Uveitis

Classification of uveitis

Uveitis: is an inflammation of the uveal tract. . The four main classifications are:

(1) **anatomical**, (2) **clinical**, (3) **aetiological** and (4) **pathological**.

ANATOMICAL CLASSIFICATION

Anterior uveitis is subdivided into **iritis**, in which the inflammation predominantly affects the iris, and **iridocyclitis**, in which both the iris and the anterior part of the ciliary body (pars plicata) are equally involved.

Intermediate uveitis is characterized by predominant involvement of the posterior part of the ciliary body (pars plana) and the extreme periphery of the retina.

Posterior uveitis is characterized by inflammation located behind the posterior border of the vitreous base.

Panuveitis is characterized by involvement of the entire uveal tract.

Anterior uveitis is the most common type followed by intermediate, posterior and panuveitis.

CLINICAL CLASSIFICATION

Classified according to the mode of onset and duration, uveitis can be **acute** or **chronic**.

Acute uveitis usually has a sudden symptomatic onset and persists for 6 weeks or less. If the inflammation recurs following the initial attack, it is referred to as recurrent acute.

Chronic uveitis persists for months or years. Its onset is frequently insidious and may be asymptomatic, although occasionally acute or subacute exacerbations of inflammation may occur.

AETIOLOGICAL CLASSIFICATION

Exogenous uveitis is caused by either external injury to the uvea or invasion of micro-organisms or other agents from outside.

Endogenous uveitis is caused by micro organisms or other agents from within the patient. The following are the main types:
1. **Associated with systemic disease** (e.g. ankylosing spondylitis).
2. **Infections** with **bacteria** (e.g. tuberculosis), **fungi** (e.g. candidiasis), **viruses** (e.g. herpes zoster), **protozoa** (e.g. toxoplasmosis) or **roundworms** (e.g. toxocariasis).
3. **Idiopathic specific uveitis entities** (e.g. Fuchs' uveitis syndrome).

4. **Idiopathic non-specific uveitis entities** which do not fall into any of the above categories. They make up about 25% of all cases of uveitis.

PATHOLOGICAL CLASSIFICATION

Classified pathologically, uveitis may be **granulomatous** or **non-granulomatous**.

Clinical features of uveitis

ANTERIOR UVEITIS

Symptoms

The main symptoms of acute anterior uveitis are photophobia, pain, redness, decreased vision and lacrimation. In chronic anterior uveitis, however, the eye may be white and symptoms minimal, even in the presence of severe inflammation.

Signs

Injection in acute anterior uveitis is circumcorneal 'ciliary' and has a violaceous hue

Keratic precipitates (KP) are cellular deposits on the corneal endothelium. Their characteristics and distribution may give important clues as to the probable type of uveitis. KP most commonly form in the mid and inferior zones of the cornea.

1. **Small KP** are characteristic of herpes zoster and Fuchs' uveitis syndrome.
2. **Medium KP** occur in most types of acute and chronic anterior uveitis.
3. **Large KP** are usually of the 'mutton fat' variety and have a greasy waxy appearance . They typically occur in granulomatous uveitis.
4. Fresh KP tend to be white and round. With age, they shrink, fade and become pigmented.

Iris nodules are a feature of granulomatous inflammation.

1. **Koeppe** nodules are small and situated at the pupillary border).
2. **Busacca** nodules are larger but less common. They are located on the surface of the iris away from the pupil

Aqueous cells are a sign of active inflammation They should be graded according to the number observed in the oblique slit beam. The light intensity and magnification of the slitlamp should be maximal and the beam 3 mm long and 1 mm wide. The cells should be counted and graded from 0 to +4 as follows:

- 5-10 cells = +1
- 11-20 cells = +2
- 21-50 cells = +3
- >50 cells = +4.

Aqueous flare is the result of leakage of proteins into the aqueous humour through damaged iris blood vessels and not necessarily indicative of active inflammation. For this reason, the presence of a flare in the absence of cells is not an indication for treatment. Aqueous flare is graded using the same setting on the slitlamp as for counting cells. The beam should be passed obliquely to the plane of the iris in order to evaluate the degree of obscuration of iris details. The flare is graded from 0 to +4 as follows:

- faint - just detectable = +1
- moderate - iris details clear = +2
- marked - iris details hazy = +3
- intense - with severe fibrinous exudate = +4.

Posterior synechiae are adhesions between the anterior lens surface and the iris. They form with ease during an attack of acute anterior uveitis because the pupil is small. They may also form in eyes with moderate-to-severe chronic anterior uveitis. Posterior synechiae extending for 360° (seclusio pupillae) prevent the passage of aqueous humour from the posterior to the anterior chamber, giving rise to a forward bowing of the peripheral iris (iris bombé), which may lead to elevation of intraocular pressure secondary to closure of the angle by the peripheral iris.

Anterior vitreous cells should be compared in density with those in the aqueous. In iritis, aqueous cells far exceed the number of vitreous cells, whereas in iridocyclitis the cells are distributed equally between the two compartments.

INTERMEDIATE UVEITIS

The symptoms are usually floaters, although occasionally the patient presents with impairment of visual acuity caused by chronic cystoid macular oedema.

Signs are cellular infiltration of the vitreous (vitritis) with few, if any, cells in the anterior chamber and no focal inflammatory lesion in the fundus.

POSTERIOR UVEITIS

Symptoms

The two main symptoms of posterior segment inflammation are floaters and impaired vision. A patient with a peripheral inflammatory lesion will complain of seeing floaters and may have only minimal blurring of vision. On the other hand, active choroiditis involving the fovea will primarily cause loss of central vision, and the patient may not notice the presence of floaters.

Signs

Vitreous changes include cells, flare, opacities and, frequently, posterior vitreous detachment. In some cases the posterior hyaloid face is covered by inflammatory precipitates comparable to KP. Coarse opacities are usually the result of severe tissue destruction.

’Snowball’ or ’cotton-ball’ opacities are characteristic of pars planitis, although they may also occur in candidiasis and sarcoidosis.

Choroiditis is characterized by yellow or greyish patches with reasonably well-demarcated borders

Retinitis gives the retina a white cloudy appearance. Because the outline of the inflammatory focus is indistinct, exact demarcation between healthy and inflamed retina may be difficult to discern.

Vasculitis is inflammation of the retinal blood vessels. The retinal veins (periphlebitis) are most frequently involved, although in some cases the arterioles (periarteritis) may be affected.

Treatment of uveitis

Aims of therapy

The aims of treating uveitis are:
1. **To prevent vision-threatening complications**.
2. **To relieve the patient’s discomfort**.
3. **If possible, to treat the underlying cause**.

The four groups of drugs currently used in the treatment of uveitis are: (1) **mydiatics**, (2) **steroids**, (3) **cytotoxic drugs** and (4) **cyclosporin**. Patients with uveitis caused by infections should be treated with the appropriate antimicrobial or antiviral agent.

Behçet’s disease

SYSTEMIC FEATURES

Behçet’s disease is an idiopathic multisystem disorder which typically affects young men from the eastern Mediterranean region and Japan, but is rare in western Europe and America, The disease is associated with an increased prevalence of HLA-B5. The basic lesion is an obliterative vasculitis probably caused by abnormal circulating immune complexes.

Presentation is usually in the third and fourth decades with recurrent oral aphthous ulceration. As there are no special confirmatory tests, the diagnosis requires the presence of **oral ulceration** in association with two of the following: **recurrent genital ulceration, skin lesions, eye involvement** and **a positive pathergy test**.

Oral ulceration is a universal finding and a very common presenting feature. Aphthous ulcers are painful and shallow with a yellowish necrotic base. They are recurrent and tend to occur in crops, which may involve the tongue, gums, lips and buccal mucosa

Genital ulceration is present in about 90% of patients and is more apparent and troublesome in men than in women

Skin lesions include erythema nodosum, pustules and ulceration. A papule developing at the site of a skin puncture (pathergy test) is characteristic.

Other features include thrombophlebitis, arthropathy, gastrointestinal lesions, CNS involvement and cardiovascular lesions.

OCULAR FEATURES

About 70% of patients with Behçet's disease develop recurrent, bilateral, non-granulomatous, intraocular inflammation. In any individual patients, either anterior or posterior segment involvement can predominate. In patients with posterior segment involvement, the long term visual prognosis is poor.

Acute recurrent iridocyclitis which may be associated with a transient hypopyon is common. Initially, it responds well to topical steroids but it may subsequently become chronic and lead to phthisis bulbi.

Posterior segment involvement includes the following:
1. **Diffuse vascular leakage** throughout the fundus is the most common and persistent finding.).
2. **Periphlebitis**, which may result in venous occlusion
3. **Retinitis** characterized by white necrotic infiltrates of the inner retina,
4. **Vitritis**, which may be severe and persistent, is universal in eyes with uveitis.

TREATMENT OF POSTERIOR UVEITIS

1. **Systemic steroids** in high doses are usually effective initially in controlling posterior segment inflammation. Unfortunately, the lesions often subsequently become steroid resistant and require alternative therapy.
2. **Chlorambucil** is initially effective in about 75% of cases.
3. **Cyclosporin** is a potent immunomodulator affecting both the cellular and humoral arms of the immune response. It may be beneficial for the acute exacerbations of both eye and mucocutaneous lesions.
4. **Plasma exchange** may be useful in some cases.