Uveitis

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Uveitis is inflammation of the uveal tract.

Classification

A- Anatomical classification.

1-Anterior uveitis may be subdivided into:

- **Iritis** in which the inflammation primarily involves the iris.
- **Iridocyclitis** in which both the iris and the pars plicata of the ciliary body are involved.

2-Intermediate uveitis is defined as inflammation predominantly involving the pars plana, the peripheral retina and the vitreous.

3-Posterior uveitis involves the fundus posterior to the vitreous base.

- Retinitis with the primary focus in the retina.
- Choroiditis with the primary focus in the choroid.
- Vasculitis which may involve veins, arteries or both.

4-Panuveitis implies involvement of the entire uveal tract without a predominant site of inflammation.

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

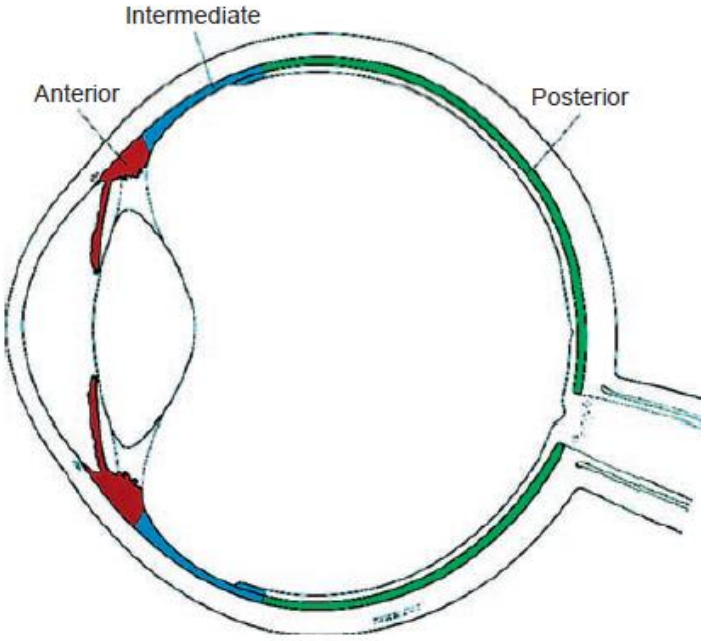

Fig. 11.1 Anatomical classification of uveitis

B-Clinical classification.

According to the mode of onset and duration

1- **Acute** uveitis ; characterized by sudden onset and limited duration.

2- **Chronic** uveitis describes persistent duration, with relapse less than 3 months after discontinuation of treatment .

C- Pathological classification.

1-Granulamatous uveitis.

2- Non granulamatous uveitis.

Clinical features

- **Anterior uveitis**

Symptoms in **Acute Anterior uveitis** (AAU); consist of the rapid onset of pain, photophobia, redness and watery discharge, sometimes preceded by mild ocular discomfort for a few days. Blurring of vision is related to severity. **Chronic Anterior uveitis** (CAU); may be of insidious or acute onset, and can be asymptomatic until the development of complications such as cataract.

1- **Visual acuity** is variably impaired depending on the severity of inflammation and the presence of complications. It is frequently only mildly reduced in AAU.

2- **'Ciliary injection'** (perilimbal injection) is circumcorneal conjunctival hyperaemia with a violaceous (purplish) hue due to involvement of deeper blood vessels and is typically seen in anterior uveitis of acute onset.

3- **Miosis** due to pupillary sphincter spasm , predisposes to the formation of posterior synechiae.

4- **Anterior chamber cells** are a dependable indicator of inflammatory activity. Grading is performed by estimating the number of cells in a 1 mm by 1 mm slit beam field, employing adequate light intensity and magnification.

Grade	Cells in field
0	<1
0.5+	1–5
1+	6–15
2+	16–25
3+	26–50
4+	>50

5-Aqueous flare is haziness of the normally clear fluid in the anterior chamber, reflecting the presence of protein due to breakdown of the blood–aqueous barrier. Flare may be graded clinically using a slit lamp to assess the degree of interference with visualization of iris and lens.

Grade	Description
0	None
1+	Faint
2+	Moderate (iris and lens details clear)
3+	Marked (iris and lens details hazy)
4+	Intense (fibrin or plastic aqueous)

6-Keratic precipitates (KP) are deposits on the corneal endothelium composed of inflammatory cells such as lymphocytes, plasma cells and macrophages. They are usually concentrated in the mid and inferior zone of the cornea, Their characteristics indicate the probable type of uveitis: typically **smaller** in the non-granulomatous inflammation typical of AAU, and **medium to large** in (classically chronic) granulomatous inflammation in which cell types may include epithelioid and multinucleated cells. Large greasy-appearing granulomatous KP are said to have a **'mutton fat' appearance.**

7- Hypopyon refers to a whitish purulent exudate composed of myriad inflammatory cells in the inferior part of the anterior chamber (AC), forming a horizontal level under the influence of gravity. Hypopyon is common in **HLA-B27**-associated AAU, when a high fibrin content makes it **immobile** and slow to absorb. In patients with **Behçet disease** the hypopyon contains minimal fibrin and so characteristically **shifts** according to the patient's head position.

8-Fibrinous exudate in the anterior chamber is common in severe AAU, and as with hypopyon is often seen with HLA-B27-related inflammation.

9- Posterior synechiae (PS) are inflammatory adhesions between the pupil margin and the anterior lens capsule. They can develop rapidly, and to prevent their formation initial prophylaxis with a mydriatic agent. Once established, every attempt must be made to break PS before they become permanent.

10- Iris nodules: **Koeppe nodules** are located on the pupillary margin. They can occur in both granulomatous and non-granulomatous anterior uveitis. **Busacca nodules** involve the iris stroma and are a feature of granulomatous uveitis.

11- **Iris atrophy & Iris neovascularization (rubeosis iridis);** can occur, particularly in chronic inflammation.

INTERMEDIATE UVEITIS

Clinical features

Symptoms. Presentation is with the insidious onset of blurred vision, often accompanied by vitreous floaters; there is usually no pain or redness.

Signs

1-Visual acuity is variably affected depending on inflammatory activity and complications, particularly cystoid macular odema (CMO).

2- Anterior uveitis. (anterior chamber cells, Keratic precipitates(KPs), Posterior synechiae (PS).

3- Vitreous. Vitreous **cells** with anterior predominance are universal, with vitreous **condensation** and haze in more severe cases. **Snowballs** are whitish focal collections of inflammatory cells and exudate, usually most numerous in the inferior vitreous.

4- Posterior segment.

- Peripheral periphlebitis.
- Snowbanking is characterized by a grey-white fibrovascular plaque which may occur in all quadrants, but is most frequently inferior.
- Others ; cystoid macular odema (CMO),Neovascularization and Optic disc swelling.

POSTERIOR UVEITIS

Clinical features

Symptoms; are mainly floaters and impaired vision.

Signs

- **Retinitis** may be focal (solitary), multifocal, geographic or diffuse. Active lesions are characterized by whitish retinal opacities with indistinct borders due to surrounding oedema .As the lesion resolves, the borders become better defined.

- **Choroiditis** may also be focal, multifocal, geographic or diffuse. It does not usually induce vitritis in the absence of concomitant retinal involvement. Active choroiditis is characterized by a round, yellow nodule.
- **Vasculitis** may occur as a primary condition or as a secondary adjacent to a focus of retinitis. Both arteries (periarteritis) and veins (periphlebitis) may be affected although venous involvement is more common.

AETIOLOY OF UVEITIS

- **Idiopathic; non specific** about 25% of all cases of uveitis.
- **Idiopathic specific uveitis entities** (e.g. Pars planitis, Fuchs uveitis syndrome).
- **Infectious;** viral (herpetic uveitis),bacterial (TB), protozoa (toxoplasmosis) or roundworm (toxocariasis).
- **Non-infectious;** HLA-B27 positivity (e.g.Ankylosing spondylitis), Juvenile idiopathic arthritis Sarcoidosis, Behçet diseas and Multiple sclerosis & external trauma.

Approach to the patient with uveitis

1- History

- Symptoms of uveitis (redness, pain, photophobia, blurring of vision, etc.)
- Systemic review.

2- Examination

- Ocular examination for the signs of ; (Anterior uveitis, Intermediate uveitis, Posterior uveitis).
- Systemic examination (heart, skin, joints, etc.)

3- Investigations; are directed according to the history & clinical examination.

Treatment

The aims of treating uveitis are

1- Prevention of vision threatening complications.

2- Relieve patient discomfort. 3-If possible ,to treat the underlying cause.

General principles

Treatment of immune-mediated uveitis involves predominantly the use of anti-inflammatory and immunosuppressive agents. Antibiotic or antiviral therapy for infectious diseases. It is important to keep in mind that drugs used to treat uveitis have potential side-effects, and this should always be weighed against the decision to treat. Also, it must be emphasized that the use of systemic therapy should be carried out in conjunction with a physician who is competent to deal with complications associated with both the underlying disease and the therapy.

1-Mydriatics ;(Tropicamide, Cyclopentolate ,Atropine). To promote comfort, prevent formation of posterior synechiae and break down recently formed posterior synechiae.

2- Steroids; (Topical. Periocular steroid injection, Intraocular and Systemic).

3- Antimetabolites. (Azathioprine, Methotrexate, Mycophenolate mofetil).

4- Calcineurin inhibitors. (Ciclosporin &Tacrolimus).

5- Biological blockers. (Interleukin receptor antagonists &Tumour necrosis factor alpha antagonists).

Miscellaneous uveitis

Uveitis in spondyloarthropathies (HLA-B27).

The AAU associated with HLA-B27 is typically unilateral, severe, recurrent and associated with a higher incidence of posterior synechiae. A fibrinous exudate in the anterior chamber is common.

Toxoplasma retinitis

Toxoplasmosis is the **most frequent cause** of infectious retinitis in immunocompetent individuals. Reactivation at previously inactive cyst-containing scars is the rule in the immunocompetent, although a small minority may represent new infection. Most quiescent lesions will have been acquired postnatally. Recurrent episodes of inflammation are common and occur when the cysts rupture and release hundreds of tachyzoites into normal retinal cells.

Clinical features

The diagnosis of toxoplasma retinitis is based on a compatible fundus lesion and positive serology for toxoplasma antibodies

Presentation is with unilateral sudden onset of floaters, visual loss and photophobia.

Signs;

1- Anterior uveitis.

2- A single inflammatory focus of white retinitis or retinochoroiditis associated with a pigmented scar ('satellite lesion') is **typical**.

3-The focus not associated with an old scar, and multiple lesions are relatively uncommon in the immunocompetent but occur more frequently in the immunocompromised..

4-**Vitritis** may be severe and impair fundus visualization. **'Headlight in the fog'** is the classic description of a white retinal inflammatory nidus viewed through vitritis.

- **Healing** in immunocompetent hosts usually occurs spontaneously within 6–8 weeks, although vitreous opacities take longer to clear. The inflammatory focus is replaced by a sharply demarcated atrophic scar that develops a pigmented border.
- **Treatment**

1-Prednisolone is given initially and tapered according to clinical response, but should always be used in conjunction with a specific anti-*Toxoplasma* agent, most frequently pyrimethamine combined with sulfadiazine ('classic' or 'triple' therapy). Systemic steroids should be avoided or used with extreme caution in immunocompromised patients.

2- Pyrimethamine is a folic acid antagonist. in combination with **oral folinic** (not folic) acid 5 mg three times a week to retard thrombocytopenia, leukopenia and folate deficiency.

3- Sulfadiazine is usually given in combination with pyrimethamine. Side effects of sulfonamides include renal stones, allergic reactions and Stevens–Johnson syndrome.

4-Other systemic options include clindamycin, Co-trimoxazole , azithromycin and clarithromycin.

Behçet disease

Behçet disease is an idiopathic, multisystem syndrome characterized by recurrent aphthous oral ulcers, genital ulceration and uveitis. Vasculitis is a key pathogenetic component and may involve small, medium and large veins and arteries. The disease typically affects patients from Turkey, the Middle and Far East with a lower prevalence in Europe and North America. Behçet disease probably has an autoimmune basis, and may be precipitated by exposure to an infectious agent with

subsequent cross-reaction. It is strongly associated with **HLA-B51**. The peak age of onset of BS is in the 3rd decade,; males are affected more frequently than females.

Diagnostic criteria

1- Recurrent oral ulcerations that have recurred at least three times in a 12-month period.

2- Plus at least two of the following:

1-Recurrent genital ulceration.

2-Ocular inflammation.

3-Skin lesions include erythema nodosum, folliculitis, acneiform nodules or papulopustular lesions

4-Positive pathergy test, which is characterized by the formation of a pustule after 24–48 hours at the site of a sterile needle prick .

Additional features

Major vascular complications, arthritis, gastrointestinal ulceration ,CNS, hepatic and renal lesions.

Ocular features

ocular inflammation is the presenting manifestation in about 10% of cases. Ocular disease is usually bilateral and typically presents during the 3rd–4th decades.

1- AAU, which may be simultaneously bilateral and frequently associated with a transient mobile hypopyon in a relatively white eye ('cold hypopyon). It usually responds well to topical steroids.

2-Vitritis, which may be severe and persistent, is universal in eyes with active disease.

3-Retinitis

4-Retinal vasculitis. may involve both veins and arteries and result in occlusion

5-End-stage disease is characterized by optic atrophy, vascular occlusion.

Treatment posterior uveitis

Immunosuppressants are the mainstay of treatment.

1-Systemic steroids and azathioprine in combination are recommended for the initial management of posterior uveitis.

2- Ciclosporin in combination with azathioprine and systemic steroids.

3- Biological blockers (Infliximab) should be considered early for vision-threatening Behçet disease.

4- Subcutaneous Interferon-alfa. with or without steroids**.** for severe disease.