UVEA / UVEITIS

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Uvea = middle layer (coat) of the globe
= vascular pigmented layer

- A. iris
- B. ciliary body
- C. choroid

- Position: the uveal tract lies between the sclera and retina.
- Arterial supply to the uveal tract is provided *by the ophthalmic artery.*

Iris

- It is a coloured, circular diaphragm with a central aperture — the pupil, measuring about 3,5 - 4 mm. It regulates the amount of light rays reaching the retina.

- Iris consists of 2 layers:
- **Anterior stromal layer**
- **Posterior pigmented epithelial layer**

  (protects the eye against excessive incident light)

- **two muscles**
  - **sphincter pupilae m.** - circular fibres – miosis
  - **dilatator pupilae m.** – radial fibres – mydriasis

IRIS

- Supplies with major and minor vascular (arterial) circle (not visible)
- Colour of the iris – depends on the amount of pigment melanin (developing – to 6th month of life)
- At the periphery, the iris is attached to the ciliary body.

Ciliary body

- **Is continuation of the iris -** *extends from the root of the iris to the ora serrata, where it joins the choroid.*
- **It consists of anterior pars plicata and the posterior pars plana**

FUNCTION :

1. **ciliary muscle (= accomodative muscle)** is responsible for accommodation.
2. **production of the aqueous humor**

   On the surface of ciliary body – approx . 60- 80 ciliary processes (pars plicata part of ciliary body)

   Nonpigmented layer of the epithelium covering the ciliary processes produces the aqueous humor
3. **Between ciliary processes - lens zonules** (suspensory ligaments) are attached

   Suspensory ligaments are attached to the ciliary processes and the equator of the lens.

ZONULAR FIBRES (Zonula ciliaris Zinii)

- = ”Suspensory ligaments ”
- Series of very fine transparent fibres which run from ciliary processes
- Attached to the lens capsule around equator
- Holds the lens in the right position
- Assist action of ciliary muscle during accommodation
- It is composed of glycoproteins and mucopolysaccharides

Accommodation = the ability of the eye to change its focus from distant to near objects (and vice versa). This process is achieved by the lens changing its shape.

Changes during accommodation:

A) contraction of ciliary muscles;

B) approximation of ciliary muscles to lens;

C) relaxation of suspensory ligament;

D) increased curvature of anterior surface of lens

Choroid – middle layer of the eyeball

- Contains vessels and lot of pigmented cells
- It is a dark , highly vascular layer situated in between the sclera and retina.
- Function : as „ lymphatic nodule“ , **supplies nourishment to the outer layers of the retina**
- The outer layers of retina are dependent for their nutrition
  (feeds pigmented epithelium of the retina and a layer of rods and cones)

- The inflammation of choroid always involves the retina.

- The blood flow through the choroid is the highest in the entire body

- 3 layers: suprachoroid / blood vessel layer / Bruch`s membrane

INFLAMMATION OF THE UVEAL TRACT (UVEITIS)

Uveitis – definition

- **The term uveitis strictly means inflammation of the uveal tissue only.**

- **However, there is always associated inflammation of the adjacent structures such as retina, vitreous, sclera or cornea.**

Uveitis- classification

- Anterior (iritis, iridocyclitis)
  - the inflammation of the iris (iritis) and pars plicata of the ciliary body (cyclitis) or iridocyclitis.
- Intermediate (pars planitis)
  - the inflammation of the pars plana part of the ciliary body.
- Posterior
  - the inflammation of the choroid (choroiditis or combination with retina as chorioretinitis - focalis, diseminnata , nekrotisans
- Panuveitis - all uveal parts

- Endophtalmitis - inflammation of the internal structures of the eye, i.e. choroid,retina and vitreous.

- Panophtalmitis- all intraocular structures : purulent inflammation of all the structures of the eye. There is inflammation of all the three coats of the eye and Tenon's capsule as well.

Types of inflamation

- **Granulomatous** - infective in nature , chronic inflammation, limited granuloma formation
  (less virulent, usually endogenous)

- **Non – granulomatous** -it is usually due to allergic or immune related reaction.
- acute onset and shorter duration
- acute diffuse damage - response to virulent pathogens, hyperergic states with a strong inflammatory and exudative response

Etiology

- In most cases, uveitis is not due to direct infection. It is usually due to allergy or hypersensitivity reaction to an infectious agent.
- **Hypersensitivity reaction**—It occurs due to hypersensitivity reaction to autologous tissue components (autoimmune reaction). Therefore uveitis occurs commonly in association with: rheumatoid arthritis, systemic lupus erythematosus, sarcoidosis, ankylosing spondylitis, Reiter's disease, Behcet's syndrome.

Classification- CAUSES

- **I.Endogenous**

  Organisms lodged in some other organ of the body reach the eye through the bloodstream.

- Infections -viral, bacterial, parasitic ,fungal disease
- Systemic inflammatory disease (reumatoid disease)
- Idiopathic nonspecific and specific (allergic, autoimmune background)

- **II.Exogenous** - after trauma or after surgery

It occurs due to a perforating wound or corneal ulcer. It causes acute purulent iridocyclitis and sometimes panophthalmitis.

General mechanism of ocular inflammation

The following mechanisms are involved in inflammation of the uveal tissue

- ***Vessel dilatation*** → deep ciliary (mixed) injection
- ***Vascular permeability*** → aqueous ***flare*** (flare in anterior chamber = cell movement) , **tyndalisation**
- ***Migration of cells*** → keratic precipitates (cell precipitates on the corneal endothelium)

→ exudate in anterior chamber

→ hypopyon (pus level on AC bottom)

Anterior uveitis (iridocyclitis)

SYMPTOMS:

- Eye-dull pain- worse at acomodation
- sensitivity to touch, increased tearing, photophobia
  (Photophobia is due to pain induced by pupillary constriction and ciliary spasm because of inflammation!)

  (without any mucopurulent discharge)
- red eye = ciliary injection = hyperaemia around the limbus due to the dilation of anterior ciliary vessels
- decreased vision - due to exudate in AC (anterior chamber)

Anterior uveitis (iridocyclitis)

SIGNS :

- Iris –decoloration or nodule formation in the iris tissue
- inflammatory exudate - Ly, Le, fibrin

(The exudate tends to stick to the damaged endothelium in the lower part of cornea in a triangular pattern due to the convection currents in anterior chamber and effect of gravity.)

- Tyndalisation - flare in anterior chamber - cell movement
- cell precipitates on the corneal endothelium
- hypopyon-pus level on AC bottom , serofibrinous
- posterior synechiae- adhesions between the iris and lens , pupil is lobed

Anterior uveitis (iridocyclitis)

- **Diagnosis:**
- subjective compliants
- biomicroscopy (at the slit lamp)-objective findings
- laboratory exams - ASLO, LATEX, CRP, Brucela, Leptospira, BWR, HSV, HIV ...
- **foci** -ORL, urology, gynecology, lung (tuberculin test), dentist
- immunological examination

Therapy of the anterior uveitis (iridocyclitis)

mydriatics (barrier formation, break adhesions between iris end anterior capsule of the lens , reduce pain, swelling).....ATROPINE drops

Cortiocosteroids – anti inflammatory, anti-allergic,anti –fibrotic activity

anti-inflammatory drugs - drops and pills (Ibuprofen)

specific anti-inflammatory therapy (antibiotics, antivirals, ...) immunomodulation, immunosuppressants

Non-steroidal anti-inflammatory (NSAIDs) :drops,pills, etc. indomethacin, diclofenac. These are safer as prolonged use of steroids which may produce complications : steroid glaucoma by reducing outflow facility, cataract and secondary infection with bacteria or fungi.

Corticosteroids

- These are anti-inflammatory in action.
- They are very useful in controlling inflammation in the acute phase.
- Due to their anti-allergic and anti-fibrotic activity they reduce fibrosis and thus prevent disorganisation and destruction of tissues.
- **Topical**—eyedrops and eye ointment
- **Subconjunctival injection**
- **Periocular injection of depot steroids**
- **Systemic steroids**
- **intravitreal injections**

Intermediate uveitis

(CHRONIC POSTERIOR CYCLITIS OR PARS PLANITIS)

It affects the pars plana of the ciliary body and often the peripheral retina and underlying choroid.

- Idiopatic, chronic , relapsing inflammatory disease

- or assocciated with systemic diseases

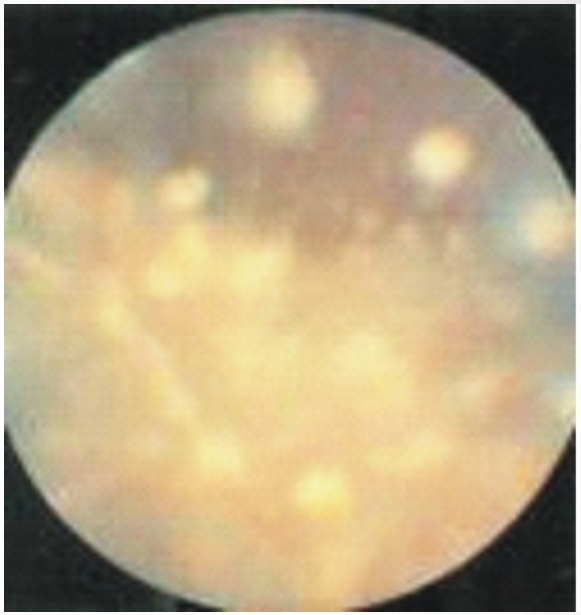
- *multiple sclerosis (MS) , sarcoidosis, Lyme disease, reumatoid arthritis*

- It is common in children and young adults

- **VITREUS is the major site of the inflammation**

Intermediate uveitis

(CHRONIC POSTERIOR CYCLITIS OR PARS PLANITIS)



SYMPTOMS : Subjectiv:

- flying flies (seeing floaters)
- deterioration of central visual acuity
- blurred vision

SIGNS: Objectiv:

- cell infiltration , condensation in vitreus , vitritis
- inflammatory cells in the anterior chamber (+ or -)
- "snowballs, snowbanking,i.e.grey-white plaques involving the inferior pars plana
- cotton ball 'bearing whitish-based pars planitis

Intermediary uveitis – THERAPY

- corticosteroids - parabulbar (periocular) injection
  - therapeutic concentration behind the lens
  - prolonged effect
- systemic corticosteroids

  *oral prednisone (*start with large dose : Prednison 1-2 mg/kg/day – maintained until clinical effect is seen ,reduce slow)

  or

  i.v. methylprednisolone 1g /day 2-3 days
- immunosuppressants
- intravitreal steroids
- vitreoretinal surgery in severe cases

Posterior uveitis

- choroiditis or chorioretinitis

Symptoms:

- Painless - no sensitive innervation
- Calm eye - often unrecognized
- Metamorphopsia—straight line appears wavy due to oedema of the retina.
- Decrease in visual acuity- due to retinal lesions and opacities in the vitreous (floaters) and especially when macular involvement
- Sensation of flying flies, cobwebs, blurred vision, vitreous exudate (Black spots are seen floating in the vitreous (vitritis).

Posterior uveitis-chorioretinitis

SIGNS:

- In early stage one or more yellowish areas (yellowish gray exudative changes)with ill-defined edges are seen deep under the retinal vessels. This appearance is due to infiltration of the choroid and presence of exudates which hide the choroidal vessels.
- There may be sheathing of retinal vessels.
- Complications: macular edema , optic disc edema
- Usually chronic disease
- Healing stage - scars - fibrous tissue (whitish scar-scleral shines), pigment clumps

(yellow lesions become white due to fibrosis and the lesions are surrounded by black pigments)

Posterior uveitis

Diagnosis:

- Subjective problems , symptoms
- objective findings(panfundoscopy)
- visual field test
- fluoroangiography, OCT (optical coherence tomography)
- sampling of vitreous material
- immunological exams

Posterior uveitis- therapy

- General treatment - according to etiology, antibiotics, antivirals ..
- Specific treatment is required for causative organism such as: toxoplamosis, toxocariasis , tuberculosis, syphilis, etc.
- Immunosuppression
- Nonspecific anti-inflammatory therapy, Wobenzym
- Corticosteroids : systemic, parabulbar , intravitreal steroids
- vitreoretinal surgery (pars plana vitrectomy) , application of antibiotics into vitreous

Clinical types of uveitis

Sarcoidosis

- chronic multisystem granulomatous disease of unknown etiology
- predominantly affects lungs and intrathoracic lymph nodes, but any organ can by involved
- involvement of the eyes and adnexa occurs in 25-80%

- ***Ocular manifestation of sarcoidosis***

- **Adnexa**l involvement:

Orbital lacrimal gland granuloma
Extraocular muscles granuloma
Lid granuloma
Conjunctiva granuloma
Conjunctivitis

- **Episcleritis, scleritis**
- **Keratitis**
- **Uveitis granulomatous or non-granulomatous**

Anterior uveitis
Intermediate uveitis
Posterior uveitis
Panuveitis

- **Optic nerve** involvement

Papilitis
Optic disc granuloma
Papilledema

Toxoplasmosis = protozoan infection

- common cause of posterior uveitis
- Toxoplasma gondii

- spread by haematogenous dissemination – to the muscles, brain, ,choroid and retina
- clinical picture – necrotizing chorioretinitis
  – often self-limited, progressively resolves, leaving scar(s)

- recurrences – unpredictable

- in immunosuppressive patient – multifocal, progressive chorioretinitis

Toxacariasis

- Toxocara canis, cati
- usually – in children
- the soil of parks and playgrounds is commonly contaminated with the eggs
- larvae migrate through the intestine wall to the blood, encystes in various tissues (eye, brain, liver,..)
- ocular finding – granuloma
- death of the larva leads to severe intraocular inflamation

Fungal uveitis

- uncommon
- causative organism – Candida, Aspergillus, cryptococcus, fusarium
- progresive intraocular inflamation
- more often in : immunocompromised patients or in organ transplant patient

Th : Amphotericin-B

Surgical : pars plana vitrectomy

Sympathetic ophthalmia

- **Specific bilateral inflammation** of the uveal tract due to chronic irritation of one eye, caused by a perforating wound to the eye or (very rare) after intraocular surgery, produces transferred **uveitis in the fellow eye**

- Tissues in the injured eye (uveal tract , lens, and retina) act as antigens and provoke an autoimmune disorder in the unaffected eye.

Etiology of sympathetic ophthalmia

- **After trauma, penetrating injury** (sympathetic eye) –

- immune response to antigens of injured uvea: iris, ciliary body or choroid
- autoimmune response to proteins of damaged tissue

---- antibodies attack uvea of the other (second) eye

- on the second (sympathizing) eye – fibrovascular chronic inflammation of the iris and ciliary body

- Iridocyclitis or chorioretinitis

Sympathetic ophthalmia

(prevalence 0,06-0,01%)

Clinical signs include :

- combined injections
- cells and protein in the anterior chamber and vitreous body,
- papillary and retinal edema, granulomatous inflammation of the choroid.
- The disorder has a chronic clinical course and may involve severe **complications** such as secondary glaucoma, secondary cataract, retinal detachment, and shrinkage of the eyeball.
- Sympathetic ophthalmia can lead **to blindness** in particularly severe cases

Sympathetic ophthalmia

Therapy :

- high doses of corticosteroids i.v., p.o. , topical
- local mydriatics
- Immunosuppression
- antibiotics
- surgery (enucleation sympathetic eye) - the injured eye, which is usually blind, must be in severe cases enucleated to eliminate the antigen and to save 2-nd eye

Severe prognosis ….. without treatment……blindness!

Endophthalmitis

(inflammation of the internal structures of the eye, i.e. choroid, retina and vitreous)

- ***Exogenous:***
- after trauma, penetrating injuries, intraocular foreign body , perforation of suppurative corneal ulcer
- after intraocular surgery (sources :bacterial flora of the eyelids, conjunctiva and lacrimal passage)
- ***Endogenous :***
- Systemic infection may cause metastatic infection(septicemboli),e.g.AIDS, viral fever, septicemia. It may occur in immunodeficient host and uncontrolled diabetic patients.

(DM gangrene, focuses in the body , lowered immunity – purulent microbes - infammation of the middle and inner layer of the eye)

Endophthalmitis – clinical features

- There is sudden onset with severe pain and redness in the eye in acute cases.
- Visual loss
- Lid oedema,chemosis and corneal haze
- Fibrinous exudate or hypopyon
- Vitritis (haze in the vitreous from inflammatory cells)
- Yellowish reflex seen behind the lens
- Inability to visualize the fundus