Uveitis

Classification

- Heterogeneous ocular diseases - inflammation of any component of uveal tract (iris, ciliary body, choroid).

Classifications

1. Intermediate uveitis (iris, ciliary body, choroid)

   - Presentation: Heterogeneous ocular disease
   - Incidence: 69% of cases
   - Features:
     1. Floaters
     2. Blurred vision

2. Posterior uveitis - choroiditis ± retinitis (chorioretinits, retinal vasculitis), optic neuritis.

   - Presentation: Heterogeneous ocular disease
   - Incidence: 5% of cases
   - Features:
     1. Floaters
     2. Blurred vision

Etiologic categories

1. Idiopathic (i.e. pars planitis) (69%)
2. Major granulomatous diseases - sarcoidosis (22%), tuberculosis, Lyme disease, lues, HLA-B27 syndromes, many articular syndromes (≈ 17%)
3. Trauma (5.7%)
4. Herpes simplex, herpes zoster disease (1.9-12.4%)
5. Iatrogenic (postoperative).

Complications

- Vision may be normal/ slightly decreased in chronic cases
- N.B. in some cases eye is normal-looking!

Acute iritis - iridocyclitis - cellular activity also involves retrobulbar vitreous

- Pain
- Photophobia
- Arthritis
- Night vision may be normal

Indirect ophtalmoscopy - cells in vitreous humor, cellular aggregates & condensations over pars plana (most frequently inferior - classic “snowbank” appearance), posterior synechiae may form.

Last updated: May 9, 2019

References:

- Online Journal of Ophthalmology
- Eye74 (1)
4.

**DIFFUSE UVEITIS (S. PANUVEITIS, ENDOPHTHALMITIS)** - involves all parts of eye.

**etiologies**

1. overwelling infections (e.g. infantile toxocariasis, postoperative bacterial endophthalmitis, severe toxoplasmosis).
2. dissemination of granulomatous uveitides.
3. **Reiter's syndrome**
   - peripheral granuloma involving pars plana (intermediate uveitis);
   - posterior granuloma near or involving optic nerve or macula;

**ETIOLOGIC CATEGORIES**

1. **Inflammatory** (primary autoimmune)
2. Infectious
3. Infiltrative (invasive neoplastic processes)
4. **Injurious**
5. **Iatrogenic** (surgery, inadvertent trauma, medication)
6. Inherited (metabolic / dystrophic disease)
7. **Ischemic**
8. **Idiopathic** (when all diagnostic means fail to reveal specific etiology)

**UVEITIS** can present with most challenging diagnostic dilemmas in all of ophthalmology! No standard laboratory evaluation exists for uveitis, except in screening for syphilis and sarcoidosis*, both of which can present in myriad of ways.

*or VDRL & FTA-ABS and ACE level & chest X-ray for all patients!

**TREATMENT**

**COMPLEX UVEITIC SYNDROMES**

**ANKLOKLING SPONDYLITIS**
- common cause of unilateral anterior uveitis.

**Reiter's syndrome**
- triad of conjunctivitis/uvetis, arthritis, urethritis.

**Juvenile rheumatoid arthritis** (particularly pauciarticular variant) - chronic bilateral iridocyclitis without pain, photophobia, conjunctival injection (“white iritis”).

**Behçet syndrome**
- rare in USA, common cause of uveitis in Middle and Far East - severe anterior uveitis with hypopyon, retinal vasculitis, optic nerve inflammation.
- clinical course is severe, with multiple recurrences.
- associated systemic manifestations - oral or genital aphthous ulcers; dermatitis (incl. erythema nodosum); thrombophlebitis; epidermitis. (see p. 1170-97)
- most patients eventually require immunosuppressive drugs (cyclosorpine, chlorambucil).

**Toxoplasmatis**
- posterior lesions (retinitis) + anterior segment involvement.

**Cytomegalovirus**
- posterior lesions (most common cause of retinitis in immunocompromised patients).

**Herpes zoster, herpes simplex**
- anterior uveitis ± dermatitis, keratitis, scleritis.
- rarely, rapidly progressing acute retinal necrosis, with dense vitreitis.

**Toxocariasis**
- one of most common causes of retinitis in childhood; classic forms of uveitis:
  - posterior granuloma near or involving optic nerve or macula;
  - peripheral granuloma involving pars plana (intermediate uveitis);
- severe and diffuse endophthalmitis.
- N.B. will worsen if given anticholinesterase drugs - patient will fare better with only anti-inflammatory treatment!

**Birdshot chorioretinopathy**
- idiopathic chronic bilateral, intermediate & posterior uveitis (moderate or severe vitritis and multiple areas of chorioiditis in distinctive radial or streak-like pattern).
- patients are in 5th to 7th decades of life.
- very strong association with HLA-A29.

**Ocular histoplasmosis**
- endemic to Mississippi-Ohio-Missouri River valleys) - multifocal choriditis (occasionally with macular hemorrhage from choroidal neovascularization).

**Syphilis**
- may cause any uveitis at any stage of disease.

**Ocular sarcoidosis**
- uveitis (anterior / intermediate / posterior) occurs in 10% patients with systemic sarcoidosis.

N.B. classically manifests as anterior granulomatous uveitis!
Vogt-Koyanagi-Harada syndrome (uveoencephalitis):

1) diffuse uveitis, commonly with exudative detachment of retina.
2) neurologic symptoms (occur early) - tinnitus, dysacusis, vertigo, headache, meningismus.
3) cutaneous findings (occur later) - patchy vitiligo, poliosis, alopecia.

- always affects more heavily pigmented persons.
- autoimmune reaction against choroidal melanocytes (moderately strong association with HLA-DR4).
- severe / prolonged attacks require immunosuppressants (e.g. cyclophosphamide, chlorambucil).

Symptomatic ophthalmia - bilateral granulomatous uveitis after penetrating trauma (0.5%) / surgery (< 0.1%) to one eye.

- autoimmune reaction against choroidal melanocytes (as in Vogt-Koyanagi-Harada syndrome).
- uveitis appears within 2-12 wk after injury.
- typically - floaters & decreased vision in sympathizing, noninjured eye.
- inflammation may be anterior / intermediate / posterior (choroiditis with overlying exudative retinal detachment is common).
- treatment - long-term corticosteroids + immunosuppressive drugs.

- prophylaxis - enucleation of severely injured, blind eye within 2 wk of vision loss!

Multiple yellowish ill defined lesions in choroid:

Endophthalmitis - acute, severe, diffuse uveitis.

- results from intracellular infection (vast majority after trauma or intraocular surgery).
- vitreous acts as superb medium for bacteria growth (in past, animal vitreous was used as culture medium!).
- severe pain and decreased vision, signs of intraocular inflammation (red eye, anterior chamber cells and flare, vitreitis, etc).
- medical emergency - infection may quickly involve orbit and CNS.

- treatment:
  1) broad-spectrum intracocular and systemic antibiotics (e.g. vancomycin, cefazolin).
  2) mydriatics.
  3) intraocular corticosteroids are used in selected cases.
  4) sometimes pars plana vitrectomy is indicated.

- N.B. visual prognosis is often poor, even with early and appropriate treatment!

- Endophthalmitis (after cataract extraction): mixed injection, corneal endothelial decompensation, Descemet's creases, flare in anterior chamber, necrosis of iris root (indicates intraoperative mechanical damage).

- Masquerade syndromes - conditions that mimic uveitis:
  1. Intraocular malignancy (primary / metastatic) in very young / elderly may be accompanied by significant intraocular inflammation (e.g. intraocular lymphoma).
  2. Retinitis pigmentosa
  3. Reactions to drugs (e.g. systemic sulfonamides, pamidronate, rifabutin).

- Vitreous opacification caused by metastasis of bronchial carcinoma. Vitreous biopsy reveals tumor cells.

BIBLIOGRAPHY for ch. “Ophthalmology” → follow this LINK >>