433 Teams

OPHTHALMOLOGY

11

Ocular manifestations of systemic diseases

Color index:

432 Team – **Important** – **433 Notes** – Not important

ophthalmology433team@gmail.com

جامعة الملك سعود
King Saud University

Diabetic retinopathy:

Introduction:

- The most common disease that involved the eye (particularly Retina) is DIABETES.
- Almost in all people with type 1 diabetes mellitus. 'due to early onset'
- Diabetic retinopathy is **most common microvascular complication of DM**
- The only means of preventing blindness due to diabetic retinopathy is through **screening and early treatment.**

Risk Factors:

1- **Duration of diabetes:** The longer the duration of diabetes, the more risk of DR
2- **Poor control of diabetes**: Raised HbA1c is associated with an increased risk of PDR.
3- **Pregnancy**
4- **Hypertension:** should be rigorously controlled (<140/80 mmHg).
5- **Nephropathy**: Renal transplantation may improve DR
6- **Others:** Hyperlipidemia, smoking, cataract surgery, obesity and anemia.

Pathogenesis:

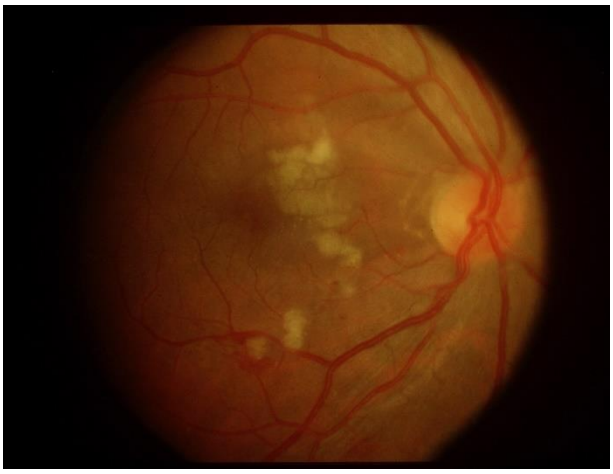
- Hyperglycemia lead to dysfunction of endothelial cells lining retinal capillaries and the loss of the inner Blood-Retina-barrier lead to edema
- Occlusion of vessels produce VGEF and lead to neovascularization

Ocular manifestation:

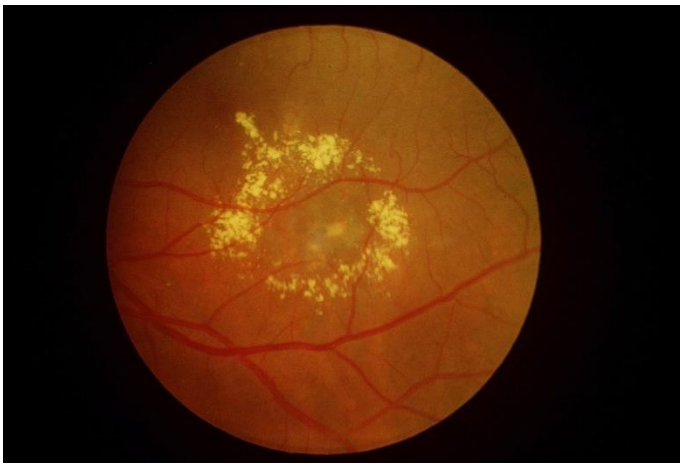
1. **Iris:** Rubeosis iridis (neovascularization in iris) **"May lead to Neovascular glaucoma"**
2. **Lens:** Cataract
3. **Uvea:** Iridocyclitis (another term of uveitis)
4. **Retinopathy: Most common one**
5. **Optic neuropathy:** diabetics have more than 25 times chance to get blindness
6. 3rd, 4th, 6th nerves palsies

RETINAL CHANGES:

1. Hard exudates
2. Macular edema
3. Microaneurysmus **'blots and dots'**
4. Soft exudates **'cotton-wool spots'**
5. Venous changes: beading vs looping **(The most reliable signs of retinal ischemia in diabetic retinopathy).**
6. Hemorrhage if neovascularization results in weak vessels and easy to break.
7. Neovascularization



Cotton wall spot



Hard exudate

Non-proliferative diabetic retinopathy:

Mild NPDR: (Control blood sugar)

Microaneurysmus only

Moderate NPDR: (Control blood sugar)

Microaneurysms – retinal haemorrhages - circumstances exudates – cotton wool spots – **minimal intraretinal microvascular anomalies (arterio-venous shunt) – minimal venous changes (looping and beading)**

Severe NPDR: (Control blood sugar + Consider panretinal laser coagulation)

All the above + **severe intraretinal microvascular anomalies (arterio-venous shunt) – severe venous changes (looping and beading)**

Diabetic macular edema 'DMO':

- Hard exudate here is in the center of the macula (can cause a loss of central vision). It's a sign of macular edema.

Optic coherence tomography (OCT) is used to diagnose early macular edema even in patients with very mild edema that you can't see it.

Treatment of diabetic macular edema (DMO):

1. **Focal laser photocoagulation:** Very effective in controlling diabetic macular edema.
2. **Intravitreal injection of anti-VEGF agents:** (Ranibizumab or Bevacizumab)

Proliferative diabetic retinopathy

Proliferative diabetic retinopathy:

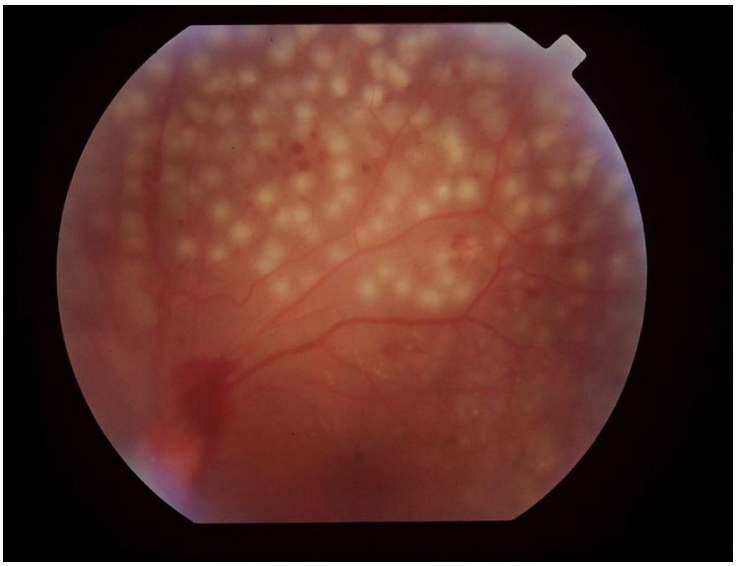
- **New vessels at the disc (NVD):** describes neovascularization on or within one disc diameter of the optic nerve head.
- **New vessels elsewhere (NVE):** describes neovascularization further away from the disc; it may be associated with fibrosis if long-standing.
- **New vessels on the iris (NVI):** also known as **rubeosis iridis,** carry a high likelihood of progression to **neovascular glaucoma.**

Treatment:

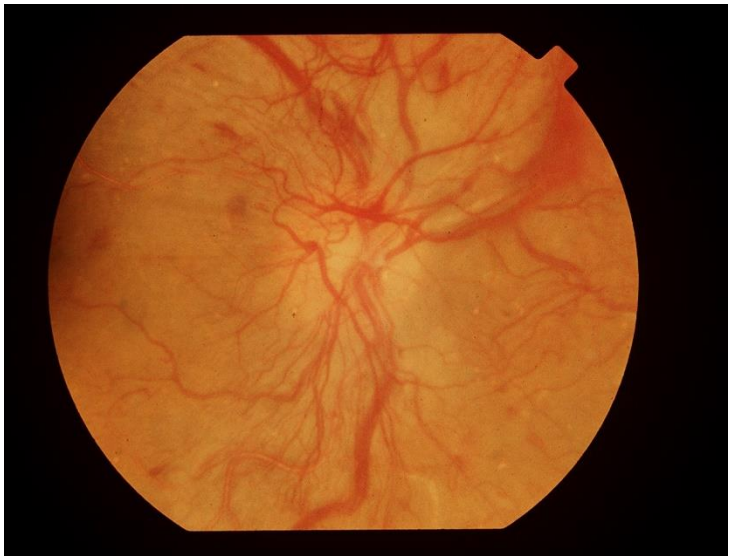
1- **Pan-retinal photocoagulation.**
2- **Intravitreal anti-VEGF injection.**

Complications of Retinal photocoagulation:

- Anterior segment complications such as corneal or lenticular opacification.
- Transient visual loss.
- Photocoagulation of the fovea.
- Macular edema.
- Hemorrhage.
- Choroidal Effusion
- Color vision alterations. Visual field defects and night vision problems.
- Hemeralopia.



Pan-retinal photocoagulation



Neovascularization

Grave's disease:

Introduction:

- Most common cause of both **bilateral and unilateral proptosis** in an adult

Pathogenesis:

- Autoimmune disease characterized with serum IgG antibodies bind to TSH receptors in the thyroid and causes overstimulation and high thyroid hormone production.
- Autoimmune antibodies infiltrate eye, **cause inflammation of extraocular muscles and associated with increased secretion of glycosaminoglycans and osmotic imbibition of water.**

Risk factors:

Smoking (most important) – family history

Systematic manifestation:

Pretibial myxedema, heat intolerance, weight loss etc…

Ocular manifestation:

Lid retraction - Proptosis and exophthalmos - Lid lag – restrictive myopathy

Investigations:

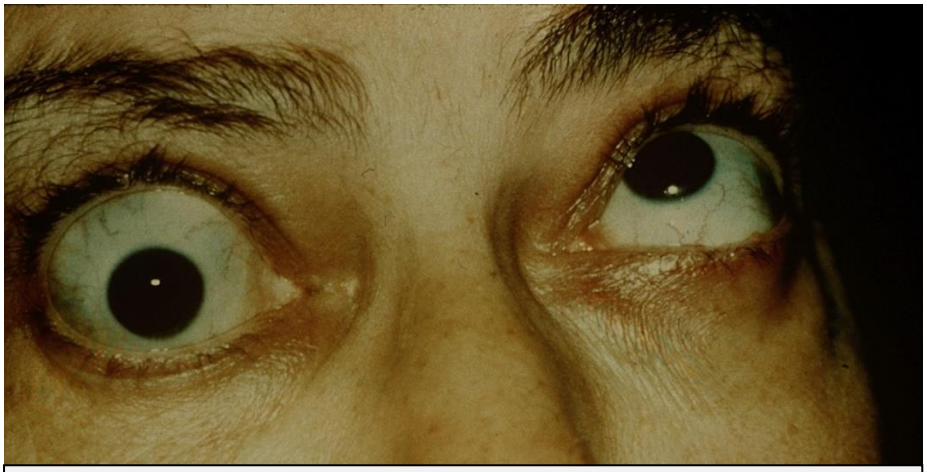
1- Thyroid function test: High T3, T4 and low TSH
2- Visual evoked potential: To exclude Optic neuropathy

Treatment:

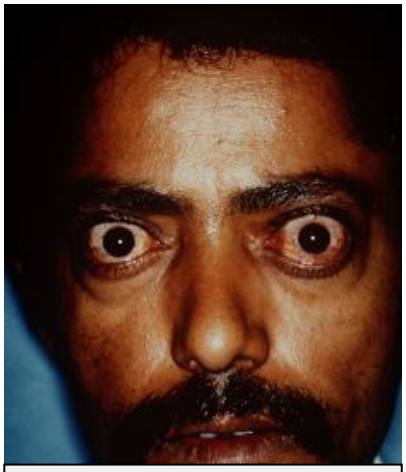
1- Anti-thyroid medications or Thyroid ablation with radioactive iodine **(for disease itself)**
2- Steroid, lubricants and eye protection before sleep **(for eye symptoms)**
3- If there is restrictive myopathy. Surgical intervention is required

Complications:

1- Optic neuropathy
2- Exposure or bacterial keratitis



Limitations of extraocular muscle



Exophthalmos with lid retraction

Tuberculosis:

Introduction:

- ❖ TB is a chronic **granulomatous infection** usually caused in humans by **Mycobacterium tuberculosis.**
- ❖ TB is primarily a pulmonary disease but may spread by the bloodstream to other sites; ocular involvement commonly occurs without clinically overt systemic disease.

Ocular manifestation:

Phlyctenular keratoconjunctivitis - Interstitial keratitis – Vitritis - Choroidal granuloma

- **Retinal vasculitis 'Eales disease'**
- **Anterior uveitis (Granulomatous uveitis)**
- **Mutton-fat keratic precipitation:** collection of inflammatory cells on the corneal endothelium appear large with yellowish color.

Investigations:

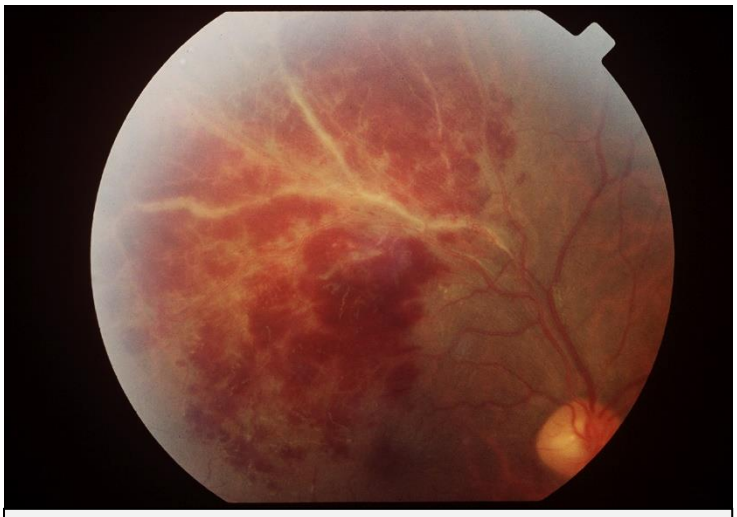
1- PCR and the interferon-gamma release assay (IGRA)
2- Aqueous or vitreous sampling rarely yields demonstrable (smear – acid-fast bacilli on Ziehl–Neelsen staining – or culture – Lowenstein–Jensen medium)

Treatment:

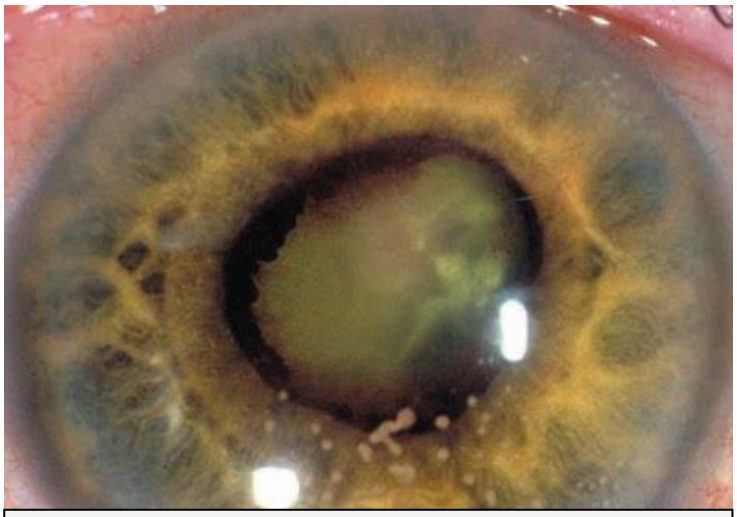
1- Prolonged Anti-TB therapy

- Isoniazid, rifampicin, pyrazinamide and ethambutol
- **ethambutol can cause optic neuropathy**

2- Topical and systemic steroids



Retinal vasculitis



Mutton-fat keratic precipitation

Sarcoidosis:

Introduction:

- It is a chronic disorder of unknown cause, manifesting with **non- caseating granulomatous inflammatory foci.**

Systematic manifestation:

The triad: **erythema nodosum - bilateral hilar lymphadenopathy - polyarthralgia**

Ocular manifestation:

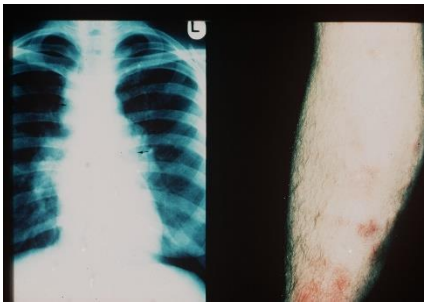
- Candle-wax exudate in the retina
- Optic nerve, Retinal, Choroidal Lid margin and conjunctival granulomas.
- **Mutton-fat keratic precipitates**

Investigations:

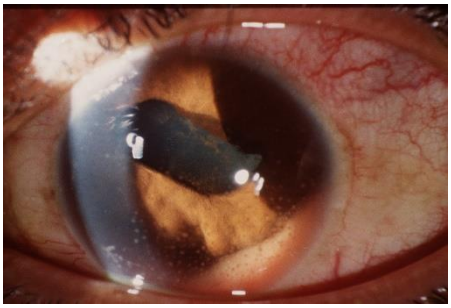
1. **Tuberculin skin test:** negative in sarcoidosis
2. **Chest X-ray:** showing bilateral hilar lymphadenopathy (BHL)
3. **Elevated serum ACE levels and/or elevated serum lysozyme**
4. **Abnormal liver enzyme tests.**

Treatment:

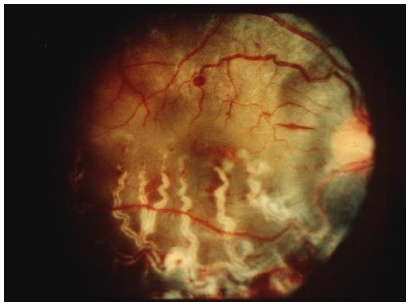
Steroid and NSAID's



Bilateral hilar lymph nodes ad erythema nodusm



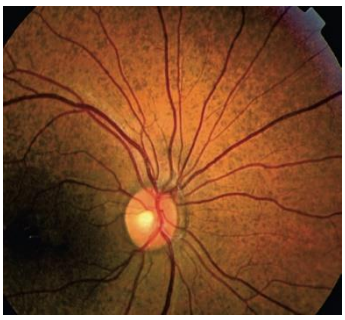
Granuloma with posterior synechia



Candle-wax exudate

Rubella:

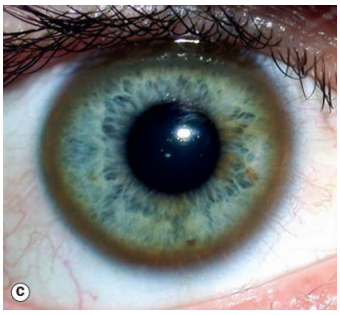
❖ Trans placental transmission could lead to congenital abnormalities.



Ocular manifestation:

- Congenital Cataract and Glaucoma
- **Microphthalmos (small eye)**
- **Pigmentary retinopathy: salt and pepper**
- **Anterior uveitis: unresponsive to steroids**

Wilson's disease 'hepatic degeneration':



Introduction:

- A rare condition involving the widespread **abnormal deposition of copper** in tissues. It is caused **by a deficiency of alpha 2 caeruloplasmin**, the major copper-carrying blood protein.

Systematic manifestation:

- Liver disease.
- Basal ganglia dysfunction.
- Psychiatric disturbances.

Ocular manifestation:

- **Kayser-Fleischer ring**: cons**ists** of a **brownish-yellow zone of fine copper dusting in peripheral descemet membrane** detected with gonioscopy (Important sign).
- **Anterior capsular 'sunflower' cataract**

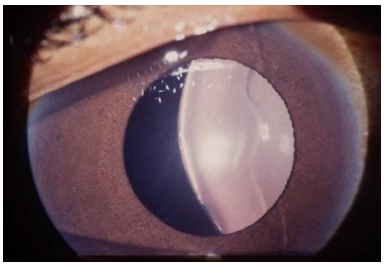
Treatment:

Penicillamine

Marfan's syndrome:

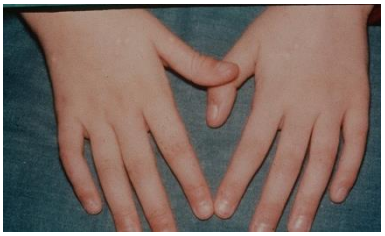
Introduction:

It is an autosomal dominant disease.



Systematic manifestation:

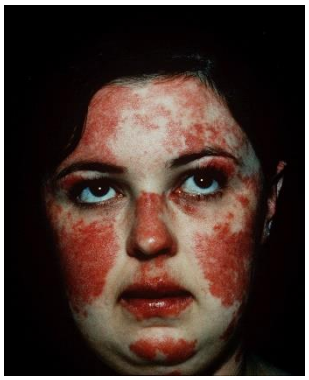
- **Arachnodactyly (Long fingers)**
- Heart diseases.
- Bone deformities



Ocular manifestation:

- **Lens subluxation:** due to weakness of the lens zonulles
- **Retinal detachment**
- **Axial myopia**
- **Angle anomaly lead to glaucoma**

Systemic Lupus Erythematosus:



- Autoimmune disease characterized by increased Anti-DNA, ANA, antiphospoholipid antibodies and decreased C3 & C4.
- **Retinal pathology:** cotton wool spots

Sjogren's syndrome:

Systematic manifestation:

Dryness of skin and mouth and arthralgia and polyneuropathy.

Ocular manifestation:

keratoconjunctivitis sicca "dryness of eye"

Investigation:

1. Schirmer tear test
2. Positive Rose Bengal staining (for keratoconjunctivitis sicca"
3. ANA, RF positive
4. Associated with **HLA-B8/DR3**

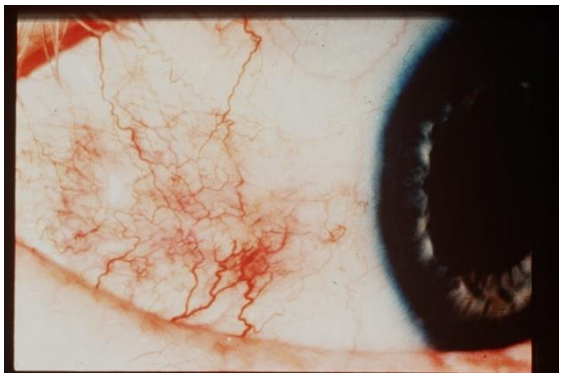
Rheumatoid Arthritis:

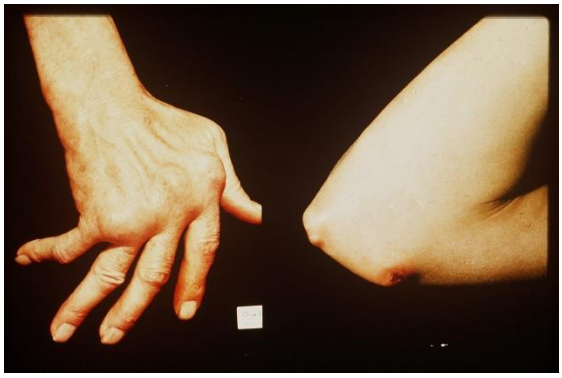
Ocular manifestation:

- **keratoconjunctivitis sicca, dryness and keratitis**

Investigation:

1. Positive Rose Bengal staining (for keratoconjunctivitis sicca)
2. RF positive





Ankylosing spondylitis:

Introduction:

Ankylosing spondylitis (AS) is characterized by inflammation, calcification and finally ossification of ligaments and capsules of joints with resultant bony ankylosis of the axial skeleton.

Systematic manifestation:

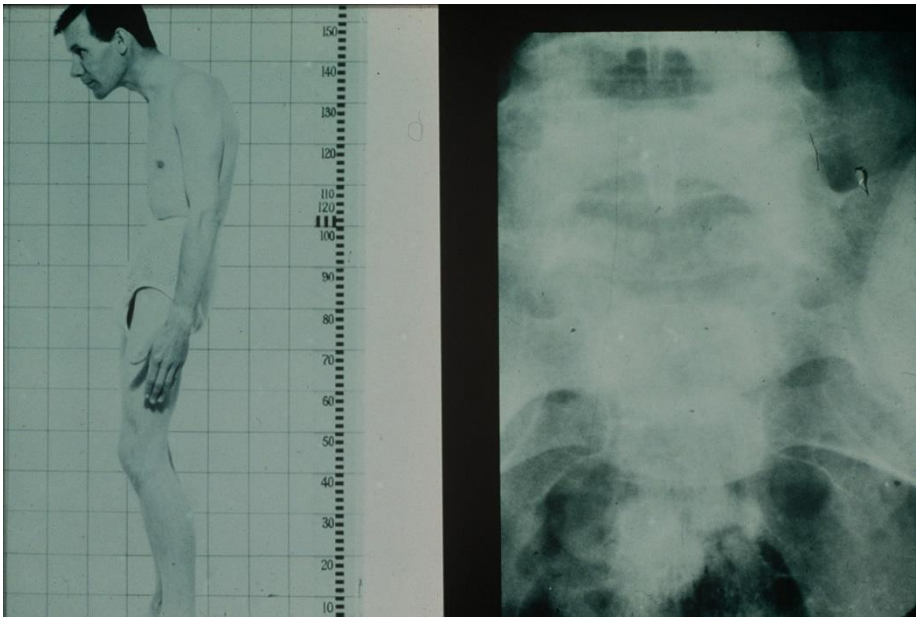
- Pain and stiffness in the lower back with limitation of movement
- Calcification of spinal ligaments gives rise to a **'bamboo spine'.**

Ocular manifestation:

- Acute **non- granulomatous** anterior uveitis

Investigations:

1. **HLA-B27-positive**
2. **X-ray: sacroiliac joints shows juxta-articular osteoporosis in the early stages**



Juvenile chronic arthritis:

Introduction:

- It is defined as **arthritis of unknown etiology** that begins **before the age of 16 years and persists for at least 6 weeks.**
- **Most common cause of anterior uveitis in children** in western countries.

Systematic manifestation:

a) **Pauciarticular form:** four or less joints are affected **(associated with 20% uveitis)**
b) **Polyarticular form:** five or more joints are affected. **(rare uveitis)**
c) **Stills disease:** fever, episodic erythematous maculopapular rash, lymphadenopathy and hepatosplenomegaly. **(extremely rare uveitis)**

Ocular manifestation:

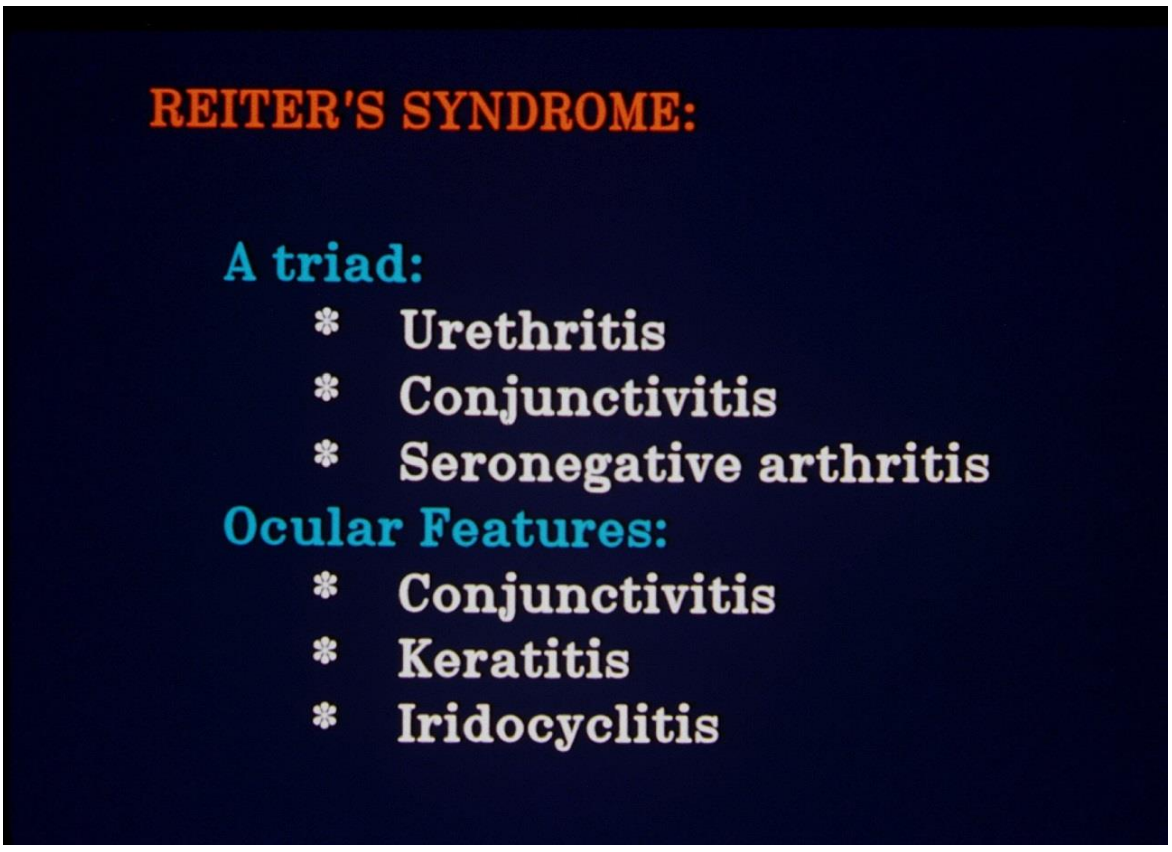
- Chronic non-granulomatous uveitis
- Band keratopathy
- Posterior synechiae

Investigations:

1. **Anti-ANA antibodies:** will be positive in majority of pauciarticular type
2. **Rehumatoid factor:** positive in some polyarticular type
3. **HLA-B27:** it will be positive in some patient

Treatment:

Topical and systematic Steroid and mydriatic agent to prevent posterior synechiae



Behçet disease:

Systematic manifestation:

- Vasculitis
- Recurrent aphthous oral ulcers
- genital ulceration

Ocular manifestation:

- Anterior non-granulomatous uveitis
- Transient hypopyon
- Retinal vasculitis
- Optic disc hyperemia

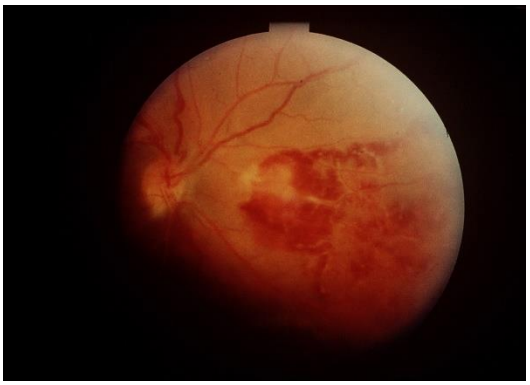
Investigations:

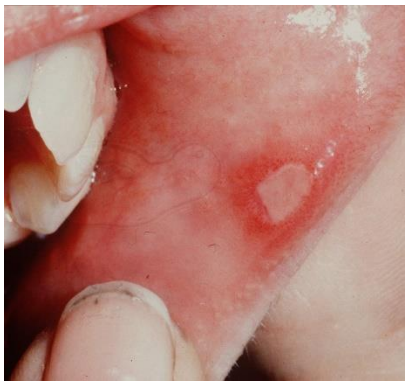
1- HLA-B51 is positive

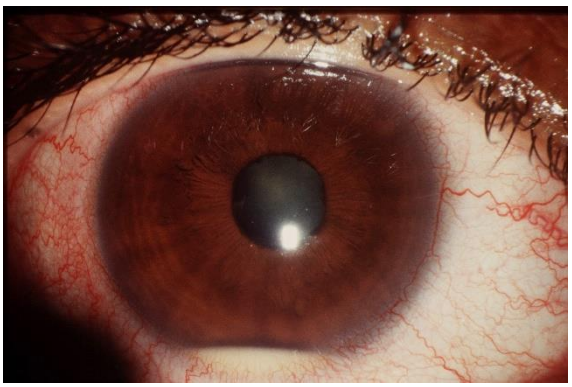
2- Pathergy test: pustule 24–48 hours after a sterile needle prick

Treatment:

- Steroid for a short time then Cyclosporine
- Anti-Alpha tumor necrosis agents ”infliximab” if case is resistant







Toxiplasmosis:

Introduction:

- Toxoplasmosis is caused by *Toxoplasma gondii after eating raw meat*
- Can be congenital: in the third trimester

Systematic manifestation:

- Congenital: Convulsions and intracranial calcification

Ocular manifestation:

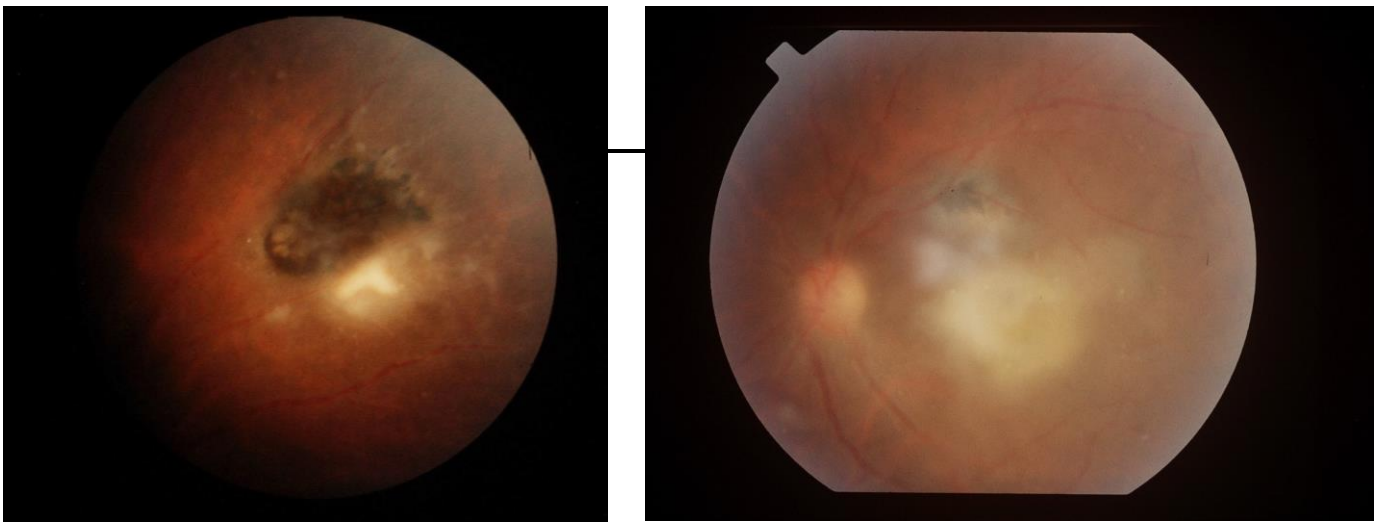
- posterior uveitis
- Macular lesion
- Retinochoroiditis ”fluffy white with pigmented scar”
- Unilateral floaters, blurring and photophobia.

Investigations:

PCR and serology

Treatment:

Clindamycin and Co- trimoxazole, 48hr later add steroid to suppress inflammation.



Vogt- koyanagi-Harada disease:

Introduction:

- Idiopathic Multisystem, autoimmune disease directed against melanocyte-containing tissues such as the uvea, ear and meninges.
- Most common cause of uveitis in the **kingdom of Saudi Arabia**

Systematic manifestation:

- Alopecia
- Vitiligo
- **Poliosis**: absence or decreased melanin in head hair, eyebrows or eyelashes.
- Deafness and vertigo
- Meningismus

Ocular manifestation:

- Bilateral granulomatous anterior uveitis
- Bilateral multifocal posterior uveitis
- Dalen–Fuchs nodules
- 'sunset glow' fundus

Investigations:

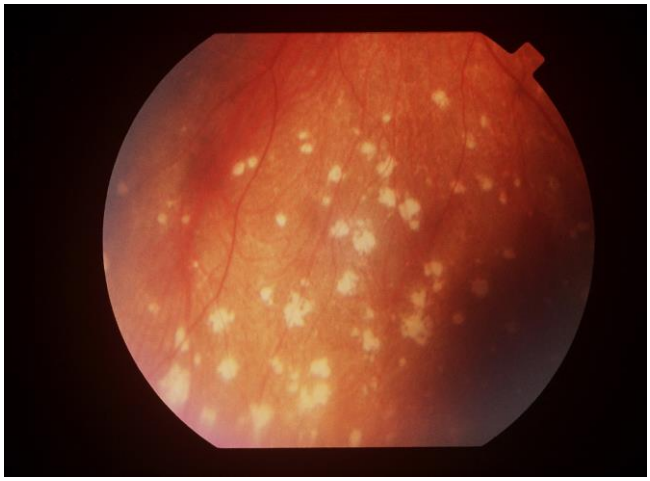
1. Associated HLA-DR1 and HLA-DR4
2. Lumbar puncture if diagnosis uncertain; CSF shows a transient lymphocytic pleocytosis, and melanin-containing macrophages.

Treatment:

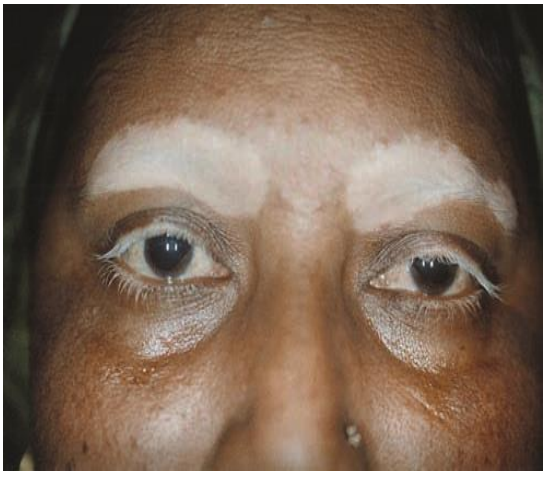
High-dose steroid or infliximab in case of steroid resistance

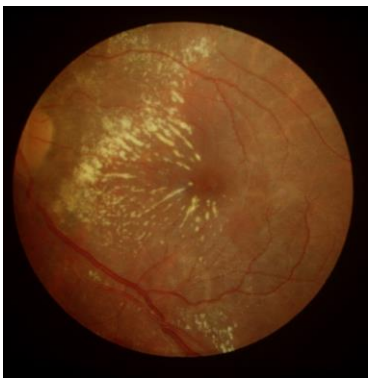
Complications:

1. Glaucoma
2. cataract
3. choroidal neovascularization,
4. subretinal fibrosis,
5. retinal atrophy



Dalen–Fuchs nodules





Hypertensive retinopathy:

Ocular manifestation:

Keith-wagener grouping:

Stage I & II: arteriolar attenuation (**silver wire and copper wire in the artery**) and increased light reflex

Stage III: Cotton-wall spots – Hard exudate – **Macular star** – retinal edema

Stage IV: All the above + Edema of optic disc

Giant cell arteritis:

Introduction:

Affect medium and large vessels. Anterior ischemic optic neuropathy is divided into two types: Arteritic , Non-Arteritic.

- **Non-arteritic anterior ischaemic optic neuropathy** (NAION): more common, caused by occlusion of the short posterior ciliary arteries resulting in partial or total infarction of the optic nerve head. Patient complains of sudden painless monocular visual loss; this is frequently discovered on awakening, suggesting a causative role for nocturnal hypotension.

- **Arteritic anterior ischaemic optic neuropathy** (AAION): Caused by giant cell arteritis (GCA). About 50% of patients with GCA have polymyalgia rheumatica (PMR)" pain and stiffness in proximal muscle groups, typically the shoulders and biceps, that is worse on waking and **jaw claudication**".

Risk Factors:

Old age, Females, smoking, low body mass index and early menopause.

Investigations:

1. Erythrocyte Sedimentation Rate 'ECR' ,
2. CBC
3. temporal artery biopsy.

Ocular manifestation:

1. **Ocular motor palsies, including a pupil-involving third nerve palsy,**
2. **Sudden, profound unilateral visual loss**
3. **strikingly pale 'chalky white' edematous disc**

Treatment:

High-dose of steroid

Done By:

Mojahed Otayf