: scleral nodule? infective etiology
- Excision biopsy after systemic exam
Scleral biopsy and patch graft
M. leprae
Three months post-op

- Sumptomatically better
  - VA: 20/60 – 20/50p
- IOP: OD – 20mmHg/ OS – 16mm Hg

Anti-lepra therapy: (restarted as patient had discontinued)
Unusual scleritis

- 14 year old girl
- C/o recurrent redness and pain for 1 yr Rt eye (more since 4 days)
- Diagnosis: scleral nodule ?infective etio
- Scleral biopsy after UBM
H&E: 10X

H&E: 40X

Tubercular scleritis
Unusual scleritis: masquerade
Laboratory Investigations

When?

- Persistent or recurrent episcleritis
- All scleritis versus bilateral scleritis
- Unilateral necrotising
- Suspected infection
Screening evaluation with:

- CBC, ESR/ CRP
- Rheumatoid factor, S. ANCA
- Anti Nuclear Antibodies*,
- FTA- ABS
- PPD and X-ray chest

*Female patients:
Laboratory Investigations

- All scleritis need to be investigated
- Diffuse anterior, unilateral scleritis: commonest
- Necrotising scleritis: may have seropositivity for ANCA, ANA, RA

Interpretation:
- Raised ESR, CRP
- S.ANCA levels
- Other tests: AntiCCCP
- HLA B27, other HLAs
- Positive Mantoux
- In case of viral suspicion
Laboratory Investigations

- **Management:**
  - CBC, RBS,
  - XRC, Mantoux,
  - S. HIV, S. VDRL

- **Etiological:**
  - RA, ANA
  - C & P ANCA, UE
  - Ds DNA, Anti-Rho, Anti-LA

- **Local:**
  - Ant FA
  - UBM, B-scan
  - Scrapings
  - Impression cyto
  - Biopsy
Scleritis Therapy

1. NSAIDS
2. Systemic Steroids
3. Therapeutic failure
4. Immunomodulators: Methotrexate, Cyclophosphamide, Cyclosporine, Azathioprine

Remission: Maintain on NSAIDS

Biologics: Infliximab, Eternecept, Rituximab
Scleral or corneal patch graft
Multi-layered amniotic membrane
Fascia lata, periosteum
Pre, peri-operative immunosuppression
Conclusions

- Diff. inflammatory vs non-inflammatory
- Infectious vs non-infectious
- Episcleritis vs scleritis
- Necrotising: vasculitic disease, needs immunospressives
- WG & PAN: Cytoxan+ systemic steroids
- Adequate medical Rx before any surgical intervention
Thank you!

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