Ocular Sarcoidosis

Careen Y. Lowder, M.D., Ph.D.
Cole Eye Institute
The Cleveland Clinic Foundation
Ocular Epidemiology

30-60% of patients with known sarcoidosis have ocular involvement at some point in the disease.

4-6% of patients with uveitis develop clinical sarcoidosis.
How do we diagnose ocular sarcoidosis?

Gold standard for diagnosis is histological proof using biopsy tissue

We do not usually biopsy intraocular structures so we rely on ancillary blood tests and imaging such as chest xray and chest CT
Skin disease

Erythema nodosum
Subcutaneous nodules
Lupus pernio (purple lupus)

Skin disease

Erythema nodosum
Subcutaneous nodule
Lupus pernio (purple lupus)
Other systemic disease

Lymphadenopathy (75%)
CNS
Bone involvement
Connective tissue
Cardiac
Renal
Sinus
Clinical Presentation

Orbital myositis
Scleritis
Lacrimal gland involvement
Conjunctival granulomas
Lacrimal infiltration
Orbital Myositis
Ptosis/lid signs
Conjunctival granulomas
Ocular sarcoidosis
Pathology

- Hallmark is the non-caseating granuloma
- Epithelioid histiocytes, monocytes, and fibroblasts
- In long-standing disease, Langerhans giant cells form
Conjunctival Biopsy

Ocular sarcoidosis
Ocular sarcoidosis
Anterior signs

Scleritis

Ocular sarcoidosis
Uveitis- Ocular Inflammatory Diseases
Anatomic classification of uveitis

Based on site of inflammation
Anatomic classification

Anterior uveitis

Primary site of inflammation is anterior chamber

Iritis
Iridocyclitis

Cells in anterior vitreous
Anatomic classification

Intermediate uveitis

Primary site of inflammation is vitreous

Vitritis

Pars planitis
Anatomic classification of uveitis

Posterior uveitis

Primary site of inflammation is retina or choroid

- Retinitis
- Choroiditis
- Chorioretinitis
- Retinochoroiditis
Anatomic classification

Panuveitis

Inflammation in anterior chamber, vitreous and retina and/or choroid

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Classification of uveitis: timing

Onset
  - Sudden
  - Insidious

Course
  - Limited – less than 3 months
  - Persistent – greater than 3 months
Characterization of uveitis

Acute
  Characterized by sudden onset and limited duration

Recurrent
  Repeated episodes activity separated by period of inactivity, off treatment, of > 3 months

Chronic
  Persistent uveitis
  Relapse with cessation of therapy
Clinical Presentation

- Anterior uveitis
- Iris nodules
Classification of uveitis

Granulomatous vs. non-granulomatous
Ocular sarcoidosis
Granulomatous iris nodules
Granulomatous iris nodules
Granulomatous KP

Ocular sarcoidosis
Granulomatous iritis
Granulomatous KP

Hyalinized (ghost) KP

Corneal edema

Pigmented old KP
Koeppe nodules

Synechiae form at nodules
Anterior uveitis: symptoms

Acute
- Sudden onset
- Pain, redness, photophobia
- Definite start date
- Self-limited

Chronic
- Insidious onset
- Asymptomatic
- Floaters, decreased vision
- History often less precise
- Long duration

Ocular sarcoidosis
Anterior Uveitis: Signs

**Acute**
- Limbal injection
- KP
- AC reaction (may have hypopyon)
- Posterior synechiae, PAS
- IOP often low

**Chronic**
- Usually no injection
- KP
- AC reaction
- Posterior synechiae, PAS
- IOP often high
Sarcoid uveitis

Classically granulomatous iridocyclitis
  Mutton fat KP
  Iris nodules
May also be non-granulomatous
Usually bilateral
Most often chronic, but may begin with acute disease
Frequent posterior segment involvement
May occur without apparent systemic disease
Panuveitis refers to intraocular inflammation involving all of the uvea with anterior segment, vitreous, retinal and choroidal inflammation.

Ocular Sarcoidosis is the most common panuveitis syndrome.
Sarcoid uveitis

Anterior segment disease
  Young
  African American
  Female

Posterior segment disease
  Older
  Caucasian

Work-up
  ACE, lysozyme
  CXR
  CT chest
  Biopsy
Ocular sarcoidosis
Normal Anterior Segment
Anterior granulomatous uveitis
Retrocorneal Membrane
Keratic precipitates and iritis
Cataract and posterior synechiae
Cataract
Clinical Presentation

Vitritis
Snowbanking and snowballs
Vitreous Snowballs
Posterior Segment

Normal optic nerve and retinal blood vessels
Ocular sarcoidosis

Posterior signs

Clinical Presentation

- Retinal periphlebitis
- Granulomas
- Preretinal nodules
- Serous retinal detachment
- Secondary cystoid macular edema
- Choroidal granulomas
- Disc granuloma
Periphlebitis
Papillitis/Candlewax
Periphlebitis

Papillitis

Ocular sarcoidosis
Severe retinal vasculitis
Retinal granulomas

Ocular sarcoidosis
Retinal Granulomas
Choroidal Granulomas
Serous retinal and choroidal detachment
Left optic nerve involvement
Cystoid Macular Edema
Macular edema
Resolved post treatment
Diagnosis

Biopsy
Chest CT – More sensitive
Less sensitive - Chest X-ray and Serum lysozyme
Angiotensin Converting Enzyme
Cutaneous Anergy
Chest CT

- Valuable test in the work-up of uveitis in the elderly
- More sensitive than standard chest x-ray in detecting mediastinal lymphadenopathy
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Treatment of acute anterior uveitis

Topical corticosteroids
- Pred Forte (prednisolone acetate 1%), hourly (or more) if severe
- Durezol (difluprednate ophthalmic emulsion 0.05%), QID

Dilators
- e.g. homatropine 5% BID
- Pain relief
- Break acute synechiae
- Prevent development of new synechiae
- Longer lasting agent if dark irides, shorter if light
Treatment of chronic iridocyclitis

Topical steroids – less frequent than for acute
May need chronic dilators
Depot steroids
e.g. subtenon triamcinolone acetonide
   TRIESENCE™ (triamcinolone acetonide injectable suspension) 40 mg/mL (Alcon)
   Kenalog 40 mg/ml. Not formulated for intraocular use

Oral steroids
   Avoid long term use

Immunosuppressive/immunomodulatory agents
Treatment of chronic iridocyclitis

Use lowest dose of anti-inflammatory agents to prevent sequelae but decrease risk of glaucoma

Concept of chronicity
  “chronic disease is chronic”

Treatment may be long term

Patients often asymptomatic

Compliance
Treatment of vitritis, posterior, panuveitis or macular edema

Posterior Sub-Tenon’s or intravitreal triamcinolone injections

Systemic Corticosteroids

Steroid sparing agents
  Methotrexate
  Leflunomide
  Cellcept
Uveitic Glaucoma

Prevalence

10 – 46% develop secondary glaucoma

Depends on type of uveitis

Age at presentation

Chronicity and severity of uveitis

Uveitic Glaucoma
Prevalence

26% - 79% uveitis patients develop CME

48.8% CME patients treated with intravitreal triamcinolone develop IOP > 5 mmHg

27.9% develop IOP > 10 mmHg

Estafanous M, Lowder CY. Patterns of macular edema in patients with uveitis. Ophthalmol 2005; 112:360
Mechanisms of Glaucoma in Uveitis

Open Angle Mechanisms

Steroid response
Clogging by inflammatory cells, proteins, debris or fibrin
Mechanisms of Glaucoma in Uveitis

Steroid Induced Glaucoma
1/3 normal population increase of 6-15 mmHg
5% normal population high response >15 mmHg
Takes several weeks (in POAG, within 4-8 hours)
Children - earlier response
Steroid Induced Glaucoma

Dose

Corticosteroid compound

Frequency and route of delivery

Duration of treatment

Patients susceptibility
Mechanisms of Glaucoma in Uveitis

Closed Angle
Pupillary block due to 360 posterior synechiae or complete pupillary membrane
Nonpupillary block angle closure due to inflamed ciliary body rotating forward leading to angle closure
Peripheral anterior synechiae
Neovascular glaucoma
Ocular Sarcoidosis

Patients do not lose vision with proper treatment and control of inflammation.