433 Teams

OPHTHALMOLOGY

11

Ocular manifestations of systemic diseases

Color index:

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

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1- Diabetic retinopathy:

➢ Introduction:

- ❖ Systemic diseases involved in Retina can lead to blindness. This can be prevented by early treatment and control of the systemic diseases.
- ❖ The most common disease that involved the eye (particularly Retina) is DIABETES.
- ❖ DM can cause retinopathy, acute angle closure glaucoma, cataract and III, IV and VI cranial nerve palsy.
- ❖ Most common ocular manifestation is retinopathy.
- ❖ Most common cause of blindness, diabetics have more than 25 times chance of getting blind than the normal people do.
- ❖ Almost in all people with type 1 diabetes mellitus. 'due to early onset'
- ❖ Microvascular complications correlate significantly with DM.
- ❖ Hypertension, obesity and the control of blood sugar are risk factors that could be modified.
- ❖ Diabetic retinopathy is most common microvascular complication of DM and can lead to retinal degeneration.
- ❖ Because diabetic retinopathy often goes unnoticed until vision loss occurs, people with diabetes should get a comprehensive dilated eye exam at least once a year.
- ❖ Diabetic macular edema is a consequence of diabetic retinopathy that causes swelling in the area of the retina called the macula.
- ❖ Controlling diabetes—by taking medications, staying physically active, and maintaining a healthy diet—can prevent or delay or vision loss.

Ocular complications of diabetes:

• Common

Retinopathy.

Iridopathy (minor iris transillumination defects). ⍰⍰Unstable refraction.

• Uncommon

Recurrent styes.

Xanthelasma.

Accelerated senile cataract.

Neovascular glaucoma (NVG).

Ocular motor nerve palsies (3^{rd},4^{th},6^{th}).

Reduced corneal sensitivity.

• Rare

Papillopathy, pupillary light-near dissociation.

Wolfram syndrome (progressive optic atrophy and multiple). Neurological and systemic abnormalities, acute-onset.

Cataract, rhino-orbital mucormycosis.

Risk Factors:

1- Duration of diabetes (non-modifiable risk factor): The longer the duration of diabetes, the more risk of DR (By 15 years of diabetes, almost all patients come with DR).

2- Poor control of diabetes (most important modifiable risk factor).

3- Pregnancy**(poor pre-pregnancy control, too rapid tightening of control during early stages).**

4- Hypertension.

5- Nephropathy (if sever, it is associated with worsening of retinopathy).

6- Others: Hyperlipidemia, smoking, cataract surgery, obesity and anemia.

Pathogenesis:

Development of progressive vasculopathy characterized by dysfunction of endothelial cells lining retinal capillaries and the loss of the inner Blood-Retina-barrier. The inner Blood-Retina-barrier contains tight- junctions that reduce the space between adjacent endothelial cells lining the fine capillaries of the retinal microvasculature to form a selective and regulatable barrier. So, if there is a breakdown of the blood-retinal barrier, which normally prevents water movement in the retina, fluid (lipoprotein) will accumulate in the retinal tissue leading to macular edema, thickening and swelling which distorts central vision.

This will also result in the occlusion of the capillaries causing to hypoxia and ischemia. The retina will react by producing angiogenesis factors like the Vascular Endothelial Growth factor (VEGF) and they will induce more breakdown of inner blood retinal-barrier (increasing the hyper permeability) and induce the formation of new vessels.

At this stage of new vessels, the disease called proliferative disease. These new vessels are not healthy; they can bleed causing loss the vision from hemorrhage in the eye (Vitreous hemorrhage) and can lead to the formation of fibrous tissue causing traction in the retina (Retinal detachment).

At the level of retinal microvasculature, the endothelial cells lining the capillaries become activated and they express adhesion of molecules (intercellular adhesion molecules (ICAMs) allowing the circulating leukocytes to come and adhere to the endothelial cells of the capillaries. This is known as leukostasis (inflammation in the diabetic retina).

In the case of sever ischemia; new vessels not only form in the retina, but in the iris as well (Rubeosis iridis). This results in neovascular glaucoma.

RETINAL CHANGES:

-Hard exudates on the retina and macular edema resulted from the leakage.

- microaneurysms ‘blots and dots’

- soft exudates ‘cotton-wool spots’.

Venous changes: beading vs looping

-hemorrhage if neovascularization results in weak vessels and easy to break.

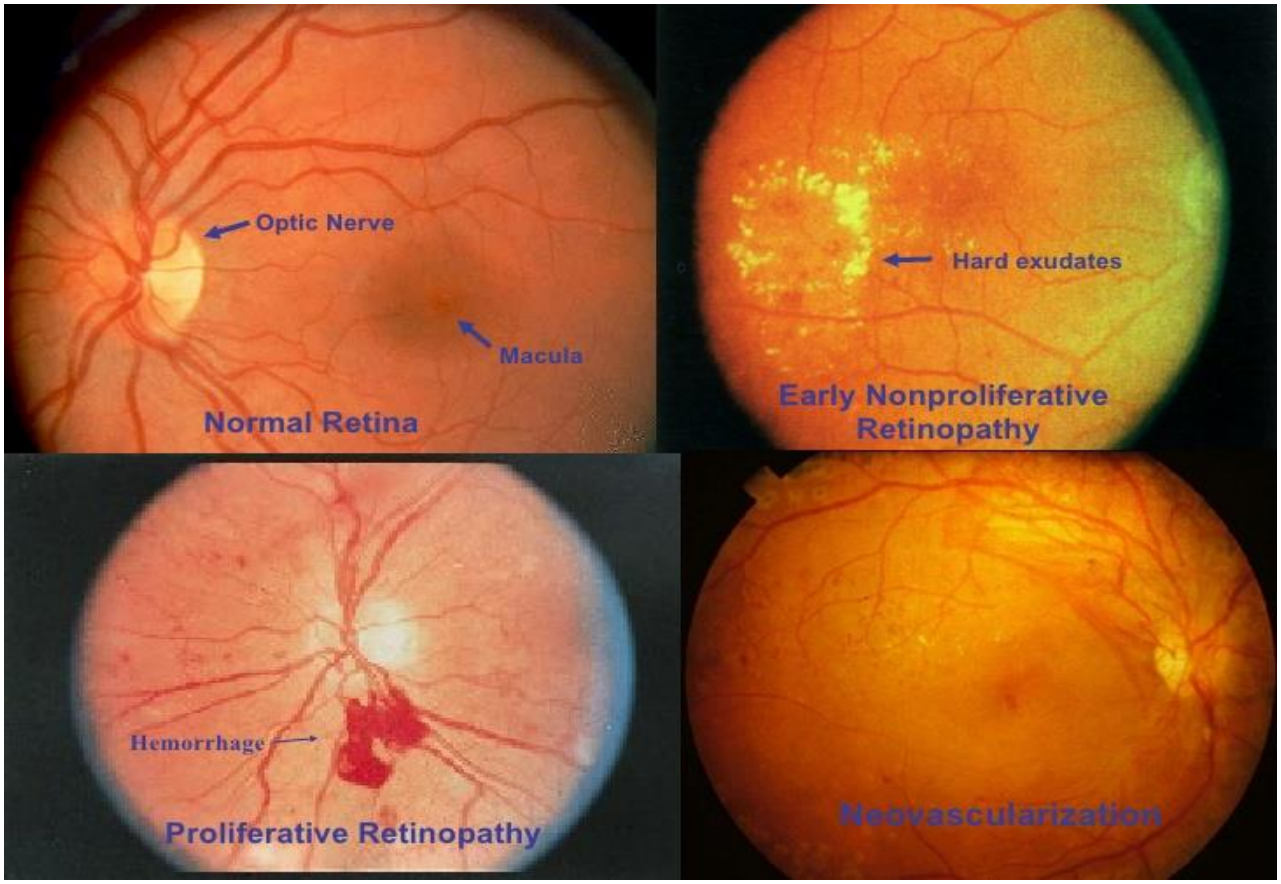


image 1: show the difference between normal eye and stages of diabetic retinopathy

Sever non-proliferative diabetic retinopathy:

Does not cause hemorrhage (no new vessels); we see only sign of ischemia.

Proliferative diabetic retinopathy:

There are new vessels. *The only means of preventing blindness due to diabetic retinopathy is through

screening and early treatment.

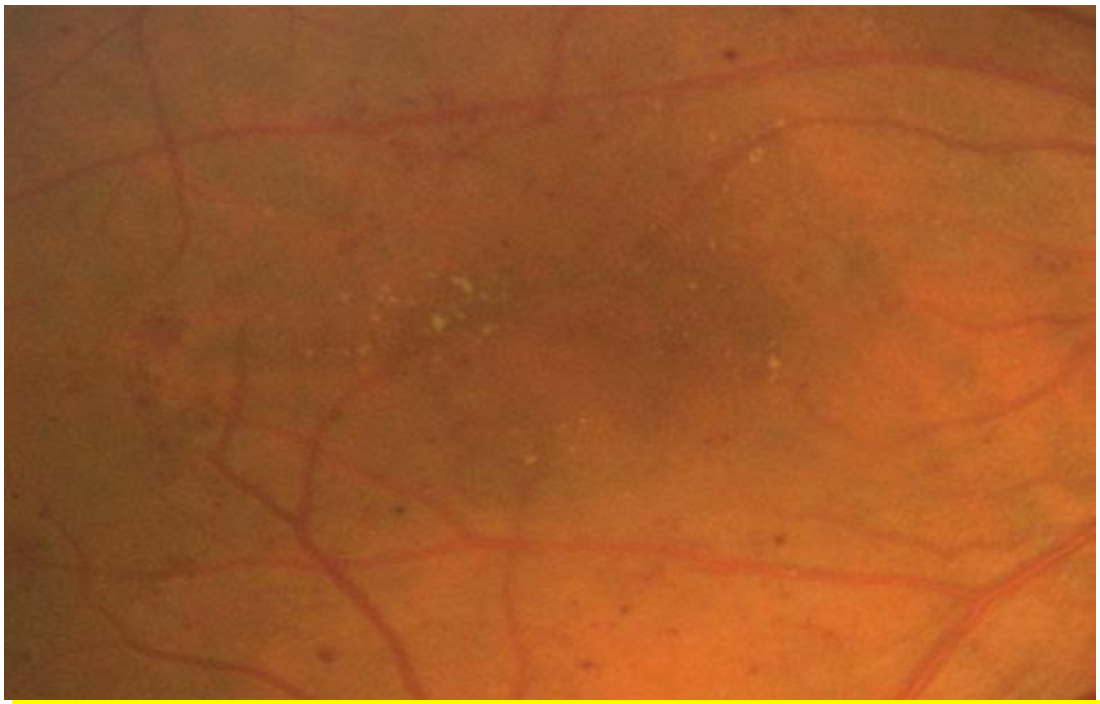


image2: we can see that there are small exudates with microaneurysims.

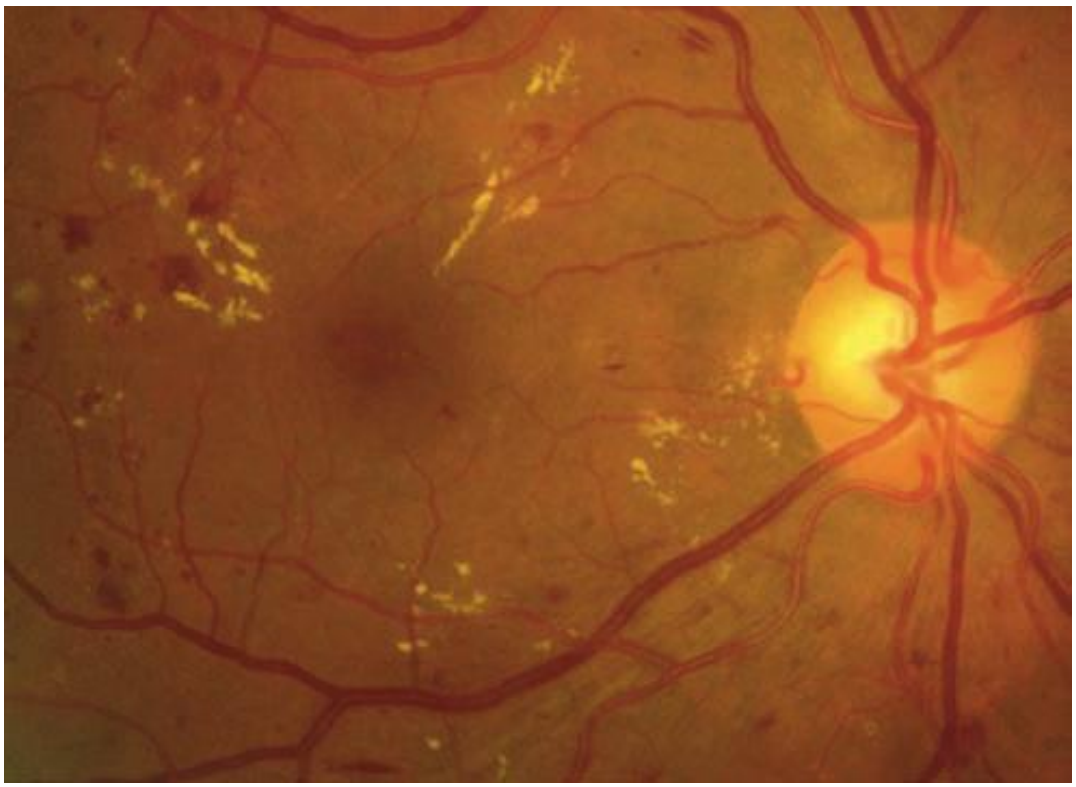


image3: more extensive exudates some associated with microaneurysims.

Diabetic macular edema 'DMO':

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

-Nowadays, 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:

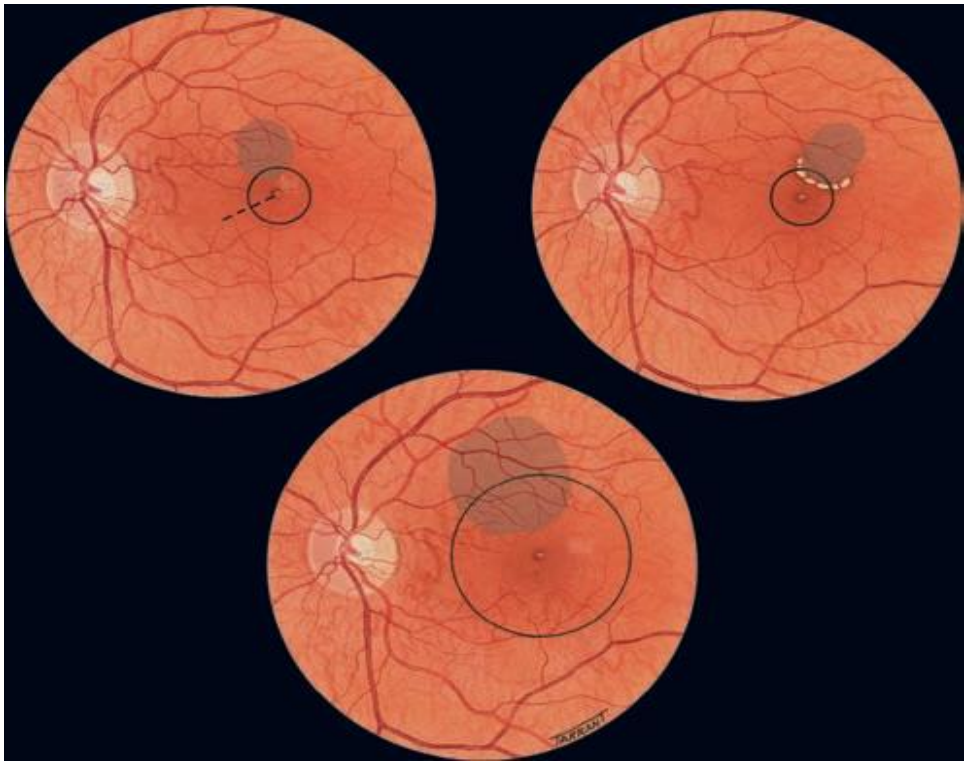
Treatment of micro aneurysms with laser (After 6 months there will be a scar from the laser treatment but no hard exudate). Very effective in controlling diabetic macular edema.

2- Intravitreal injection of anti-VEGF agents (anti- vascular endothelial growth factor): (Ranibizumab or Bevacizumab) In addition to laser, we frequently inject Intravitreal injection of anti-VEGF agents to

help control diabetic macular edema.

* Diabetic retinopathy patients have progressive macular vascular occlusion which will cause ischemia of retina (hypoxia).

image4: we can see significant macular edema progression.



Signs of Retinal Ischemia (Patient is at high risk to develop new vessels):

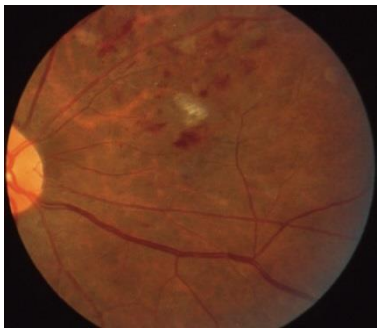
At this stage, sever non-proliferative diabetic retinopathy (Non-PDR). Patient should receive laser Panretinal photocoagulation.

- Cotton wool spots (less important sign and not used in the classification of non-PRD).

- Venous changes (The most reliable signs of retinal ischemia in diabetic retinopathy).

- Intraretinal microvascular abnormalities (IRMA): are arteriolar–venular shunts that run from retinal arterioles to venules, thus bypassing the capillary bed and are therefore often seen adjacent to areas of marked capillary hypoperfusion. * Dilated telangiectatic capillaries.

- Intraretinal Hemorrhage: The extent of involvement is a significant marker of the likelihood of progression to proliferative diabetic retinopathy.

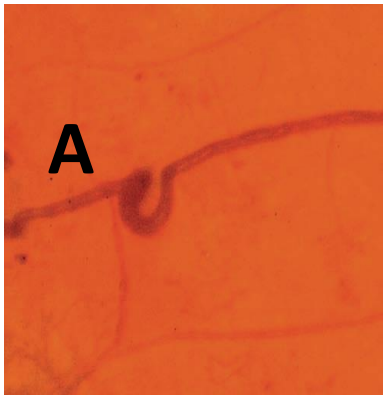


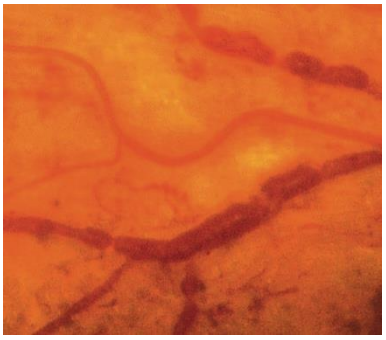
Images3,4,5 &6 show cotton wool spots and Venous changes.

(A) Looping;

(B) beading;

(C) severe segmentation





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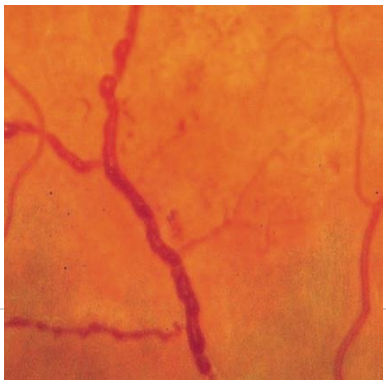


C

proliferative diabetic retinopathy

-Fibrous tissue, initially fine, gradually develops in association as vessels increase in size. They are most commonly seen at the posterior pole:

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- Scatter laser treatment panretinal photocoagulation. (The mainstay of PDR treatment) *Protect the patient from blindness. *Apply scattered laser beams throughout the retina starting from the vascular arched, don't reach the macula and extend up to the periphery.

2- Intravitreal anti-VEGF injection.

- Complications of Retinal photocoagulation:

- **1-** Anterior segment complications such as corneal or lenticular opacification.
- **2-** Transient visual loss. **3-** Photocoagulation of the fovea.
- **4-** Macular edema. **5-** Hemorrhage. **6-** Choroidal Effusion. **7-** Color vision alterations. **8-** Visual field defects and night vision problems. **9-** Hemeralopia.

2- Grave's disease:

- Autoimmune disease characterized with serum IgG antibodies bind to TSH receptors in the thyroid and causes overstimulation and high thyroid hormone production.
- Ocular manifestations of Grave's include th following:
  a) Lid retraction
  b) Proptosis and exophthalmos
  c) Lid lag
  d) Limitation of extraocular muscles movements
  e) Infiltration of hormones in the globe
- Also other symptoms and signs of hyperthyroidism e.g. pretibial myxedema, heat itolerance, weight loss etc…

3- Tuberculosis:

❖ The three most common causes of intraocular inflammation in the kingdom:

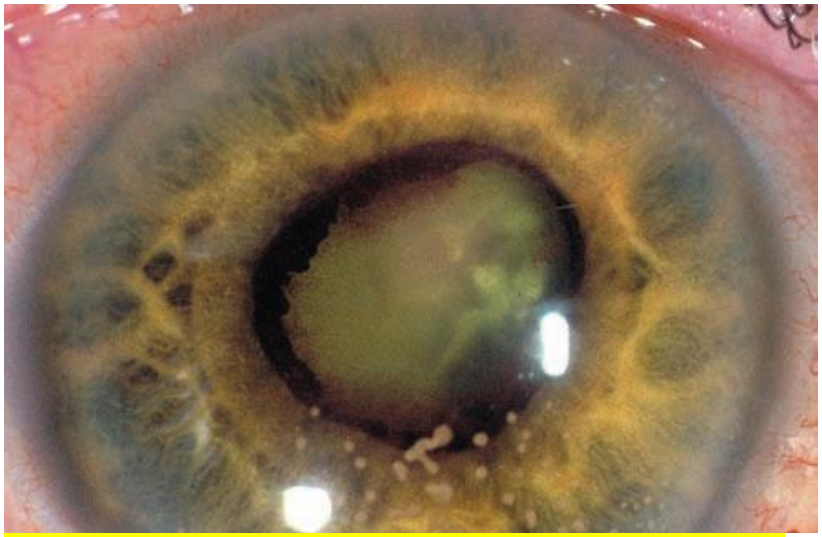
1) Vogt–Koyanagi–Harada (VKH) syndrome
2) Tuberculosis
3) Behçet disease

❖ 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 manifestations present as:

a) Anterior uveitis: causes blindness if not treated
b) Phlyctenular keratoconjunctivitis: small, circumscribed lesions at the corneal limpus.
c) Interstitial keratitis
d) Retinal vasculitis ‘Eales disease’
e) Vitritis
f) Choroidal granuloma
g) Mutton-fat keratic precipitation: collection of inflammatory cells on the corneal endothelium appear large with yellowish color.



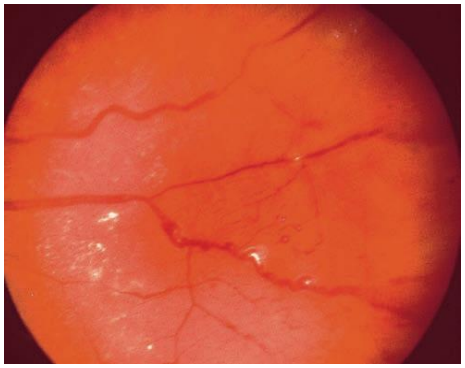


image: shows large mutton-fat keratic precipitates

Image: Eales disease: Fundus photo of the peripheral retina, revealing vascular tortuosity and peripheral retinal neovascularization.

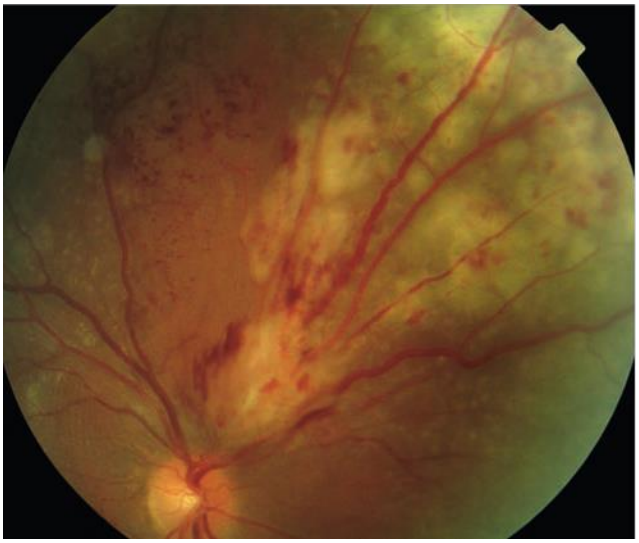


Image shows progressive retinal vasculitis in a patient with TB.

❖ Treatment:

1- Prolonged multi-drug therapy (4 drugs in 2 months, then continue for 6 months with 2 drugs): isoniazid (with Vitamin B6 (pyridoxine) to prevent the development of peripheral neuropathy), rifampin, ethambutol, pyrazinamide.

2- Topical and systemic steroids may be used concomitantly to reduce inflammation- induced damage.

4- Syphilis: not common in our region

- could be either congenital syphilis 'transplacental infection' or acquired.
- Congenital syphilis:
  - Interstitial Keratitis
  - Chorioretinitis
- Acquired syphilis:
  - Ocular chancre.
  - Iridocyelitis.
  - Interstitial Keratitis.
  - Chorioretinitis.
  - Neuro-ophthalmic (nerve palsies, optic neuritis...).

5- Sarcoidosis:

- It is a chronic disorder of unknown cause, manifesting with non- caseating granulomatous inflammatory foci.
- It more frequently affects Japanese people, it increases with good hygiene and it is one of the most common systemic associations of uveitis.
- **Eye Lesions:**
  1) Lid margin and conjunctival granuloma.
  2) Acute iridocyclitis.
  3) Chronic granulomatous iridocyclitis.
  4) Peripheral retinal periphlebitis.
  5) Choroidal granulomas.
  6) Retinal granulomas.
  7) Optic N. granulomas.
  8) Mutton-fat keratic precipitates

❖ Diagnosis confirmed by: CXR,ACE, serum lysozyme and Ca and liver function.

Image shows CXR: bilateral hilar lymphadenopathy in patients with granulomatous uveitis

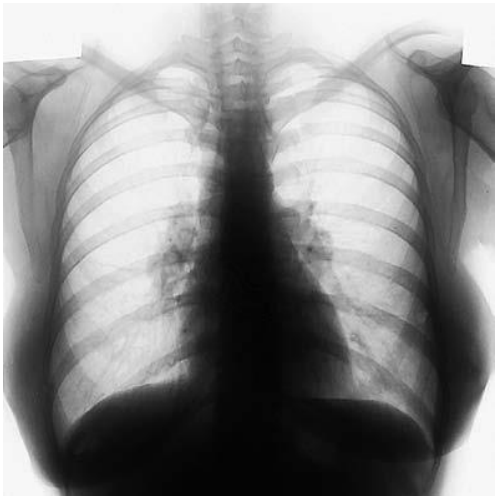


Image show erythema nodosum

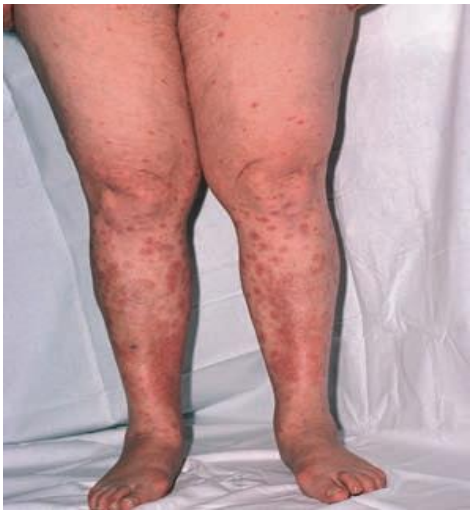
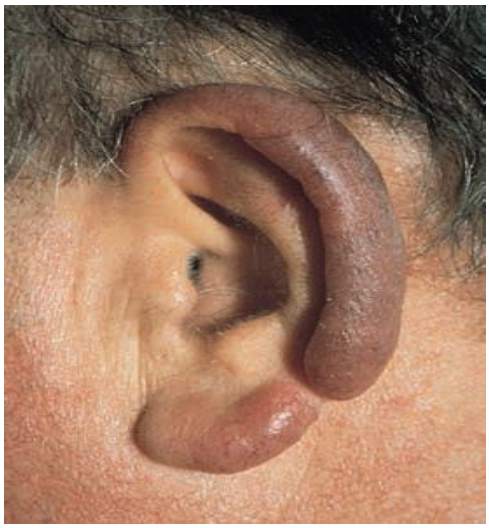


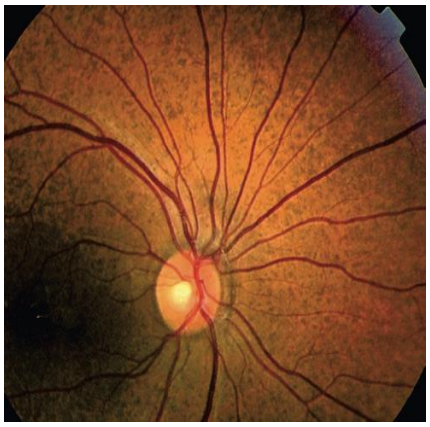
Image show lupus pernio



6- Rubella:

- Transplacental transmission could lead to congenital abnormalities,
- Ocular manifestations include:
  a) Cataract
  b) Microphthalmos
  c) Pigmentary retinopathy: salt and pepper
  d) Glaucoma
  e) Anterior uveitis: unresponsive to steroids

Image shows rubella retinopathy



7- Wilson's disease 'hepatic degeneration':

- 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.
- Presentation:

1- liver disease.

2- basal ganglia dysfunction.

3- psychiatric disturbances.

4- Kayser-Fleischer ring (in 90% of patients with neurological signs): consists of a brownish-yellow zone of fine copper dusting in peripheral descemet membrane detected with gonioscopy (Important sign).

5- Anterior capsular 'sunflower' cataract (Not common): Copper deposition.

8- Marfan's syndrome:

It is an autosomal dominant disease; the most important feature in the eye in this patient is lens subluxation due to weakness of the lens zonulles. This patient can also have anomalies in the angle of the anterior chamber causing glaucoma. They are also at risk of developing retinal detachment.

9- Systemic Lupus Erythematosus:

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

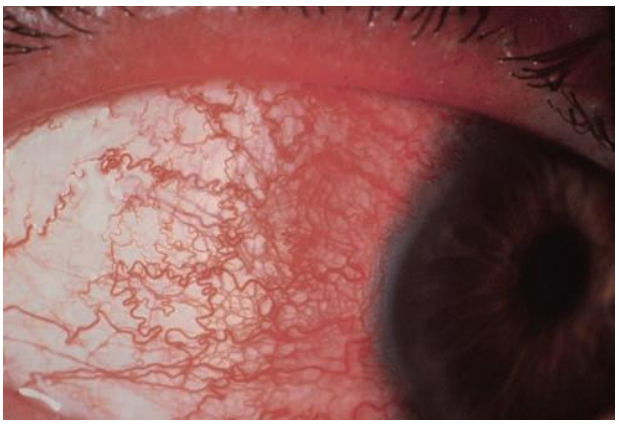
10- Rheumatoid Arthritis:

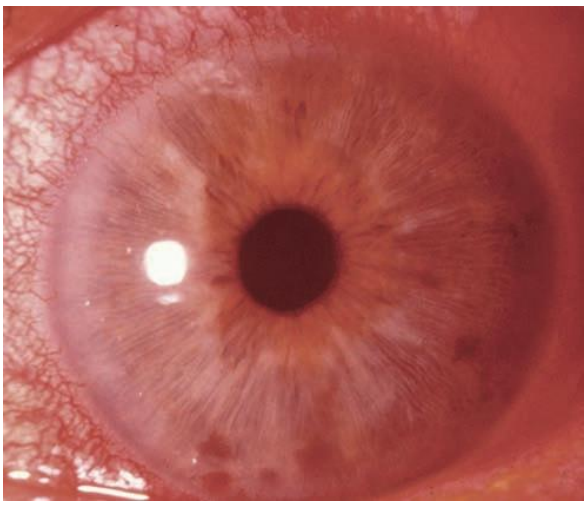
- Not common here in our region, it's rare. The patient had positive Rheumatoid factor
- - It manifest in the eye as keratoconjunctivitis sicca, dryness and keratitis

11- Ankylosing spondylitis:

- 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. It more commonly affects males, of whom 90% are HLA-B27-positive.
- Unilateral usually
- Ocular manifestation most commonly in a form of acute non- granulomatous anterior uveitis
- Pain and stiffness in the lower back with limitation of movement with calcification of spinal ligaments gives rise to a 'bamboo spine'. Radiological changes often predate clinical symptoms
- HLA positive disease: IBD, psoriasis, Reiter and ankylosing spondylitis

Images show signs of anterior uveitis: ciliary injection and meiosis





12- Juvenile chronic arthritis:

- 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.
- The patient may get no complains until they get blind.
- Can have several forms:
  a) Stills disease: fever, episodic erythematous maculopapular rash , lymphadenopathy and hepatosplenomegaly.
  b) Polyarticular: five or more joints are affected.
  c) Oligoarticular form: four or less joints are affected
- Risk factors of uveitis:

-Oligoarticular onset

-Being a girl

-Young age < 4

-Positive antinuclear antibody

- screening: ANA, RF & HLA-B27
- complications include:
  (a) glaucoma
  (b) amblyopia
  (c)maculopathy
  (d) phthisis

13- Behçet disease:

- Behçet disease (BD) is an idiopathic, multisystem syndrome characterized by recurrent aphthous oral ulcers, genital ulceration and uveitis. It's not in whites but is common here.

- It is non-granulomatous with transient mobile hypopyon in a relatively white eye Hypopyon in HLA-B27 is solid " doesn't move".
- Patients with relapsing/remitting acute onset of bilateral panuveitis with retinal vasculitis and often spontaneous resolution even without treatment
- **Manegment:** Steroid for a short time then Ciclosporin Anti Alpha tumor necrosis agents"infliximab" if case is resistant

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14- Toxiplasmosis:

- Most common cause of infectious uveitis in many countries. Common in France due to their habits." they eat raw meat"

- Constitutes 20-60% of all posterior uveitis.
- Can be congenital: in the third trimester Showing ophthalmologic manifestation retinochoroiditis "fluffy white with pigmented scar", which has a predilection for the posterior pole.
- Bilateral in most cases.
- **Symptoms:** Unilateral floaters, blurring and photophobia.
- **Management:** Clindamycin and Co- trimoxazole, 48hr later add steroid to suppress inflammation.
- Recurrent retinitis may cause blindness

15- sjogren's syndrome:

- ❖ The syndrome of dry eyes 'keratoconjunctivitis sicca'.
- ❖ Associated with HLA-B8/DR3
- ❖ Accompanied with dryness of skin and mouth and arthralgia and polyneuropathy.
- ❖ Investigated by:
  - (a) Schirmer tear test
  - (b) Rose Bengal staining
  - (c)ANA, RF positive

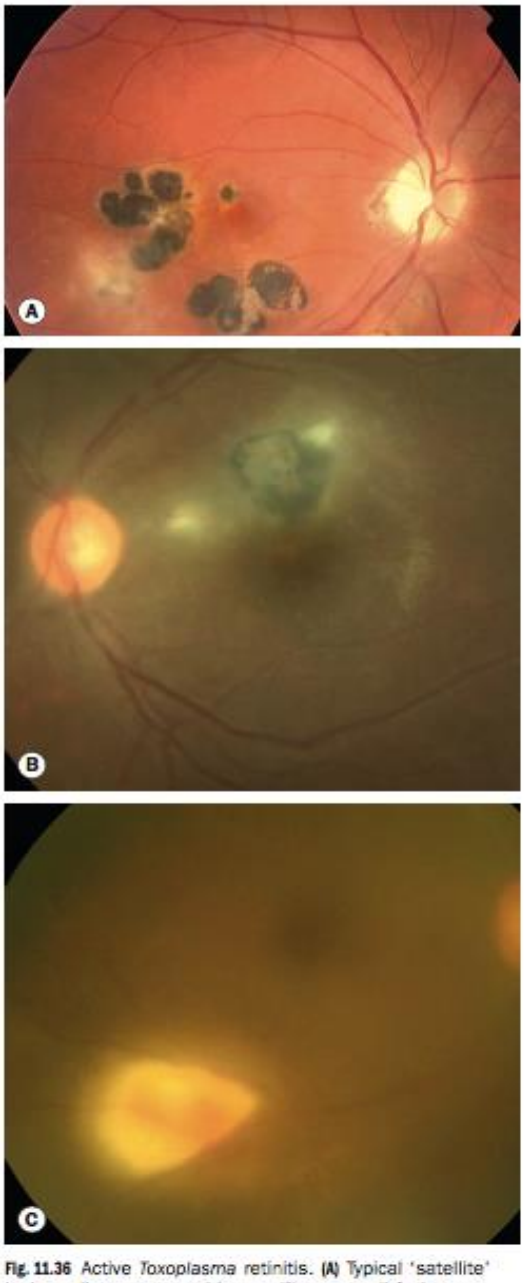


Fig. 11.36 Active *Toxoplasma* retinitis. **(A)** Typical 'satellite' lesion adjacent to an old scar; **(B)** two small foci; **(C)** severe vitreous haze and 'headlight in the fog' appearance of lesion
(Courtesy of S Chen – fig. A; C Pavesio – figs B and C)

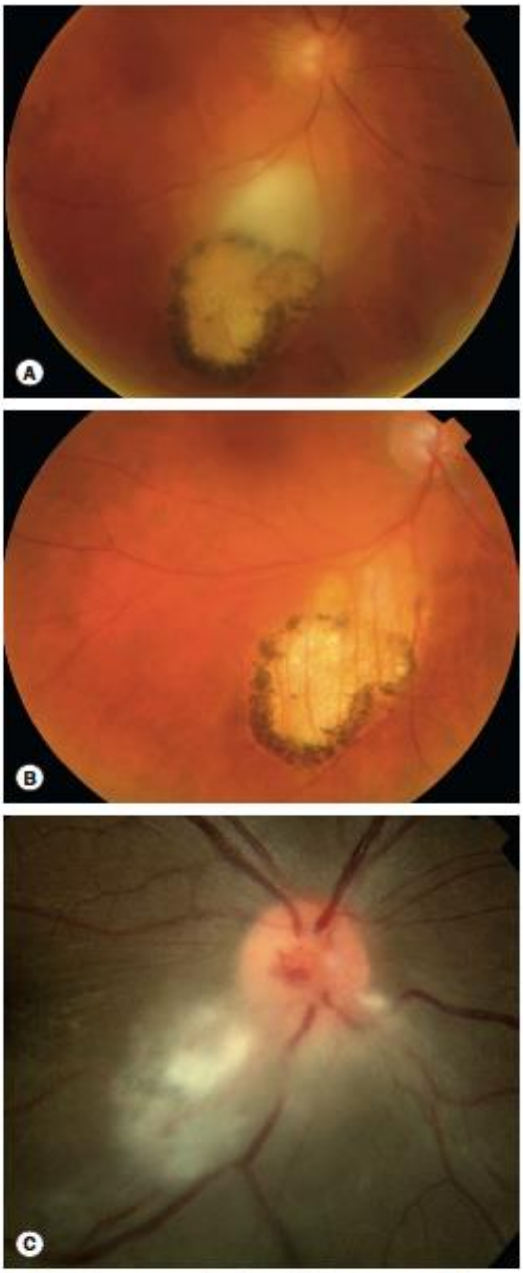
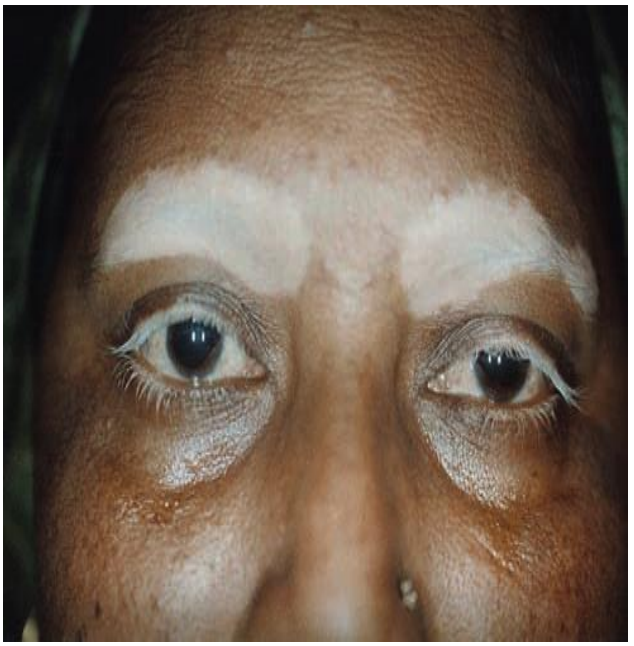


Fig. 11.37 Common complications of *Toxoplasma* retinitis. **(A)** Macular involvement, at presentation and **(B)** following treatment; **(C)** juxtapapillary lesion involving the optic nerve head

16- Vogt- koyangi Harada disease:

- Most common cause of uveitis in the kingdom
- Mean age is 28 year old
- Most common cause of intraocular inflammation
- Idiopathic Multisystem, autoimmune disease directed against melanocyte-containing tissues such as the uvea, ear and meninges.
- Symptoms:
  i) Alopecia
  ii) Vitiligo
  iii) Poliosis : absence or decreased melanin in head hair, eyebrows or eyelashes.
  iv) Deafness and vertigo
  v) Meningismus



- **Complication:**
  (a) Glaucoma,
  (b) cataract,
  (c) choroidal neovascularization,
  (d) subretinal fibrosis,
  (e) retinal atrophy

Image shows: Depigmented fundus appearance ('sunset glow' fundus)

Image shows: Vitiligo and poliosis in Vogt–Koyanagi–Harada syndrome

❖ 17- Sickel cell disease: The doctor passed the slides and said not important

- ❖ Retinopathy due to sickling of RBC causing peripheral vascular occlusion of the circulation and neovasculization in the periphary. Looks like “Sea fans”

18- Hypertensive retinopathy:

- ❖ the primary response of the retinal arterioles to systemic hypertension is vasoconstriction; this is less marked in older individuals due to involutional sclerosis conferring increased rigidity. Arteriolosclerosis refers to hardening and loss of elasticity of small vessel walls, manifested most obviously by arteriovenous (AV) Nipping(nicking) at crossing points; its presence makes it probable that hypertension has been present for many years, even if the BP is currently controlled. Nipping: Thinning of the vein after the artery and it's changing direction, to avoid pressure by the artery.
- ❖ **Signs of hypertensive retinopathy:**
  - (a) Attenuation of the arterioles, increase light reflex as a result of chronic hypertension there will be thickening of arterioles giving the appearance of **silver wire and copper wire in the artery.**
  - (b) Cotton wool spot
  - (c) Hard exudates may result in a macular star if deposited around the fovea
  - (d) Retinal hemorrhage
  - (e) Bilateral edema of the optic nerve which seen in malignant hypertension

19- Giant cell arteritis:

- ❖ 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. More common, comes with people with diabetes, hypertension, hyperlipidaemia, collagen vascular disease, antiphospholipid antibody syndrome, hyperhomocysteinaemia, sudden hypotensive events, cataract surgery, sleep apnoea syndrome and erectile

dysfunction." 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".

- Risk factors: Old age, Females, smoking, low body mass index and early menopause.

- **Ocular features:** Ocular motor palsies, including a pupil-involving third nerve palsy, Sudden, profound unilateral visual loss not uncommonly preceded by transient visual obscurations (amaurosis fugax) (Latin *fugax* meaning *fleeting*, Greek *amaurosis* meaning *darkening*, *dark*, or

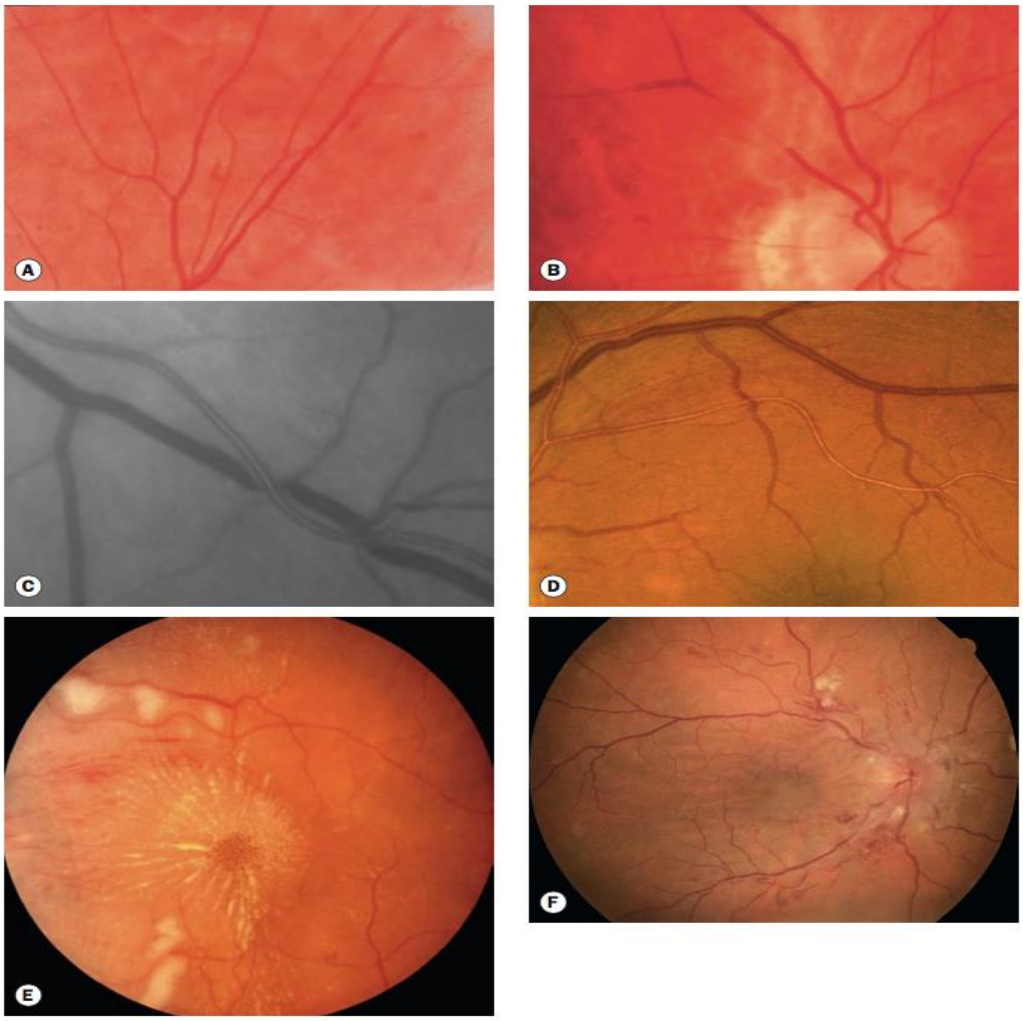


Fig. 13.43 Hypertensive retinopathy. **(A)** Generalized arteriolar attenuation; **(B)** focal arteriolar attenuation; **(C)** red-free photograph showing arteriovenous nipping; **(D)** 'copper wiring'; **(E)** grade 3 retinopathy with macular star; **(F)** grade 4 hypertensive retinopathy

obscure) which is a

painless transient monocular visual loss. And sometimes by double vision. Periocular pain is also common**. Signs**: strikingly pale 'chalky white' edematous disc is particularly suggestive of GCA.

- ❖ Once the vision is lost you cannot restore it.
- ❖ Investigated by: Erythrocyte Sedimentation Rate 'ECR' ,

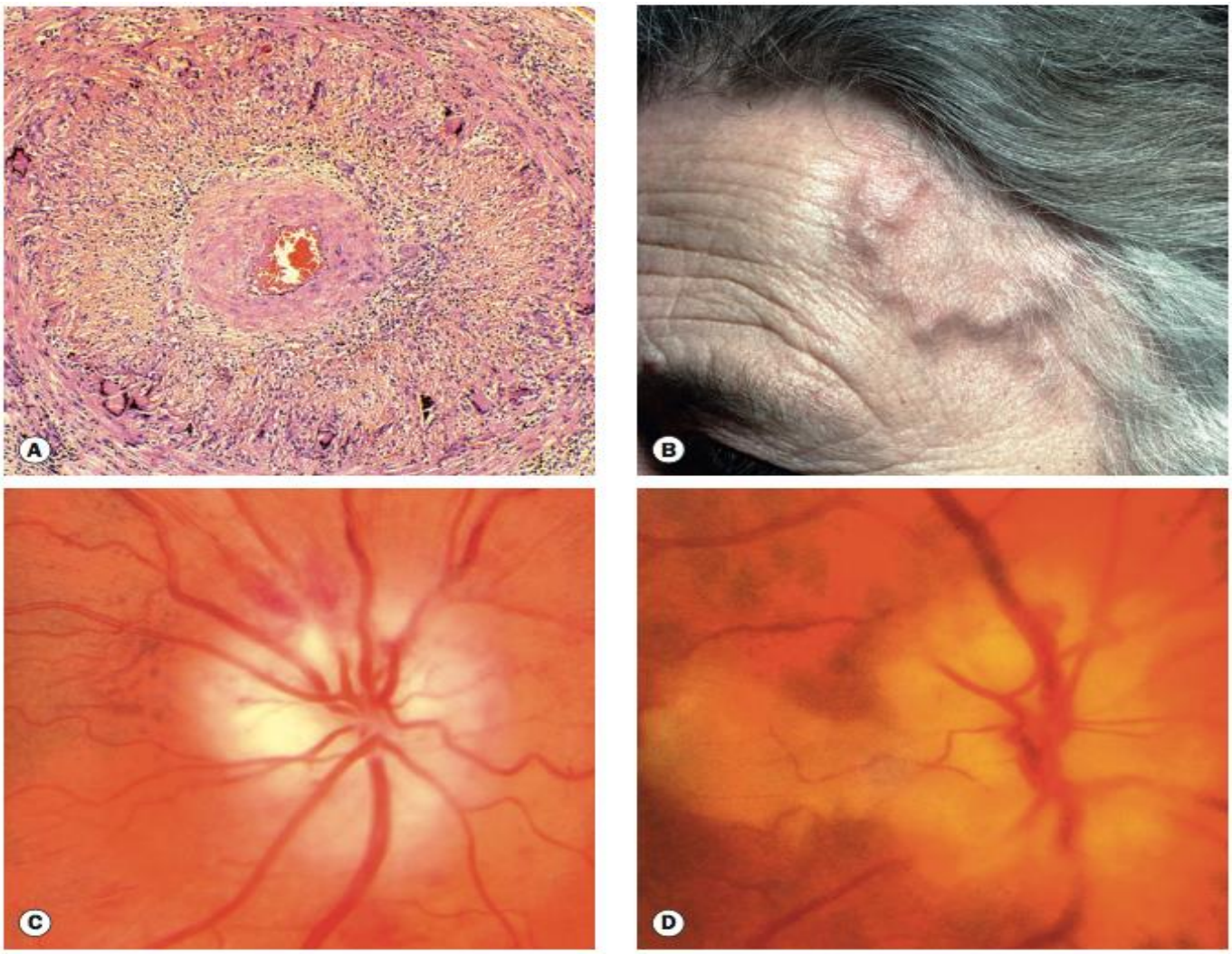


Fig. 19.14 Giant cell arteritis. **(A)** Histology shows transmural granulomatous inflammation, disruption of the internal elastic lamina, proliferation of the intima and gross narrowing of the lumen; **(B)** the superficial temporal artery is often pulseless, nodular and thickened; **(C)** pale swollen disc in arteritic ischaemic optic neuropathy; **(D)** ischaemic optic neuropathy and cilioretinal artery occlusion

(Courtesy of J Harry and G Misson, from Clinical Ophthalmic Pathology, *Butterworth-Heinemann 2002 – fig. A; S Farley, T Cole and L Rimmer – fig. B; SS Hayreh – figs C and D)*

CBC and temporal artery biopsy.

- ❖ Treatment: large doses of steroids and protect the eye

Summary:

Disease	Structure involved and presentation	Comments
Diabetes Mellitus	-The Retina: 2 changes: 1-Break down of the blood retinal barrier → early loss of peri-cytes and early loss of tight junction proteins between endothelial cells → vessels leak fluid, blood and lipoproteins → retinal edema→ visual loss 2- progressive occlusion of retinal microvasculature → ischemia and hypoxia leakage → neovascularization → angiogenesis → bleeding → vitreous hemorrhage → fibrous tissue formation → traction of the retina → detachment. Central signs of retinal ischemia: Cotton wool spots, Hard exudates, Venous beading and looping -The Iris: Neo-vascularization of iris (rubeosis iridis): Sign of severe retinopathy. -The Extra-Ocular Muscles & Optic Nerve: -Usually CN III infarct -Pupils usually spared in diabetic CN III palsy.	2 types: i. Proliferative: neovascularization of iris, disc, retina and vitreous → vitreous hemorrhage from bleeding of fragile new vessels → fibrous tissue can contract causing tractional retinal detachment. ii. Non-proliferative: -Venous changes -Intra retinal microvasculare collaterals -Presence of multiple hemorrhages in 4 quadrants. Treatment: 1-Non proliferative: Focal laser + monoclonal antibodies 2- proliferative: Pan retinal photocoagulation
Grave's Disease	Lid retraction: Due to hyper stimulation of the sympathetic nervous system and contraction + fibrosis of levator palpebrae superioris muscle. Exophthalmos: Due to an	
	increase in the bulk of the ocular muscles and the orbital fat Opthalmoplegia: Restrictive myopathy is due to fibrosis of extra-ocular muscles.	

	Retinal and optic nerve changes: Compression of the globe may lead to elevation of intra-ocular pressure Corneal changes: keratitis due to dryness and formation of ulcers in the lower 1/3 of cornea during sleep because the patient can't close his eyes which lead to excessive exposure	
Tuberculosis	-Tuberculosis can mimic any type of eye inflammation -Whenever you see a patient with chronic uveitis → rule out tuberculosis. - It can cause - granulamatous inflammation manifested as keratic preciptates on the cornea, iris nodules, inflammation of the vitreous, systoid macular edema, retinal vasculitis (Eale's disease), choroiditis.	nd -2 most common cause of uveitis is Tuberculosis. - Affect the eye by: 1- Direct infection: choroidal granuloma (appears as pinkish nodules) 2- Type 4 hypersensitivity immune reaction to tuberculin protein causes retinal vasculitis.(whitish) - Tx: anti-tuberculosis drugs +steroids
Syphilis	Important cause of uveitis Ocular features: In congenital syphilis: Interstitial keratitis and Chorioretinitis In acquired syphilis: 1.Ocular chancre 2.Iridocyclitis 3.Interstitial keratitis 4.Chorioretinitis 5.Neuro-ophthalmic manifestations	Any patient with uvietis rule out syphilis→ by doing VDRL test and fluorescent treponemal antibody absorption test (FTA-Abs).
Sarcoidosis	-Sarcoidosis is a multisystem disease characterized by non- caseating granulomatous infiltration of affected tissues. Patients may present with pulmonary, ocular, joint or reticuloendothelial system manifestations. Ocular features: 1) Lid margin and conjuctival granuloma 2) Acute iridocyclitis	The most common cause of uveitis in Japan & Blacks. Tx :corticosteroids

	3) Chronic granulomatous iridocyclitis 4) Peripheral retinal periphlebitis 5) Choroidal granulomas 6) Retinal granulomas 7) Optic nerve granulomas 8) Retinal vasculitis: less aggressive than T.B -Mutton fat keratic precipitate is diagnostic feature -Posterior synechiae	
Rubella	Maternal rubella during the 1st trimester causes serious congenital anomalies. Eyes: small eyes (microphthalmos), cataracts (most common), glaucoma, retinopathy	
Wilson's Disease	-Also called hepato- lenticular degeneration. -Kayser-Fleischer rings (copper deposits in Descemets membrane); more common in patients with CNS involvement, present in 50% if only	
	liver involvement	
Marfan's Syndrome	Ocular features: 1. Lens sublaxation (Ectopia lentis), Angle anomaly, Glaucoma, Hypoplasia of the dilator muscle, Axial myopia, and Retinal detachment	
SLE	Ocular features: -Most important and most common is retinopathy that occurs only during active disease, "flare-ups" and affects the cornea causing corneal melting. - Cotton wool spots and large areas of infarction and ischemia in the peripheries (signs of arteriolar occlusion), intra retinal hemorrhage.	Diagnosis: elevated ESR, anti-nuclear antibodies (ANA), anti-double- stranded DNA antibodies, anti-phospholipid antibodies and Low complements 3 and 4.
Rheumatoid Arthritis	Ocular features: 1. Keratoconjuctivitis sicca 2. Scleritis, lead to melting of the sclera 3. Keratitis 4. Melting of the cornea - Dryness of the cornea and conjunctiva because it destroys the accessory lacrimal glands and it's diagnosed by	

	Rose Bengal staining. - The scleritis may lead to scleromalacia perforans (thinning of sclera)	
Ankylosing Spondylitis	Presentation could be a young man with acute recurrent non- granulomatous anterior uveitis.	Investigations: HLA B27 detection. x-ray of sacroiliac joints
Juvenile idiopathic Arthritis	Mostly causes anterior uveitis. Types: Systemic "Still's disease, Polyarticular, and Pauciarticular (oligoarticular).	Risk factors of eye involvement: 1.Girls are more prone than boys (x3). 2. before the age of 4 3. Associated with
		+(ANA) 4. Oligoarticular type.
Behchet's Disease	- Is a very aggressive type of eye inflammation and a blinding disease if not treated; it causes severe occlusive retinal vasculitis. -Anterior uveitis, occasionally with hypopyon (present in about 30% of patients with Behchet's Disease).	-HLA B51 association -Common in areas of the "Ancient Silk Road' - Diagnosed clinically: recurrent oral ulcers (aphthous ulcers) and 2 of the following: 1-Recurrent Genital ulcers 2-Skin lesions 3-Anterior uveitis, occasionally with hypopyon 4- Positive Behcet's test -Tx: Corticosteroids combined with immunosuppressive therapy. If no response, infliximab (anti α-TNF).
Sjogren's Syndrome	Ocular features: keratoconjuctivitis sicca : dryness of the conjuctivia	
Vogt-Koyanagi-Harada Syndrome	-Disease of Pigmented individuals (common here in the Kingdom) - is a multisystem disorder presenting with a bilateral panuveitis with exudative retinal detachments and followed by neurological and cutaneous manifestations such as baldness and loss of lash and skin pigments (poliosis and vitiligo). - The inflammation could be granulamatous leading to Mutton fat keratic precipitates (due to anterior uveitis).	Most common cause of eye inflammation in our country. Tx: Should be very early in the acute phase by large doses of systemic steroids for at least 1 year + immunosuppressive therapy like mycophenolate mofetil (Cellcept) and cyclosporin, to prevent permanent visual loss.
Giant-Cell Arteritis	-Autoimmune vasculitis of large and medium sized arteries -sudden loss of vision due to ischemic anterior- posterior optic neuropathy	- One of the top emergencies in ophthalmology

		- investigations: Erythrocyte Sedimentation
	- Disc will be pale and swollen. -Other symptoms: Pain over the temporal artery ,Jaw claudication, Scalp tenderness (gangrene of the scalp may occur), Polymyalgia rheumatica (muscular pain, esp. neck, shoulders and hips) and Constitutional symptoms	Rate (ESR: very high), C reactive protein (CRP: high), temporal artery biopsy (gold standard). Tx: immediately and urgently with large doses of systemic steroids to prevent visual loss in the other eye.
Toxoplasmosis	-Typically causes necrotizing retinitis -If the mother is infected during 3rd trimester, the baby will present with congenintal toxoplasmosis with: 1 .Convulsions 2. Chorioretinitis 3. Intracranial calcifications Acquired: Either reactivation of old infection or new infection.	-Common in Brazil -Most common cause of infectious uveitis. Tx: It's sensitive to many antibiotics: clindamycin, azithromycin, minocycline sulfonamides, cotrimoxazole pyrimethamine (Daraprim) prophylactic treatment in pt. with recurrent attacks.
Hypertensive Retinopathy	Ocular features: Stage 1: Attenuation of arteries (↓ diameter) and increase light reflex , sometimes referred to as "copper/silver wiring" Stage 2: as grade 1 + Nipping of the retinal veins where they are crossed by arterioles Stage 3: as grade 2+ Narrowing of arterioles → breakdown of blood-retina barrier: hard exudates, hemorrhages, macular star and retinal edema Stage 4: Optic disc edema (bilateral = papilledema) → blurring of vision and temporary visual loss	

Done By:

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