

BOARD and OKAP REVIEW COURSE

Diagnosis - Management of Anterior Uveitis Syndrome

Lawrence V. Najarian, M.D.

Revised 2004

Diagnosis – Management of Anterior Uveitis Syndrome

Classification of Uveitis

- Oldest clinical classification scheme involves the differentiation between granulomatous and non-granulomatous uveitis.

<u>Intraocular Inflammation</u>		
General	More frequently involved anterior and Posterior uveal areas. Insidious onset, chronic course	Anterior Uvea most frequently involved. Acute onset, short course.
Anterior Segment	Insidious onset, mild, eye nearly white. Nodules, Floccules on iris. Medium and large (often “mutton-fat”) Keratic precipitates. Slight flare.	Acute onset, Red eye, no nodules or floccules on iris; heavy fibrinous exudates, intense flare.
Posterior segment	Common in choroids retina. Heavy vitreous exudates.	Rare Choroidal, retinal lesions. Fine vitreous opacities.

Examples of granulomatous uveitis etiologies: TB, syphilis, sarcoid, brucellosis, toxoplasmosis.

Endogenous vs. infectious

Anatomic Classification: International Uveitis Study Group, 1989

Anterior uveitis

Intermediate uveitis

Posterior uveitis

Pan uveitis

Guidelines for the IUSG recommended classification of Uveitis

Onset and duration of disease

“Acute” 2-6 weeks

“Short” Under 3 months

“Chronic” Greater than 3 months

Unilateral or bilateral

Demographics (age, sex, race)

Response to therapy

Is the disease steroid-responsive, or dependent or resistant?

Anatomic position

Pattern, i.e., single vs. recurrent episodes

Granulomatous vs. non-granulomatous

Severity of visual loss: severe vs. mild

Severe = visual loss of 50% or more compared to predisease state

General Medical History

Pediatric Anterior Uveitis

Uveitis in children under age 16 uncommon.

Comprise only 5-6% of all uveitis cases.

Etiology found in about 50% of all anterior and posterior cases

Presentation

Acute: Photophobia, decreased vision, pain, redness

Insidious: Increased lacrimation, decreased vision found on routine vision screening (JRA).

Differential diagnosis:

Trauma/child abuse

Sympathetic ophthalmia

Juvenile Rheumatoid Arthritis (see below)

Leading cause, by far, of anterior uveitis.

Risk Factors for uveitis include both pauciarticular disease & ANA positivity.

Sarcoid: Characteristic triad. No lung involvement

Granulomatous uveitis

Arthritis – non tender effusion of knees and wrists

Rash

Non specific – erythema nodosum

Specific – non caseating granulomas

Herpes Simplex – may be associated with hyphema, glaucoma & iris atrophy

Herpes Zoster – Usually develops during convalescence from a varicella infection

Syphilis:

Hutchinsons' Triad: 8th Never deafness, interstitial keratitis, widely spaced & notched teeth

Salt and pepper fundus

Saber shins

Saddle nose deformity

Kawasaki Disease: Mucocutaneous Lymph Node Syndrome

Characterized by 5-day fever, rash, lymphadenopathy, conjunctivitis, mucous membrane involvement, in most Oriental children.

Acute, bilateral, non-granulomatous inflammation in 80% of patients, self limited.

Anterior Uveitis Syndrome

Rheumatoid Arthritis

Adult Rheumatoid Arthritis:

NOT associated with increased risk of anterior uveitis

Sjogren's Syndrome

Scleritis

Sclerokeratitis

Juvenile Rheumatoid Arthritis (JRA)

Leading cause of pediatric uveitis

Leading cause of chronic crippling disease of children

Predictive indicators for increased risk of uveitis include:

Pauciarticular (less than 4-joint involvement) JRA

Seronegativity for rheumatoid factor

ANA positivity: 80% of patients with a positive ANA & JRA

ALL ANA positive, pauciarticular JRA patients should be routinely screened by an ophthalmologist every 3 months

Three (3) major forms of JRA

Pauciarticular disease – 40% with ANA

Polyarticular disease – 40%

Systemic Disease – 20%

Pauciarticular JRA – 2 types

Type I

Onset: early childhood

25% of all JRA

Girls:boys – 7:1

ANA Positive in 50% of pts

RF negative

Major complications is
iritidocyclitis

Type II

Onset: late childhood

15-20% of all JRA

Girls:boys – 1:10

ANA negative

RF negative

Hip and sacroiliac
involvement common

Ocular Symptoms

Insidious – 56% of children

Photophobia

Epiphora

Injection

Mild pain

Seronegative Spondylarthritis: Eye involvement – non granulomatous uveitis.

Conditions leading to Sacroiliitis

*Ulcerative Colitis

*Reiters Syndrome

*Ankylosing spondylitis

*Crohn's Disease

*Psoriasis

*High association with LFA-B27 and uveitis

Frequency of HLA-B27 in general population: 1.4-6%

Frequency of HLA-B27 in patients with acute uveitis: 50-60%

Excluding

Inflammatory Bowel Disease:

Frequency of uveitis greater in those patients with sacroiliitis and HLA-B27 positivity.

Terms Ulcerative Colitis & Crohn's Disease frequently confused in literature

Uveitis rare with ulcerative colitis but see it in 20% of patients with Crohn's Disease.

For board purposes uveitis occurs with each disease.

Ankylosing Spondylitis = Marie-Strumpell Disease

Symptoms: lower back pain & stiffness after inactivity.

May be infectious; triggers Klebsiella.

Frequency of HLA-B27 approaches 90%

Order Sacroiliac x-rays, NOT lumbosacral films

50% of patients asymptomatic but x-rays positive

5% of patients develop aortitis.

Rieter's Syndrome: frequently diagnosed retrospectively.

Triad of urethritis, conjunctivitis and non gonococcal polyarthritis.

May occur after dysentery without urethritis.

Possible triggers: chlamydia, shigella, salmonella & Yersinia

Occurs primarily in males and is bilateral

80% association with HLA-B27

Psoriatic Arthritis

The Uveitis is usually seen in those patients who also have arthritis.

Behcet's Syndrome – Type 3 Immune Response

Classic triad: acute hypopyon uveitis, aphthous, stomatitis, and genital ulcerations.

Common Japan, Near East, Mediterranean

Blindness results from retinal vasculitis.

25% of Patients develop stroke

Associated with HLA-B5 (subtype of MGA B-51)

Connective tissue disorders and eye involvement:

Systemic Lupus Erythematosus: Type 3 Immune Response

Retinal findings: Vasculitis, hemorrhage, cotton-wool spots.

More Uveitis Syndromes:

Fuchs' Heterochromic Iridocyclitis

Diffuse iris stromal atrophy with variable pigment layer causing heterochromia

Small, white keratic precipitates equally distributed over entire endothelium

Minimal aqueous cells and flare

Synechiae almost never form

Usually unilateral

Glaucoma and cataract common

Minimal symptoms

Glaucomatocyclitic crisis – Posner-Schlossman Syndrome

Unilateral, mild, acute iritis

Markedly increased intraocular pressure

Recurrences common

Granulomatous Uveitis:

Sarcoid – Type 4 Immune Response

Ocular inflammatory disease seen in 25-50% of patients with systemic sarcoid

More common in African Americans, Scandinavians

Multi-system disease primarily affect pulmonary function.

CNS, Liver and cutaneous involvement common

Ocular involvement:

Orbital disease: lacrimal gland, etc.

Anterior segment: "mutton-fat" keratic precipitates

Posterior segment: vitreous "snowballs", "candle wax drippings"
(Periplebitis)

Pathology:

Noncaseating granuloma: conjunctival biopsy

Schaumann's or lamellar bodies, asteroid bodies

Anergy: 90% of patients

Testing limited gallium scan of head/neck
serum lysozyme
biopsy

Tuberculosis – once very common

Diagnosis: high index of suspicion

Treatment: INH, Rifampin, Pyridoxine

Syphilis - :The Great Imitator”

May simulate all patterns of anterior and posterior uveitis

May be VDRL negative, FTS-ABS positive

High degree of suspicion in sexually active population

Up to 70% of males with acquired syphilis are homosexual

Treatment:

Benzathine penicillin, IM, weekly x3

Aqueous procaine penicillin IM with probenecid

Intravenous penicillin C.

Lens induced uveitis – Type 3 Immune Response

Phacoanaphylactic endophthalmitis (phacoantigenic uveitis)

Immune response to lens protein release seen after injury to lens capsule

or

ECCE.

Autologous lens protein may become autoantigenic after exposure to aqueous humor

A zonal granulomatous inflammation

Macrophages with lens material

Associated with sympathetic ophthalmia in 25% of cases

Treatment: steroids, cycloplegia, removal of lens material

Vogt-Koyanagi-Harada Syndrome

Seen in pigmented individuals with Indian heritage

Vitiligo, alopecia, poliosis, dysaesia, meningismus

Exudative retinal detachment with vitreous cells

“Sunset fundus” characteristic of past inflammation.

Treatment of Anterior Uveitis

Goal of therapy is to suppress inflammation and thereby minimize its destructive sequelae.

Specific treatment of causative condition

Example – treat syphilitic uveitis with penicillin.

Non-specific treatment: Immunosuppressives and Cycloplegics

Steroids: mode of administration and complications:

Topical:

Glaucoma

Posterior subcapsular opacities

Opportunistic corneal infections

Transient mydriasis and ptosis

Periocular:

- Growth retardation
- Central vasculature emboli
- Subconjunctival collection

Systemic:

- Peptic ulcer
- Osteoporosis
- Increased severity of diabetes and hypertension
- Mental changes
- Electrolyte imbalance
- Reactivation of infection (TB)
- Asceptic necrosis of femoral + humoral head

Cycloplegia – Goals of treatment

- Stop Pain of ciliary spasm induced by acute iritis
- Prevent posterior synechiae and resultant risk of papillary block glaucoma

Complications of Uveitis: secondary to disease process vs. medical treatment:

- Band keratopathy (JRA)
- Corneal edema
- Cataracts
- Glaucoma
- Synechia formation: anterior and posterior
- Hypotony
- Retinal edema and detachment
- Optic neuritis
- Neovascularization
- Myopia from ciliary spasm
- Hyperopia from macula edema

- Macular edema is the most common finding seen in posterior & intermediate uveitis.

- Macular edema is the most common cause of decreased vision associated with chronic inflammations.

Masquerade Syndromes – Anterior uveitis

- Retinoblastoma
- Leukemia
- Intraocular foreign body
- Malignant melanoma
- Juvenile xanthogranuloma can present with inflamed retina
- Retinal detachment
- Spillover syndromes: all causes of posterior uveitis
- Ocular ischemia