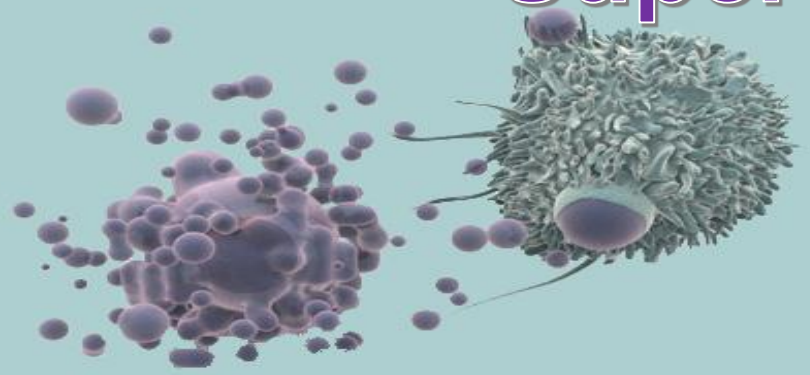
Challenges
makes you more
responsible. Always
remember that life
without struggle is
a life without success.
Don't give up and learn
not to quit.

Good Morning.

Corneal immunological disorders

Supervised by Dr. Buraa kubaisi



Presented by Dr. Alaa Arrat


KEEP YOUR
EYES OPEN

Hypersensitivity

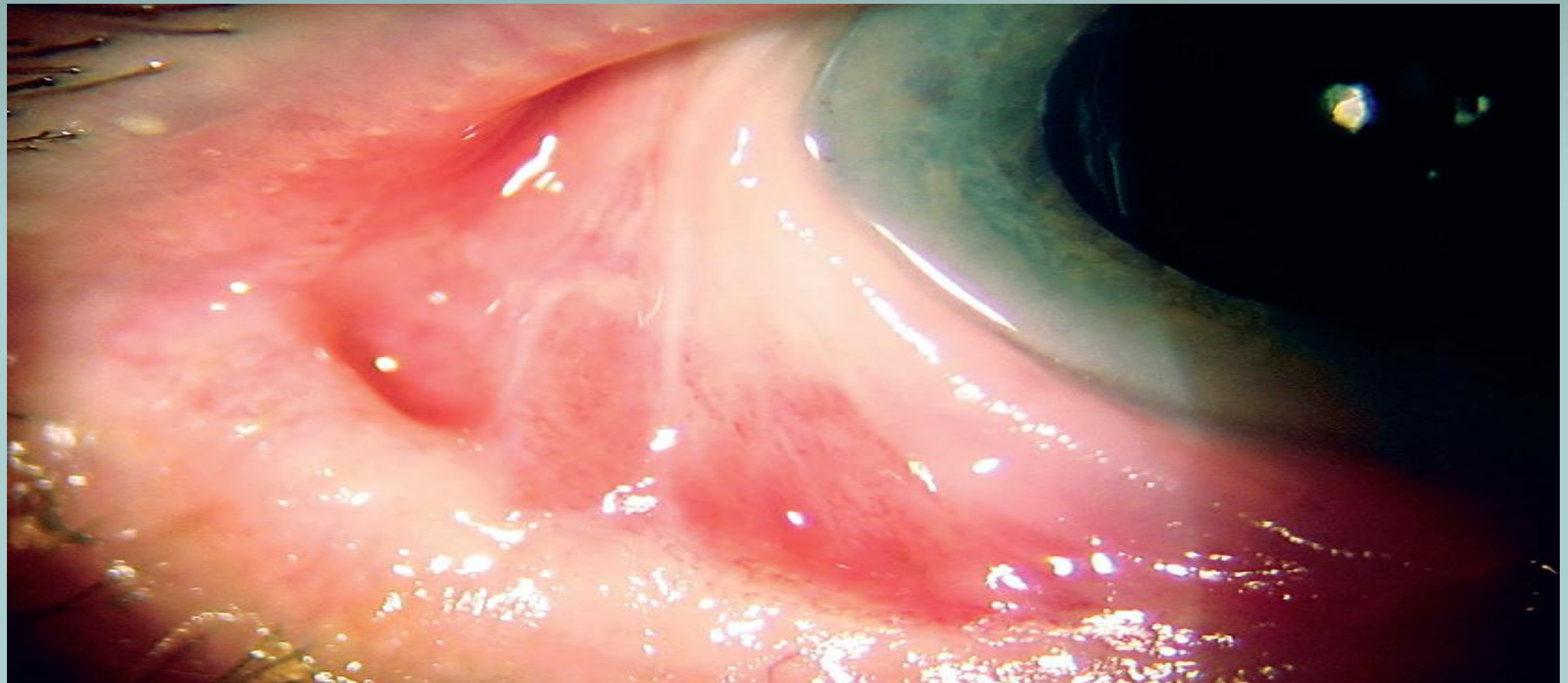
Type	Mechanism	Ocular surface disease
I	IgE cross-links receptors on mast cells → degranulation → chemical mediators	VKC
II	Antigen-antibody dependent cytotoxicity IgM or IgG activates complement causing tissue damage- could be isoimmune or autoimmune	Ocular cicatricial pemphigoid
III	Persistent antigen antibody complex formation→ complement activation-influx of polymorphs- platelet aggregation	PUK & Mooren. Steven-Johnson syndrome
IV	Exaggerated T cell-mediated immune response	VKC AKC Staph hypersensitivity disorders IK & Cogan Drug allergy Corneal transplant rejection

CASE:

A 72-year-old woman presents to your office with a report of red eyes and foreign body sensation for several months.

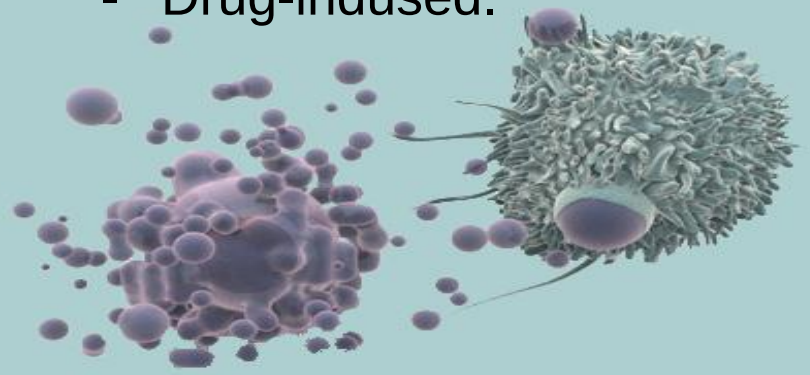
She also recently notes the onset of dysphagia.

Her slit-lamp appearance is shown



D.D OF SYMBLEPHARON

- chemical burn.
- Stevens-Johnson syndrome (SJS)
- Ocular cicatricial pemphigoid &pseudopemphigoid.
- Trachoma.
- herpes zoster, adeno.
- atopic keratoconjunctivitis.
- Scleroderma.
- graft versus host disease.
- Drug-indused.

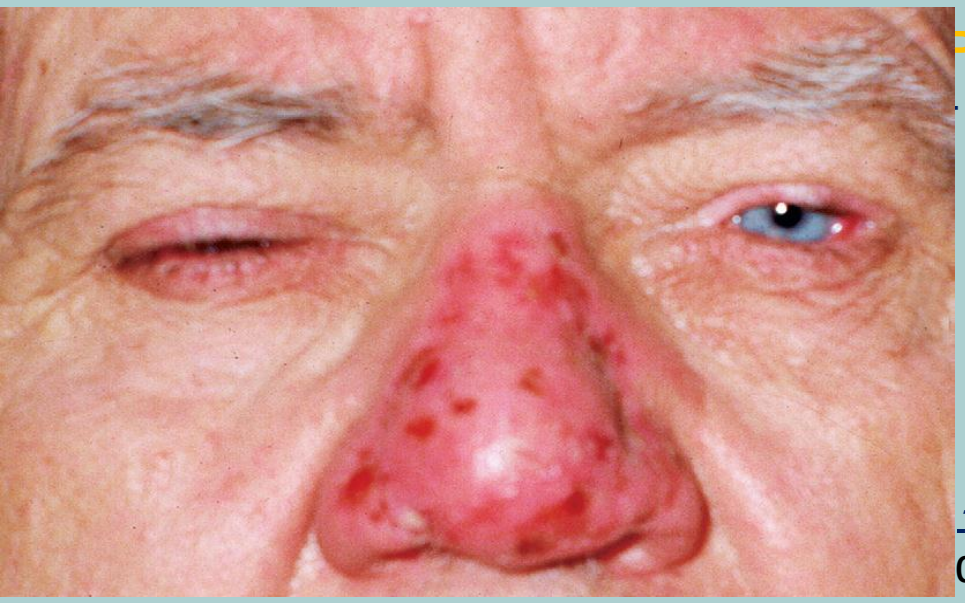


Ocular cicatricial pemphigoid

Definition:

- Mucus membrane pemphigoid is a systemic autoimmune disease characterized by chronic blistering and scarring of the skin and mucus membranes.
- If it affects the eye (40% of cases) it is termed ocular cicatricial pemphigoid (OCP).
- It is a slowly progressive, bilateral (can be asymmetric), and potentially blinding condition.
- other mucous membranes frequently involved, including oral (up to 90%), esophageal, tracheal, and genital; skin involved in up to 30% of cases.

- usually occurs in females (2 : 1) over 60 years old.



Pathophysiology:

- *autoimmune mechanism* cytotoxic (type II) hypersensitivity reaction with autoantibodies directed against antigens of the basement membrane complex. The conjunctival inflammation results in subepithelial scarring, destruction of conjunctival goblet cells, and obstruction of lacrimal ducts. Resulting in mucus and aqueous tear deficiency and keratinization of the conjunctiva and corneal epithelium.

- *drug induced* (epinephrine, timolol, pilocarpine, echothiophate iodide, or doxuridine).pseudopemphigoid..

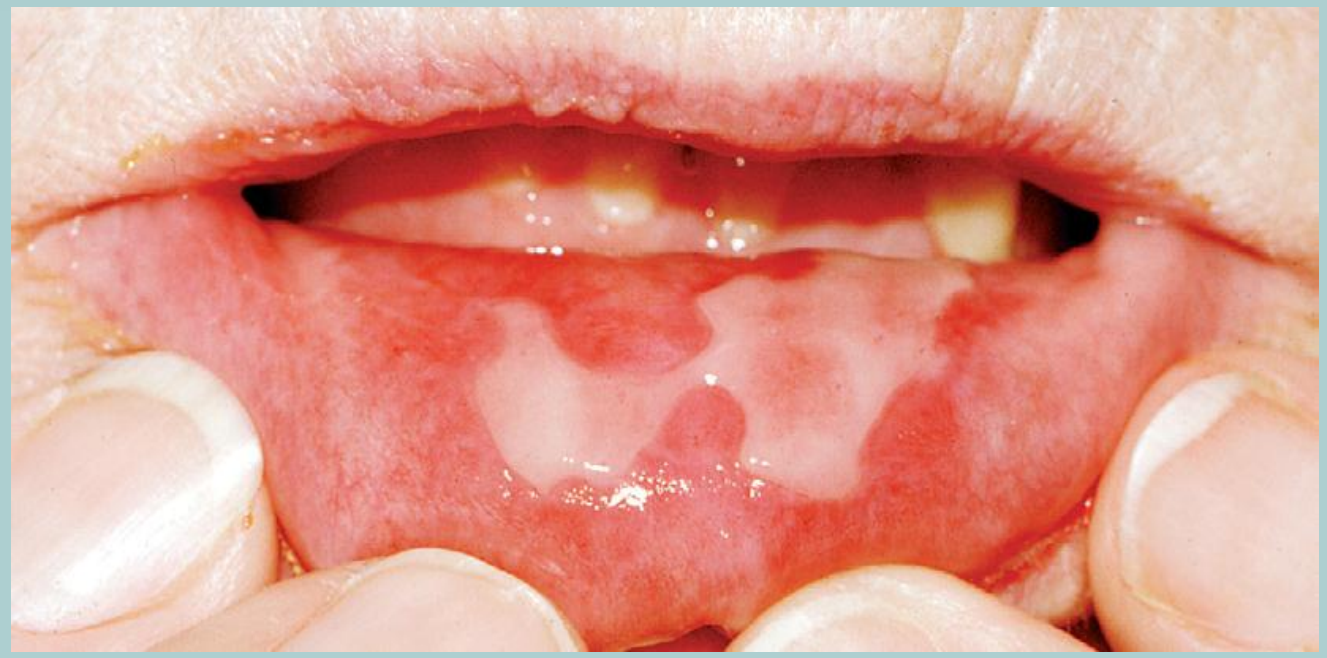
History

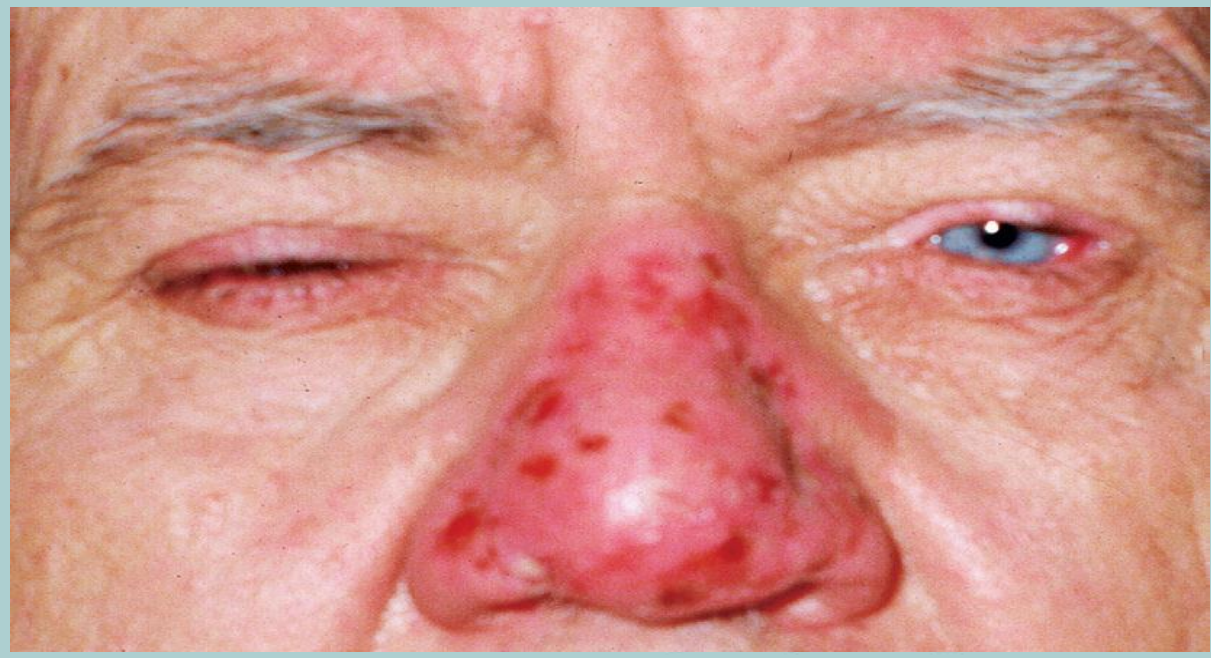
- The course of disease can be **chronic** and characterized by remissions and exacerbations.
- Onset is with **redness, foreign body sensation, watering, and photophobia**.

Misdiagnosis as blepharitis or allergic eye disease often delays appropriate treatment.
Progressive disease results in decreased vision.
- **symptoms of systemic disease** (e.g. dysphagia).

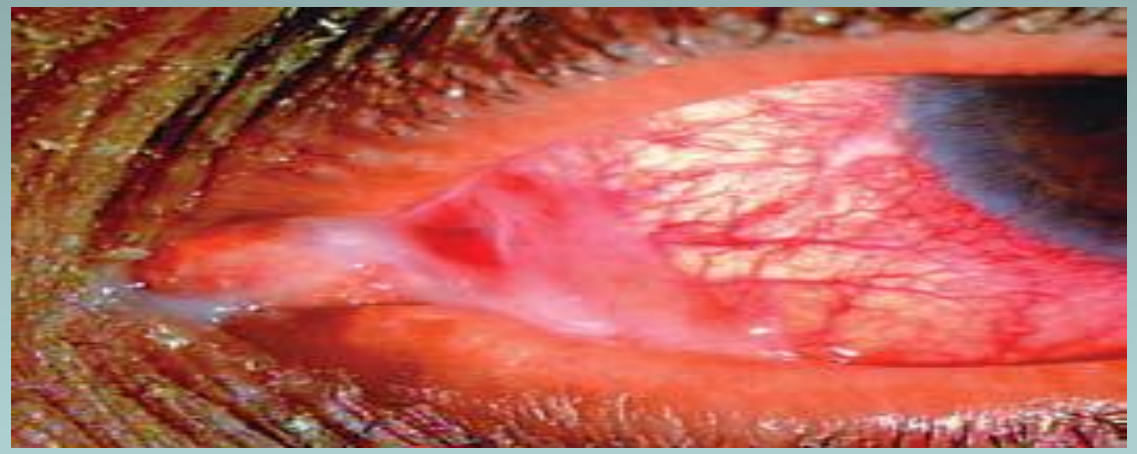
Examination

The progression of disease can be categorized into four stages,as follows:

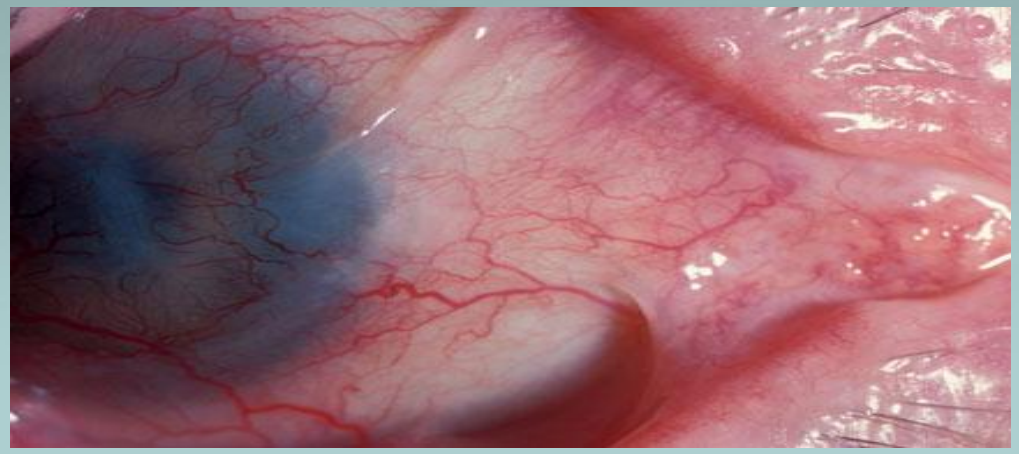




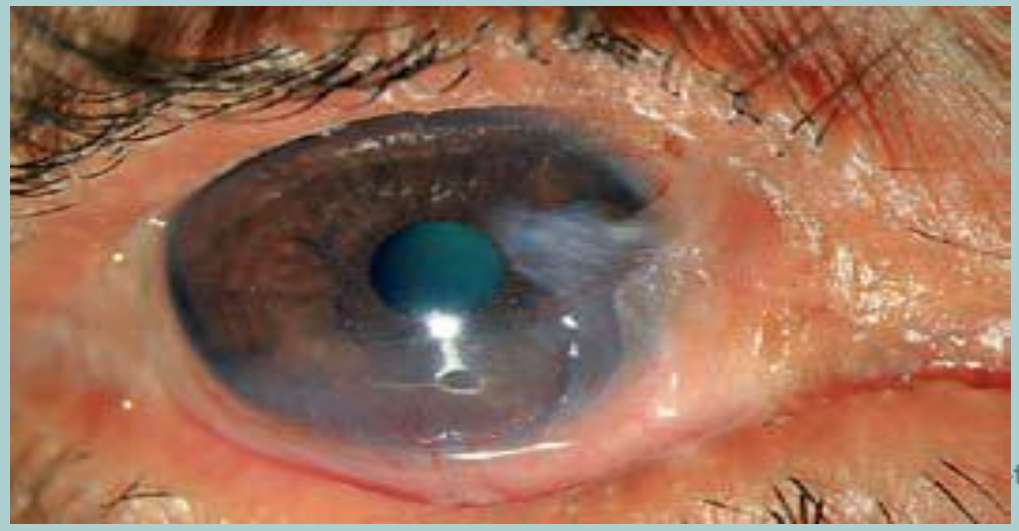
Stage I: chronic conjunctivitis with subepithelial fibrosis. Loss of the plica and flattening of the caruncle is an early sign.



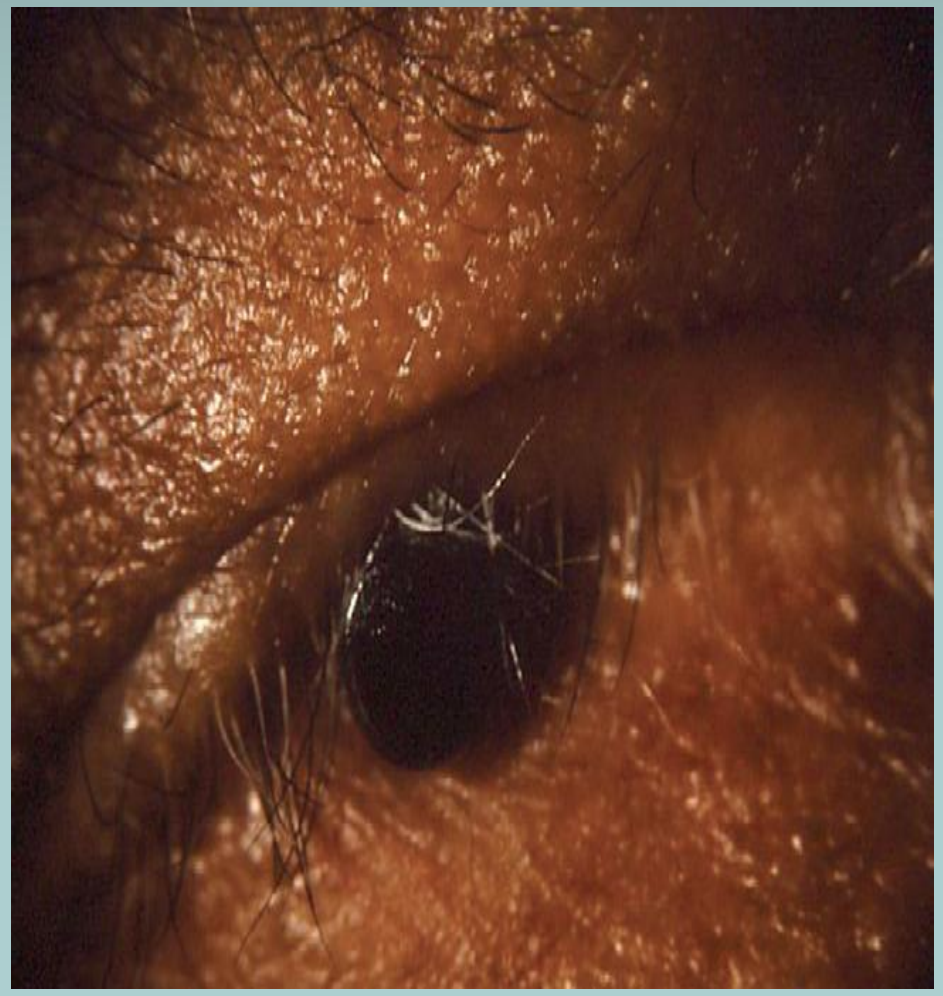
Stage III: symblepharon formation.

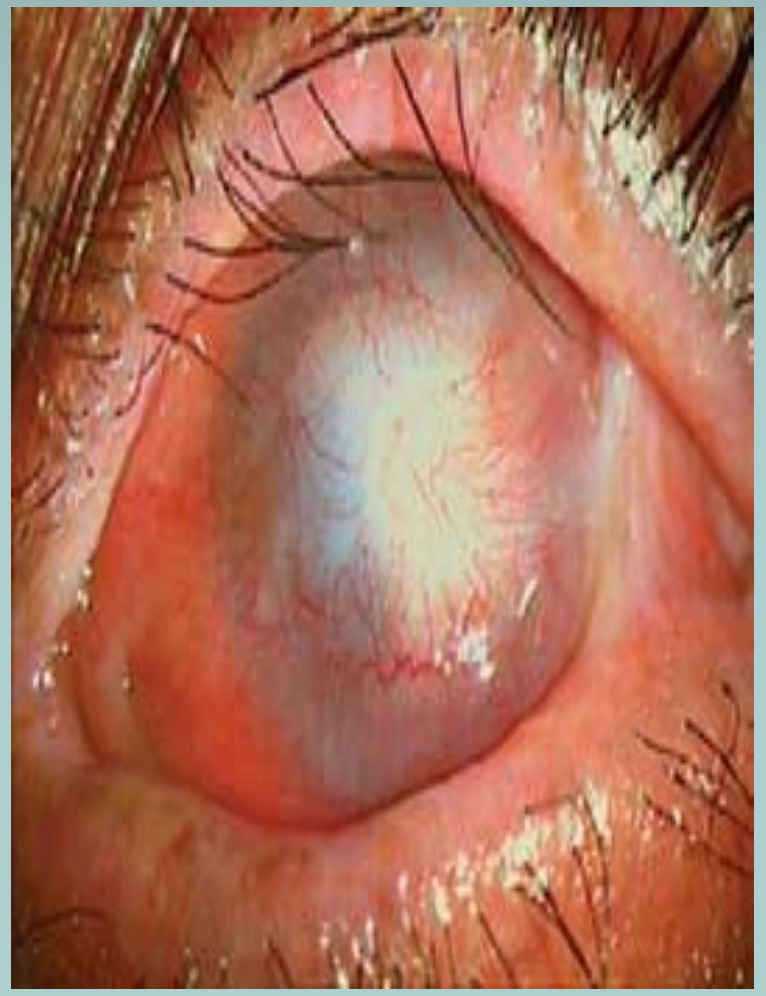


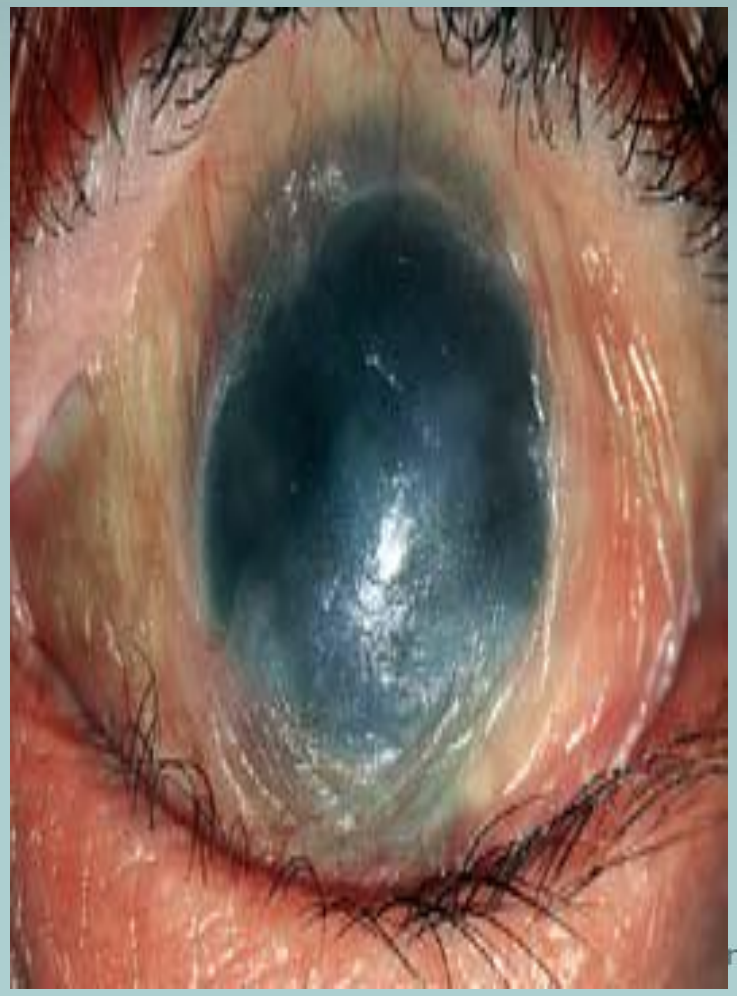
Stage II: cicatrization with shortening of the inferior fornix (depth <8 mm).



Stage IV: ankyloblepharon: dense adhesions to the lid margin,
limiting eye movement (**frozen globe**).
Severe keratopathy develops due to eyelid/lash malposition, aberrant eyelash growth and cicatricial entropion, dry eye, exposure and limbal stem cell deficiency. Persistent epithelial defects, ulceration, and neovascularization

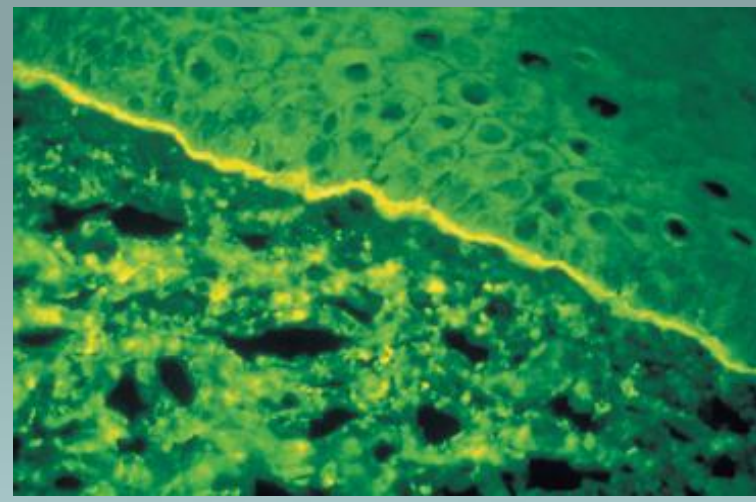




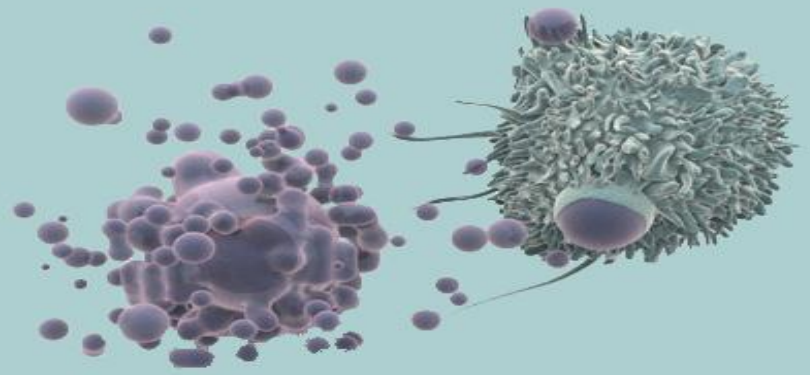


Investigations:

To support the clinical diagnosis, a **conjunctival biopsy** from an area of active inflammation(not symblepharon) should be examined for immunofluorescent antibody labelling of immunoreactants (IgG, IgA) deposited in a linear pattern at the basement membrane zone.



BACK TO THE CASE



All of the following statements are **true *except***:

A) She should be asked about use of topical medications.

B) Conjunctival scrapings may reveal eosinophils.

C) Corneal scarring and vascularization are successfully treated with penetrating keratoplasty.

D) Immunoglobulins are deposited along the conjunctival basement membrane.

Which **treatment** would be ***most* effective** for this condition?

A) Dapsone

B) Topical steroid drops

C) Lysis of membranes

D) Topical cyclosporine

Treatment

- Long-term **systemic treatment is** the mainstay.
- **Dapsone** is the front-line agent for **mild to moderate inflammation** (contraindicated in (G6PD) deficiency).
- **Alternatives** to dapsone include mycophenolate, or azathioprine.
- **For severe inflammation** (conjunctival necrosis, limbitis) cyclophosphamide is used.

High-dose systemic corticosteroid is very effective but only used for short periods of time for control of severe disease.

Adjuvant therapy to the above includes:
treatment for dry eye (tear-film supplements, lubricants, punctual occlusion (if not already stenosed from scarring));
topical steroids can be used for **maintenance** of mild disease or for acute exacerbations.

Surgery in OCP??

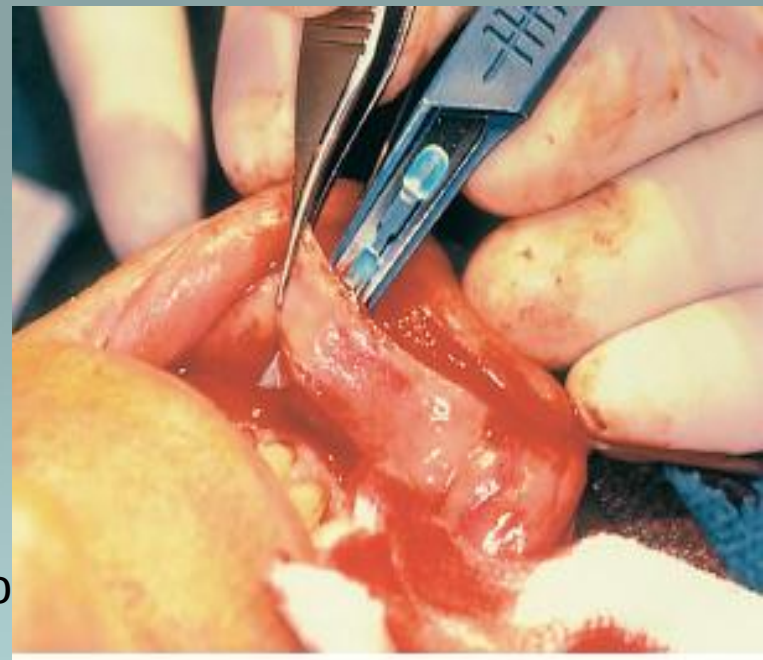
surgery often initiates exacerbations

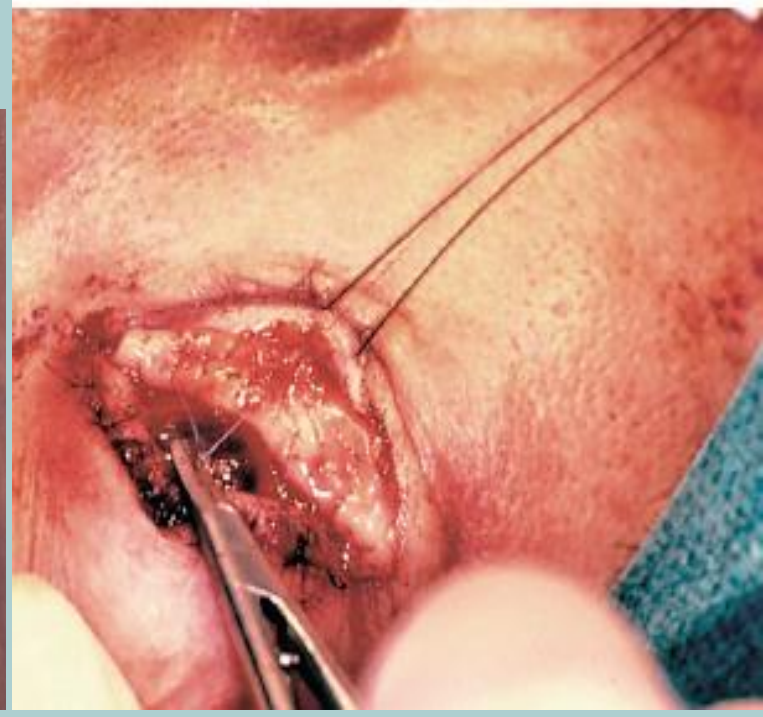
Surgery may be required in the following cases:

- Cryotherapy or electrolysis of trichiasis.
- Correction of entropion.

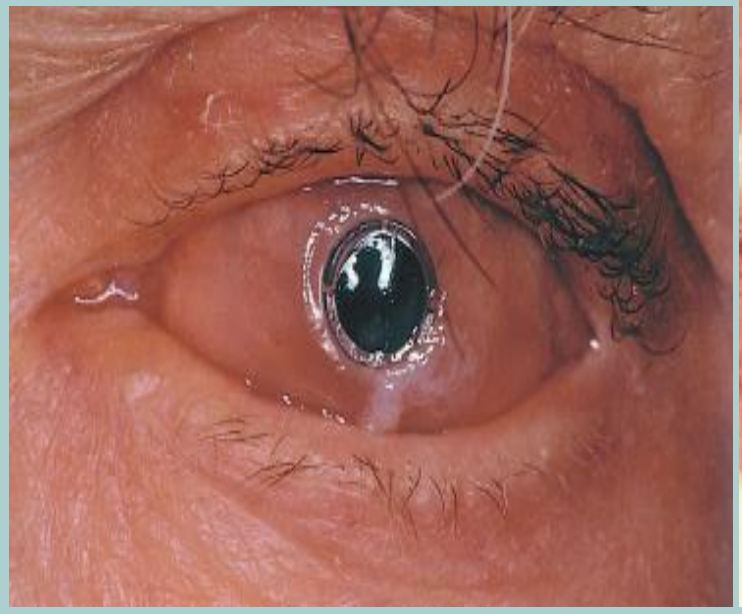
Mucous membrane grafts (e.g., buccal or amniotic membrane graft) can be used to reconstruct the fornices if needed

- penetrating keratoplasty has an extremely guarded prognosis .
- A keratoprosthesis (artificial cornea) may be the only option to restore vision.





Prognosis Poor



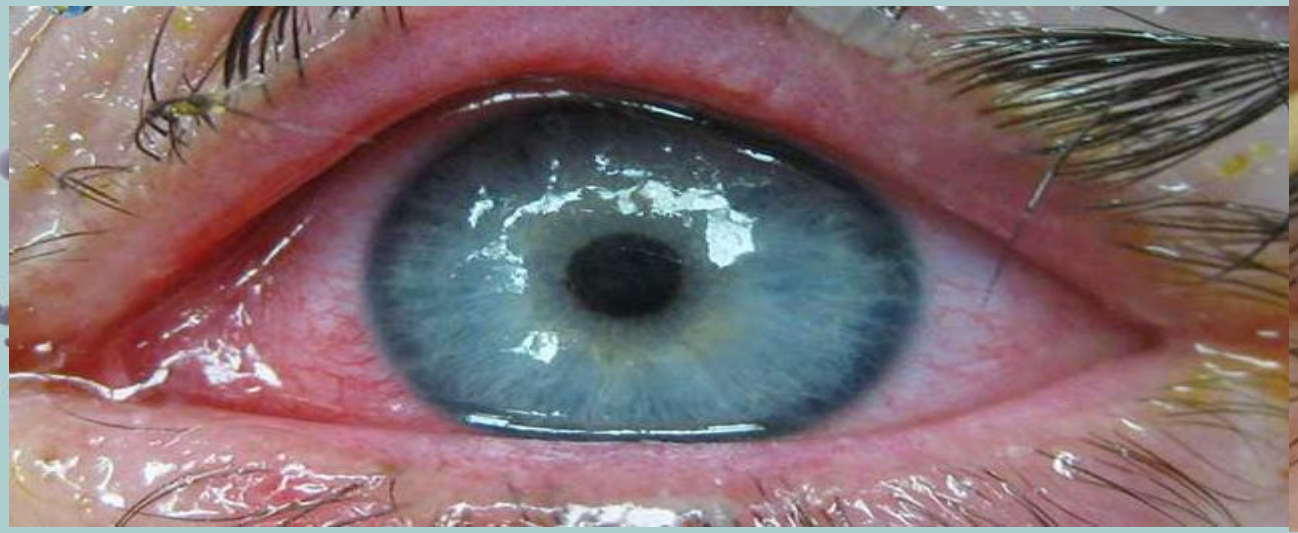
Follow-Up

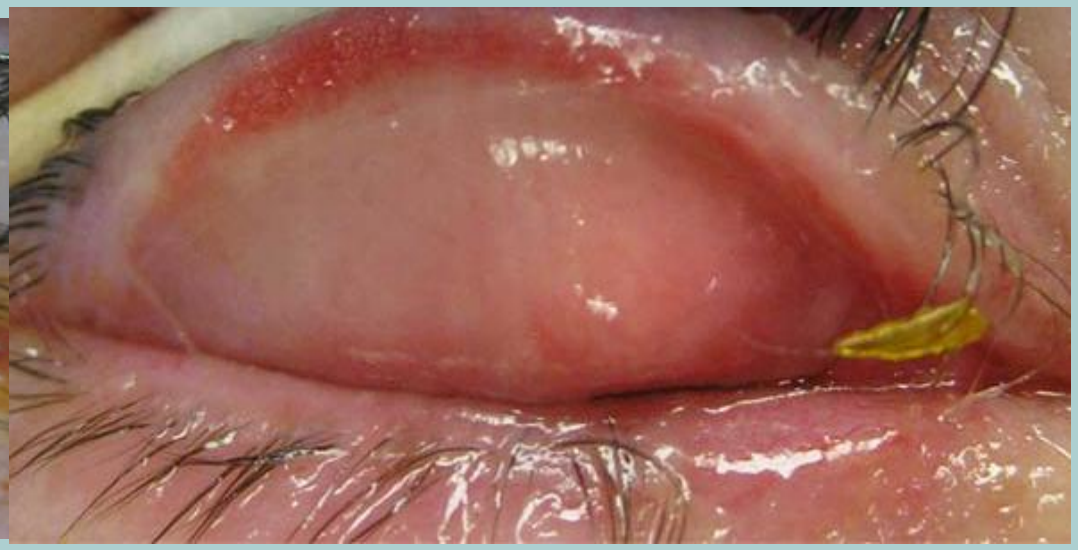
Every 1 to 2 weeks during acute exacerbations, and every 1 to 6 months during remissions.

case

A 13-year-old female was prescribed trimethoprim-sulfamethoxazole by her primary care provider for a lower extremity infection. In the following days, she developed a fever of 103° F, conjunctival injection, and an erythematous rash involving >60% body surface area with bullae involving <10% body surface area. The antibiotic medication was immediately discontinued, but her symptoms continued to worsen.

She was subsequently admitted to the pediatric critical care unit, and an ophthalmic consult was requested.





Stevens–Johnson syndrome

- Definition:

potentially fatal acute mucocutaneous disorder. The most severe manifestation of this disease is toxic epidermal necrolysis.

SJS is defined as <10% total body surface area, SJS-TEN overlap as 10-30%, and TEN as >30%.

- Pathophysiology:

type III hypersensitivity reaction with immune-complex deposition in the skin and mucus membranes.

- Etiology:

The most common known etiological factors include:

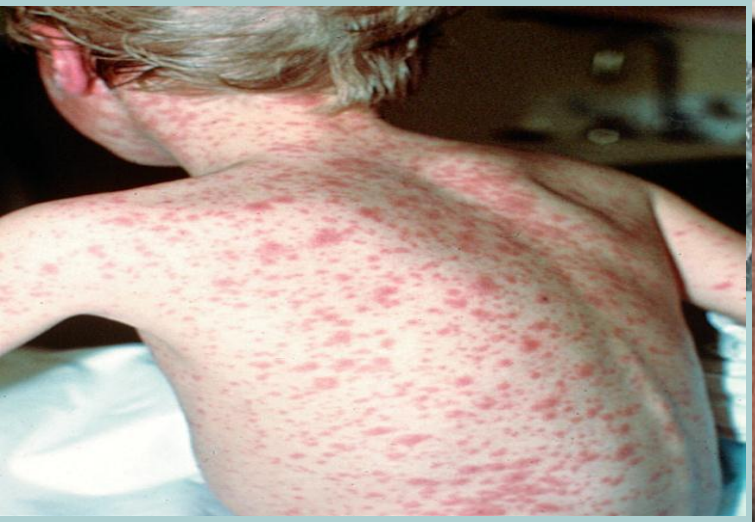
- drug hypersensitivities **80%(**sulfonamides, penicillin, phenytoin).
- viral infections (herpes simplex, AIDS, influenza),
- malignancies

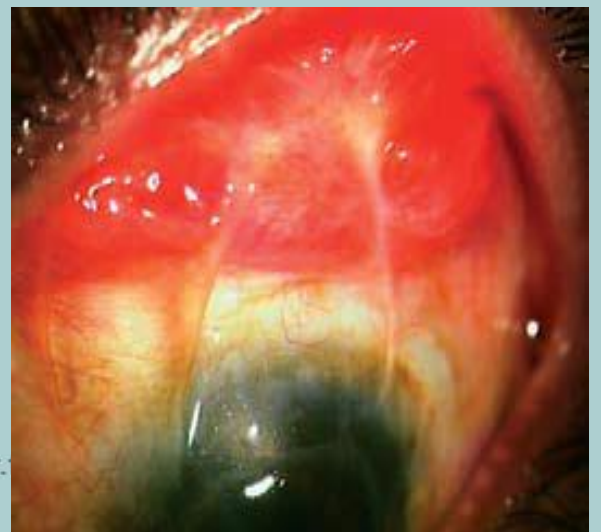
The precipitating cause is not identified in up to half of cases.

Clinical evaluation

- Prodromal phase 1–14 day upper respiratory-tract symptoms. Mucocutaneous lesions develop abruptly. Skin lesions are classically erythematous maculopapules described as ‘target lesions’.
- ocular acute phase lasts 2–3 weeks and consists most commonly of a transient self-limiting bilateral membranous conjunctivitis. Occasionally there may be more severe conjunctival involvement with bullae, necrosis, and membrane formation.
  There may be secondary ocular surface infection.
- Long-term ocular surface scarring may occur depending on the severity of the disease. The features of OCP stages II–IV may all occur but these are **usually non-progressive** after the acute event.

Ocular surface disease can lead to progressive corneal scarring and vascularization.



Diagnosis is clinical.

Treatment

The mainstay of treatment is supportive. systemic immunosuppresion for acute disease is best administered by an intensive care physician/ dermatologist.

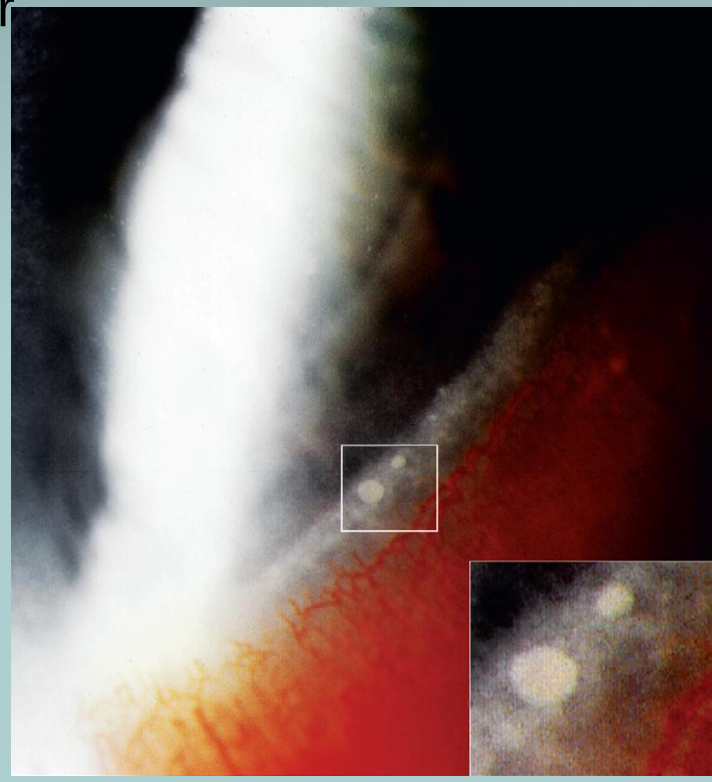
Ocular treatment involves intensive use of artificial tears and lubricants, prophylactic topical antibiotics, and possibly topical steroids. Any associated long-term dry eye, lash/lid abnormality are managed as for OCP.

Sytemic immunosuppression is only indicated if there are signs of **recurrent** conjunctival inflammation..

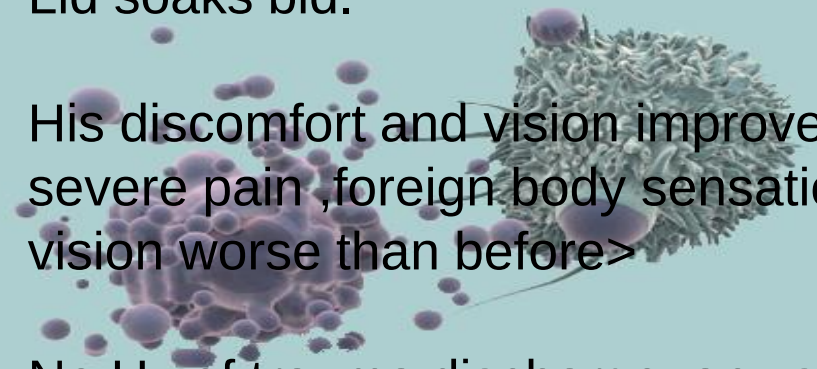
Prognosis Fair; not progressive (in contrast to ocular cicatricial pemphigoid), recurrences are rare, but up to 30% mortality

case:

A **61** years old man with a 16 years of **RA** taking prednisone came to your office.
He previously had discomfort, erythema and blurry vision for 6 days.
His general ophthalmologist treated him for severe staphylococcal blepharitis.



He was taking:
Bacitracin oint bid
Ofloxacin e.d every hour.
Lid soaks bid.



His discomfort and vision improved slightly but after few days he came severe pain ,foreign body sensation , photophobia, tearing and blurred vision worse than before>

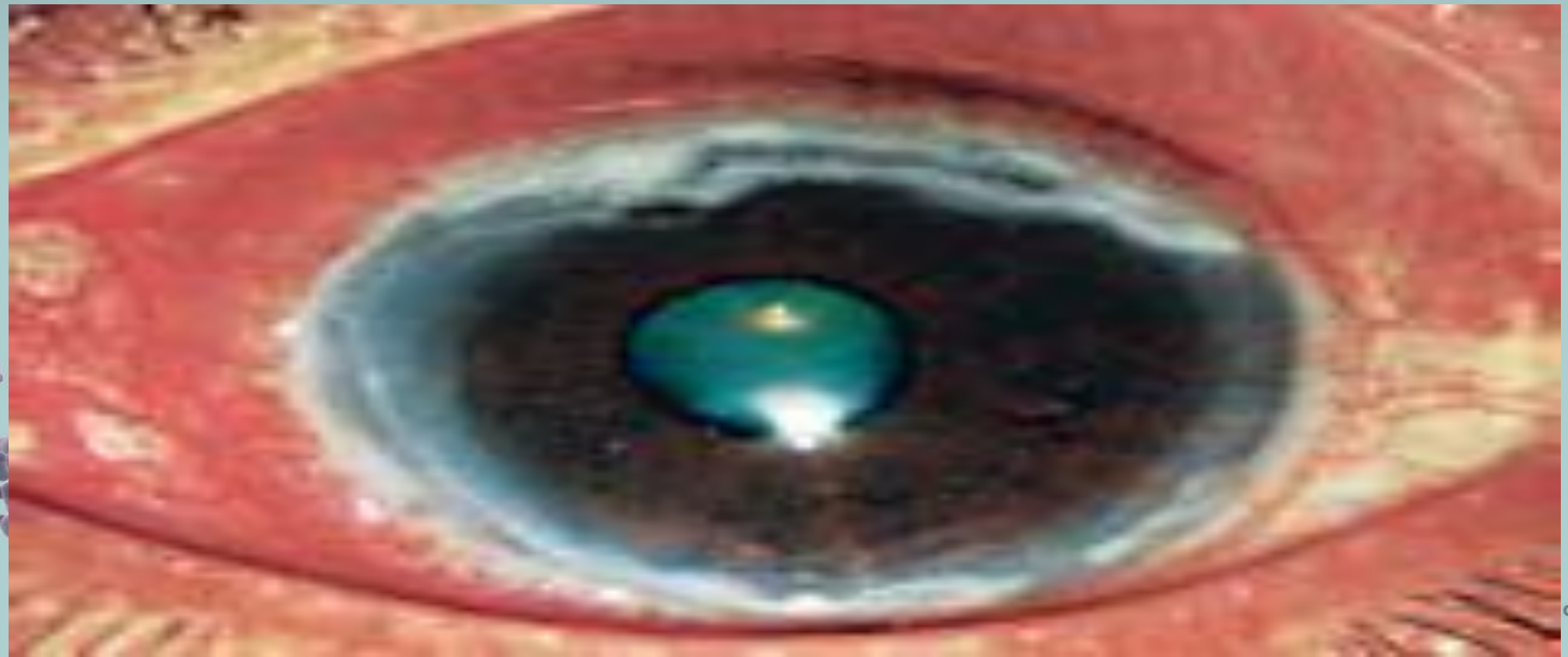
No Hx of trauma discharge, any sick contact or URI.
No known drug allergy.

Ophthalmic examination:

BCVA: 0.5 OD, 1.0 OS

The lid and lashes were significant for severe lid crusting with telangiectasia. The conjunctiva was injected +2.

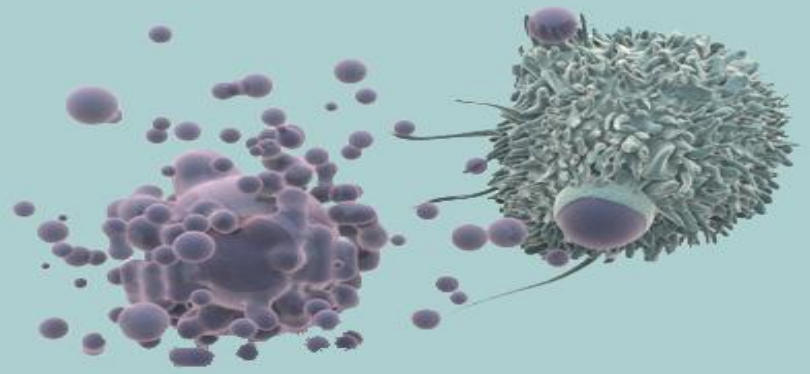
The right cornea showed a circumferential stromal opacity . The limbal vessels were injected and dilated with an epithelial defect and melting.



Differential Diagnosis

- **autoimmune**:
  Peripheral ulcerative keratopathy.
  Mooren ulcer.
  Staphylococcal marginal keratitis.

- **Infectious**:
  Herpetic keratitis.
  acanthamoeba



Peripheral ulcerative keratitis

Definition: uncommon potentially blinding disorder consisting of crescent-shaped destructive inflammation of the peripheral corneal stroma, with an epithelial defect.

What is peripheral cornea?

an arbitrary central limit beginning around 3.5–4.5 mm from the visual axis and extending out to the junction of the ill-defined transition between limbus and the sclera and conjunctiva.

Distinct anatomic differences of the peripheral cornea include:

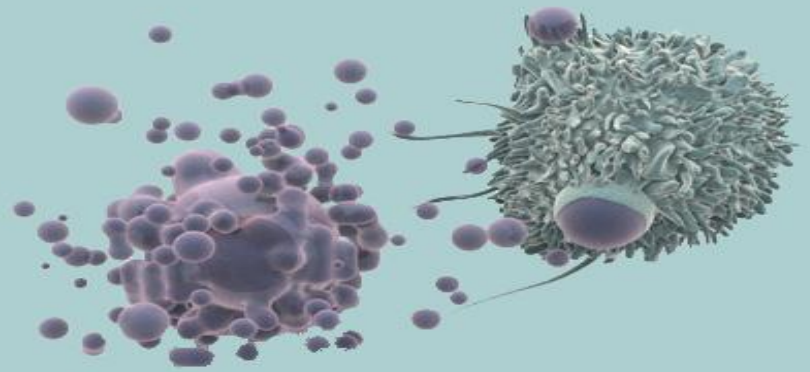
- the greater thickness (up to 0.7 mm) with tight collagen bundle packing; a vascular arcade that originates from the anterior ciliary arteries and extends approximately 0.5 mm into the clear cornea (providing the nutritional supply but also access to the efferent arm of the immune response and the accompanying lymphatics , which drain to the regional lymph nodes.

immunologic characteristics:

- More Langerhans' cells , IgM and C1 (important in the activation of the classic complement pathway), likely due to limited central diffusion from the limbal vessels of the latter two molecules due to their large size.
- Additionally, the adjacent conjunctival blood vessels and lymphatics are source of inflammatory effector cells and cytokines involved in the production of collagenase and proteoglycanase, which may contribute to stromal thinning and eventually perforation.

Etiology

- ***Majority- local and systemic immunological processes(50% CVD) (34% RA)***
- ***Few cases – infectious & neoplastic conditions***



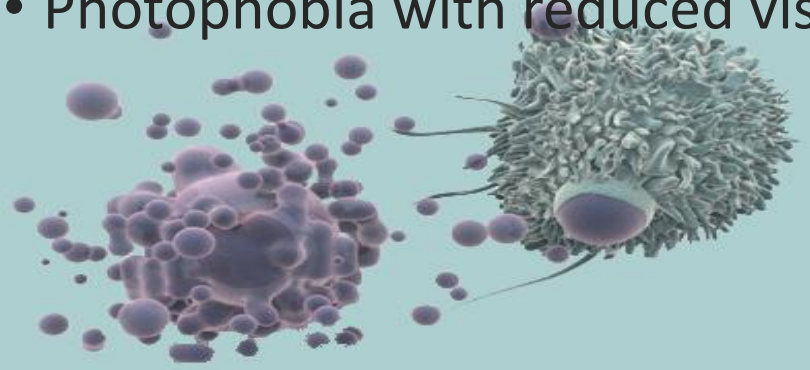
OCULAR MANIFESTATIONS:

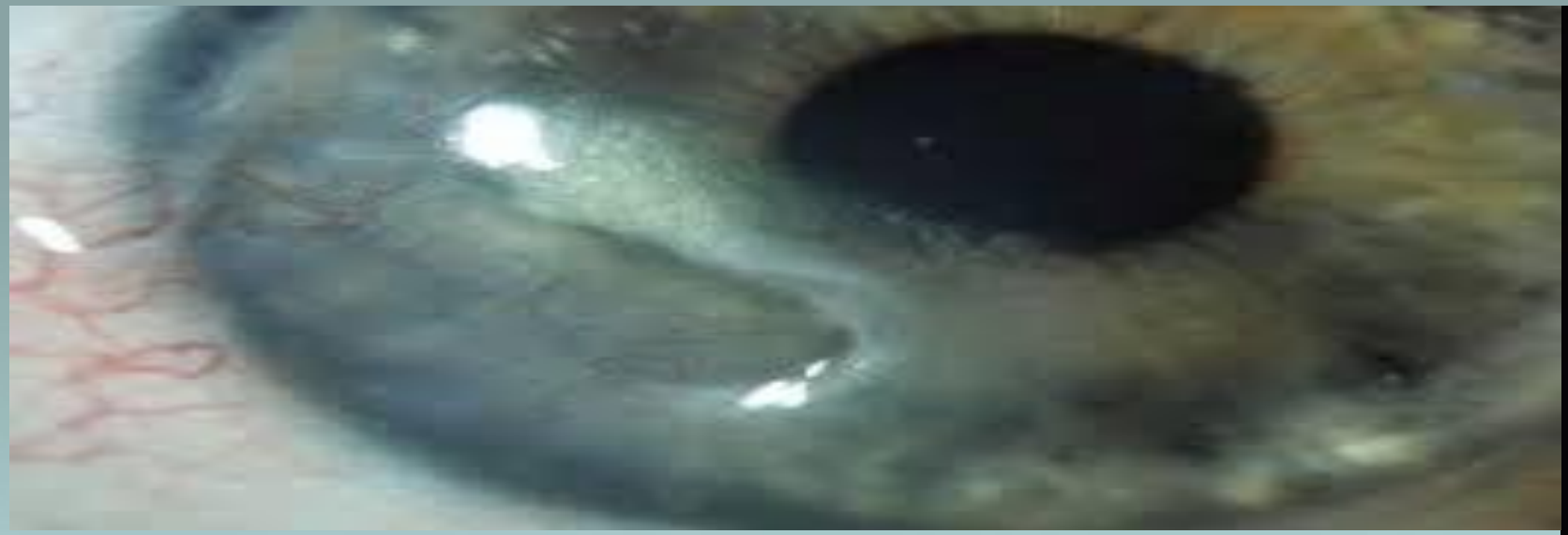
 Presenting symptoms of PUK -**not specific**.

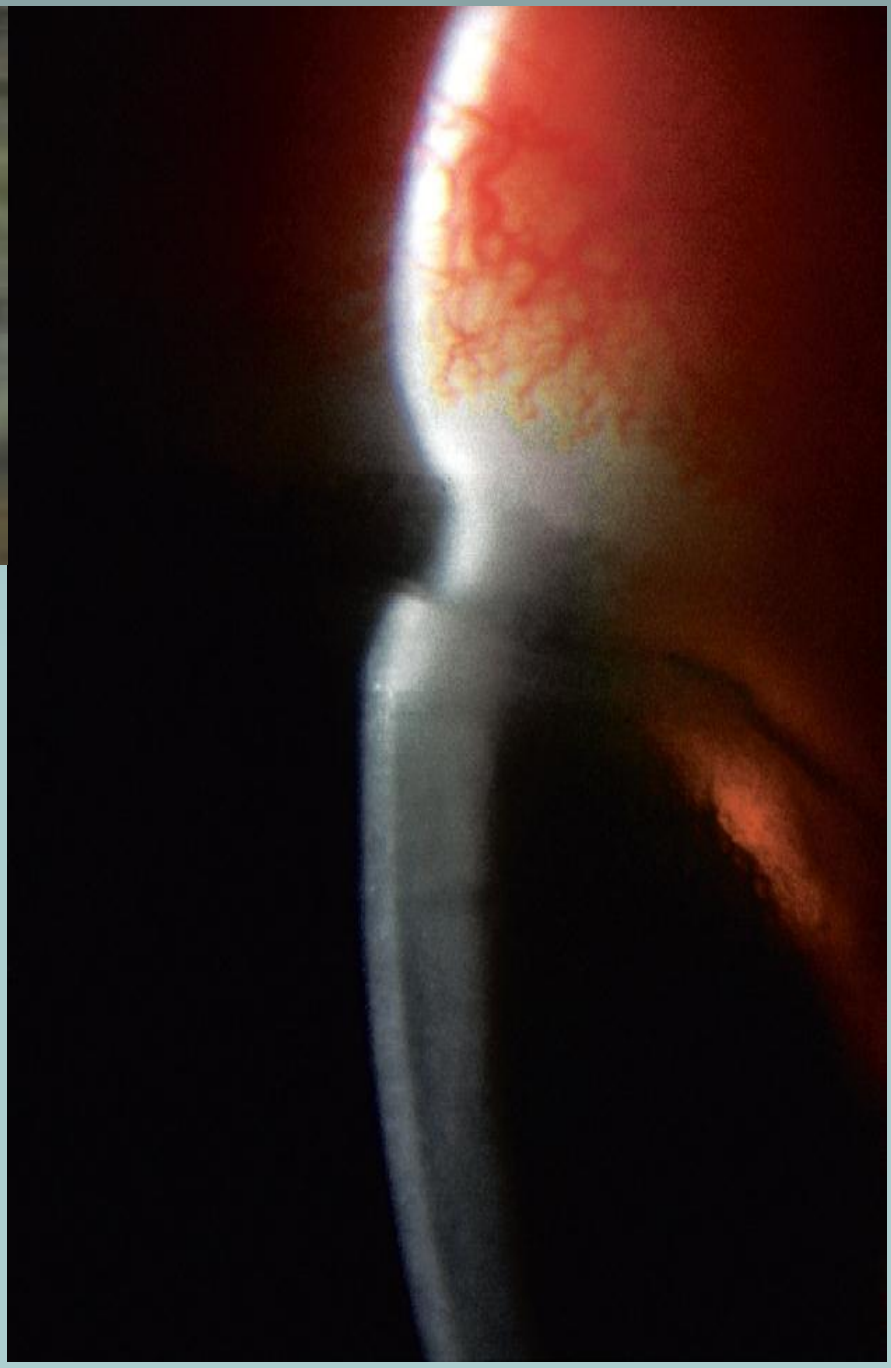
- Foreign body sensation, watering, pain, and photophobia –
- Pain is severe - associated ***scleritis*** (RA, WG, PAN, RP)

Pain without scleral involvement- ***Mooren's Ulcer***

- Decreased Vision - inflammatory process proceeds centrally.
- Photophobia with reduced visual acuity - ***Associated anterior uveitis***







Ocular Examination:

Examination of **lids and conj**:
Blepharitis , Telengiectasis (rosaecae)
varying degrees of adjacent conjunctival injection.
Limbal ischaemia due to vasculitic vaso-occlusion may also be present.
The **cornea**: as a crescent-shaped epithelial defect with stromal thinning and infiltrate,
often limited to one quadrant of the juxtalimbal cornea. The disease may extend to involve the paralimbal sclera.
AC: calm and quiet or may be with reaction
Posterior segment: signs of Posterior scleritis, Vasculitis of CVD.

SYSTEMIC EXAMINATION

- Thorough systemic history & examination mandatory
- Important Questionnaire?

Weight loss, fatigue Skin – facial rashes, ulcers, (SLE)

- Respiratory symptoms (WG, SLE)
- GI symptoms- pain diarrhoea (SLE, WG)
- Musculoskeletal symptoms- joint pain (RA, SLE)
- Neurological – seizures, Raynauds (WG, RP, SLE)
- Genitourinary- hematuria (PAN, SLE)
- Swollen ear lobes (RP, SLE)
- Deafness (WG)
- Nasal ulcers/ bleeds (WG)
- Saddle nose (WG, RP)

Clinical course of the ulcer

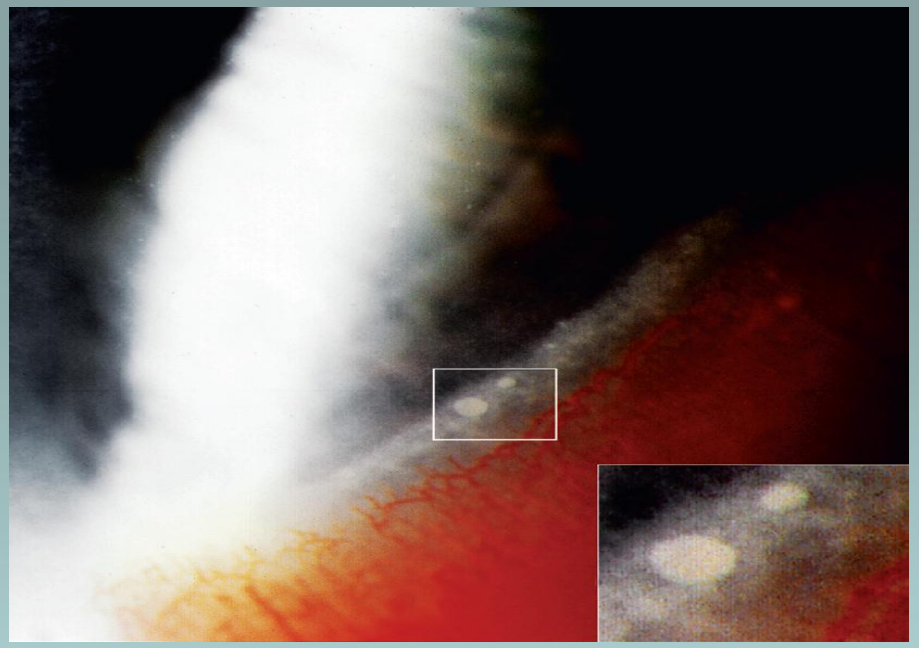
Stromal infiltration of peripheral cornea

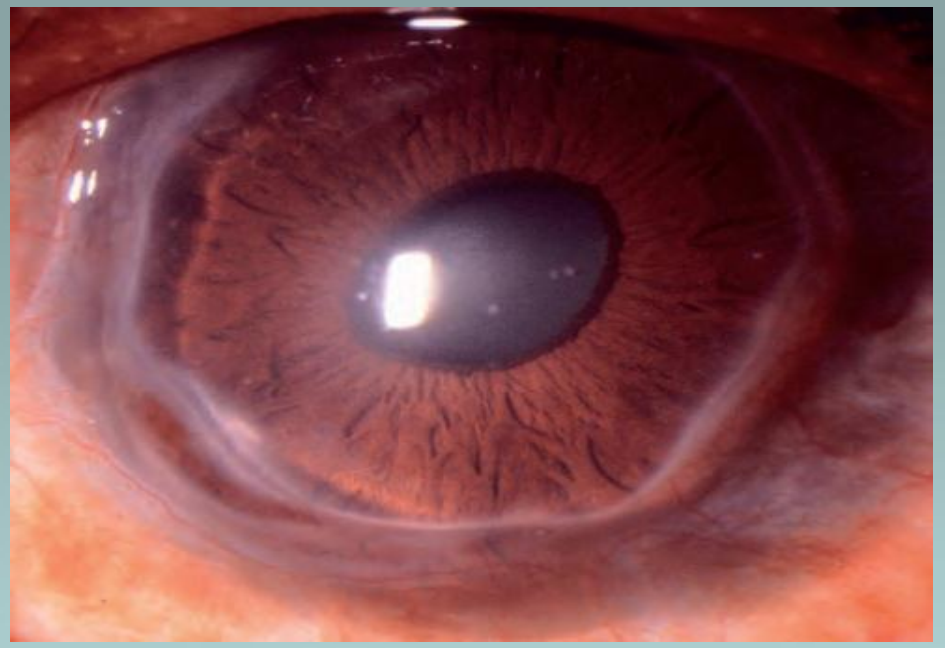
Superficial ulceration

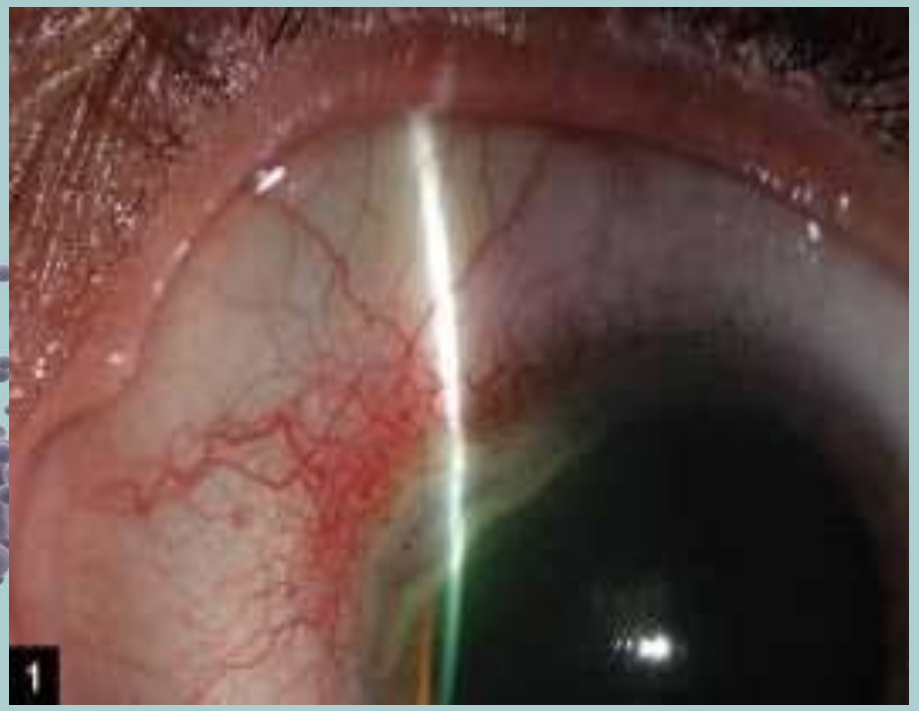
Peripheral spreading and deeper ulceration

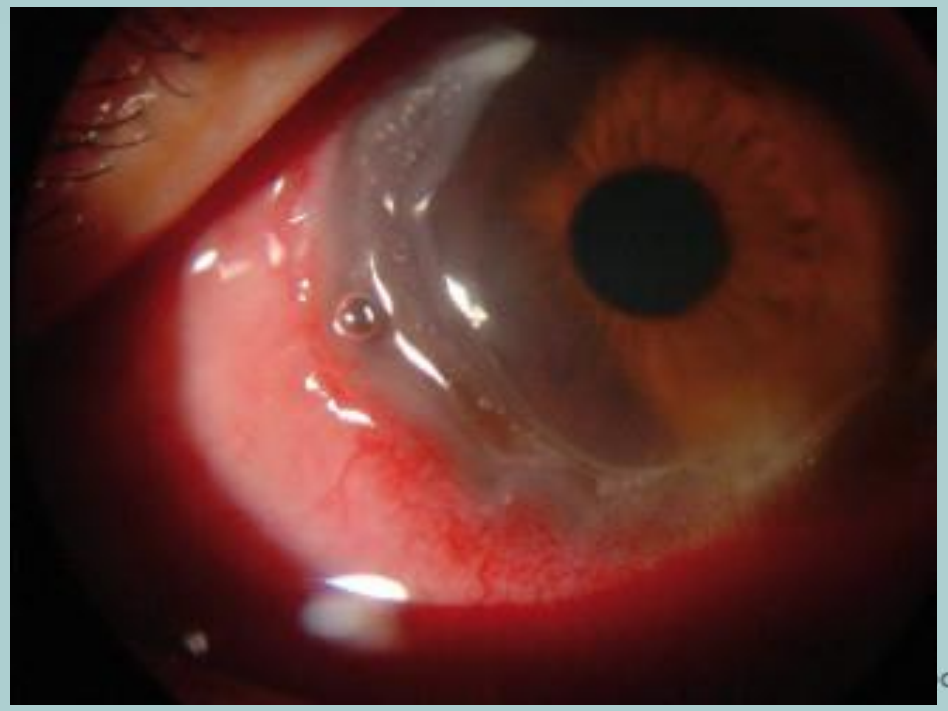
Continued circumferential spreading

Hour glass cornea (contact lens cornea)









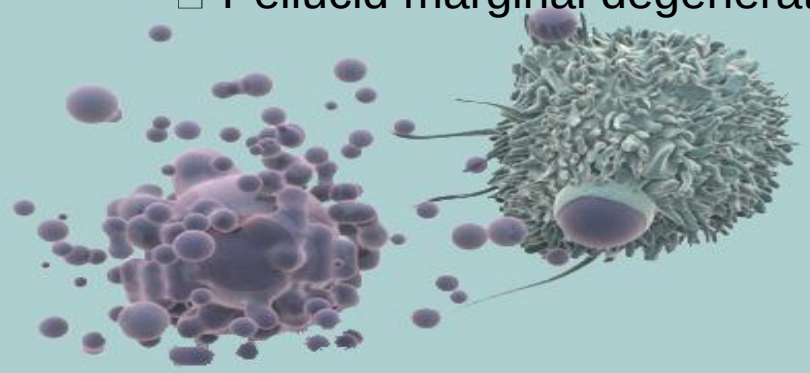
Differential diagnosis of PUK:

Infectious causes:

- HSV,VZV
- Acanthamoeba keratitis
- Bacterial keratitis(staph, strep, gonococcus, Moraxella..)
- Fungal keratitis.

Corneal degenerative conditions:

- Terrrien’s marginal keratitis.
- Pellucid marginal degeneration.

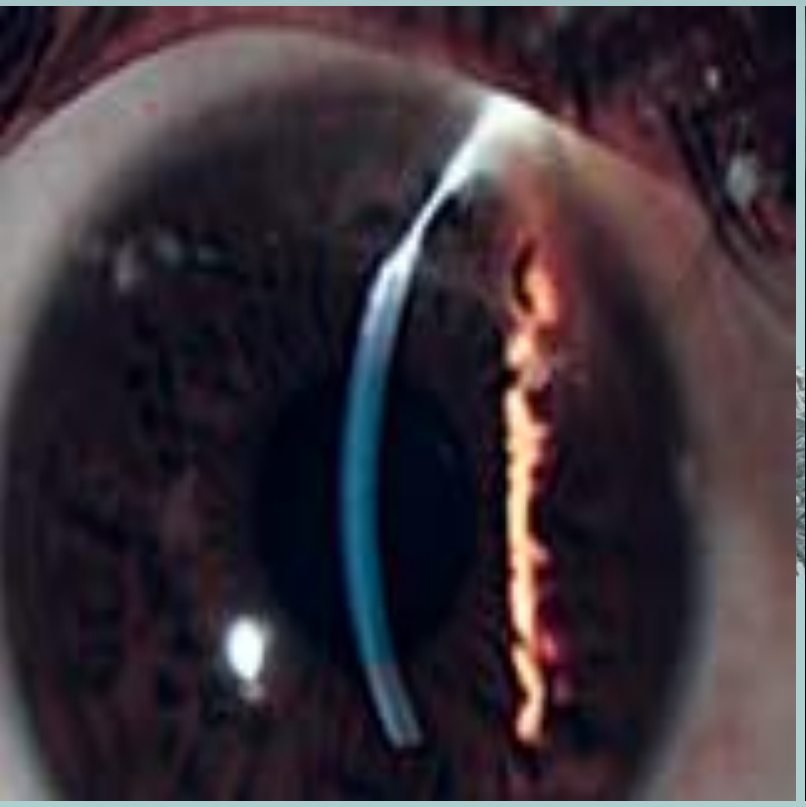


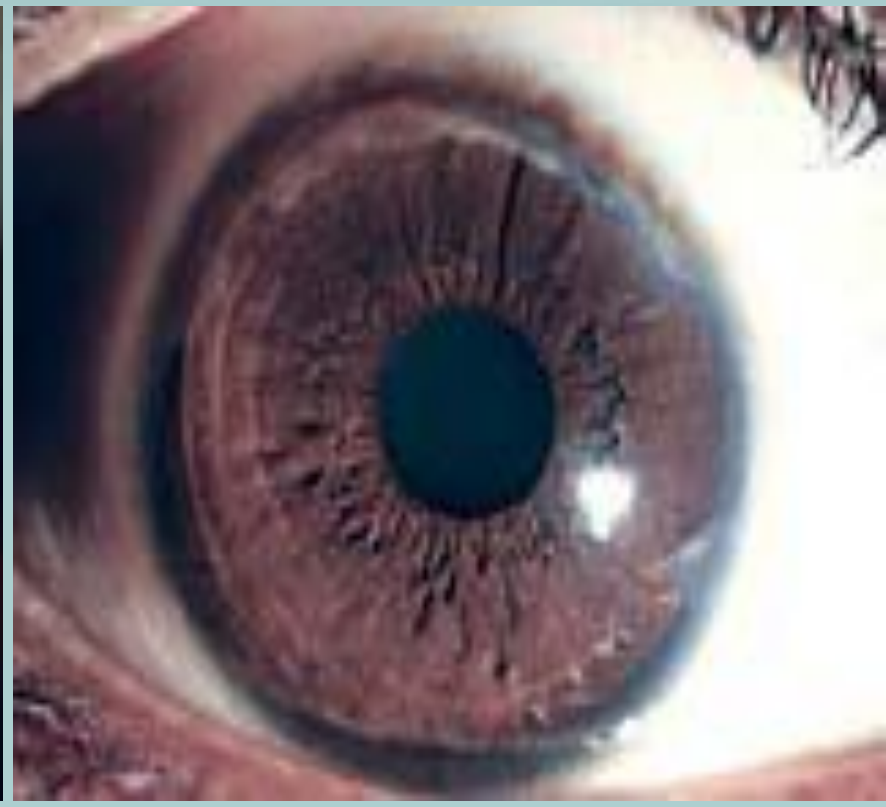
Others:

- Traumatic,postsurgical.
- Mooren ulcer.
- Exposure keratopathy.
- Rosacea.

TERRIENS MARGINAL DEGENERATION

- both are progressive, peripheral corneal thinning may cause decreased V/A
- Both can perforate.

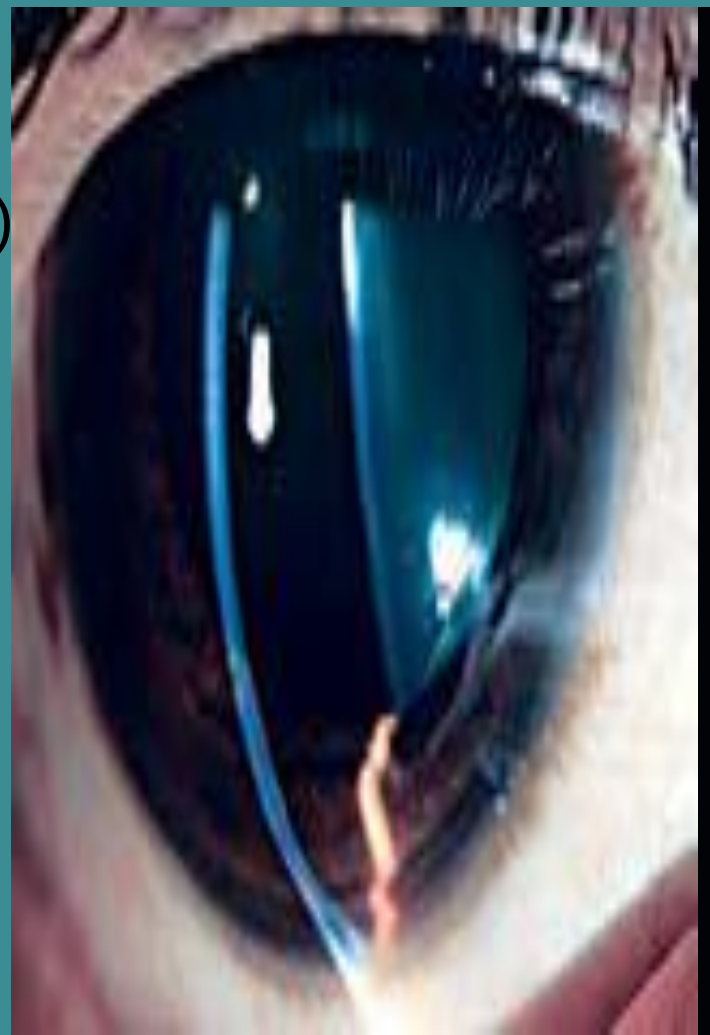


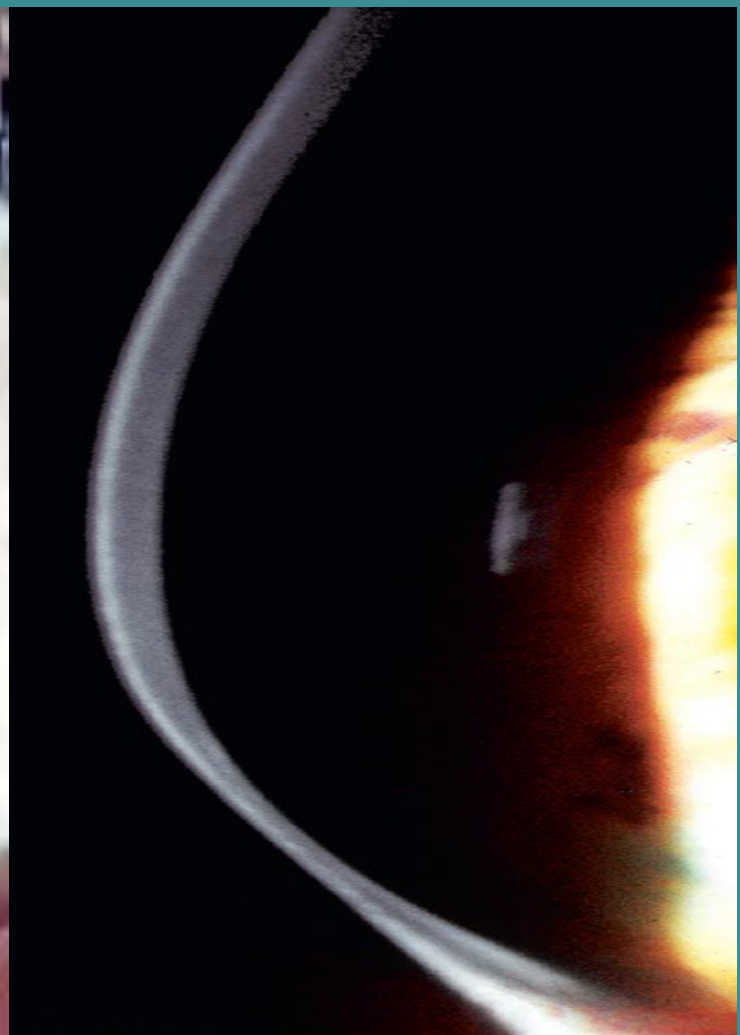


Terriens marginal degeneration	PUK
Asymptomatic (painless)	symptomatic
Corneal epithelium intact	Epithelial defect
Non inflammatory condition: i.e no conjunctival injection or AC reaction	inflammatory
bilateral	Usually unilateral but can be bilateral
Begins superiorly	Mooren: nasally or temporally
More in men	More in women when CVD associated
A yellow line (lipid) may appear,with a fine pannus over the thinned areas& neovascularization.	Inflammatory infiltrates

PELLUCID MARGINAL DEGENERATION

- Bilateral, painless
- Inferior corneal crescent thinning usually from 4- to 8-o’clock portions)
- Progressive
- Clear zone of cornea
- Epithelium intact
- Adjacent conjunctiva no inflammation.
- Corneal ectasia above thinning
- High against the rule astigmatism
- Corneal topography





LABORATORY INVESTIGATIONS for PUK

1. CBC , ESR
2. RF(+ve in 80 % patients with RA)
3. Angiotensin Converting Enzyme (Sarcoidosis)
4. Anti Nuclear Antibody(ANA) (SLE ,RA)
5. Antibody to Double Strand DNA (anti –ds DNA)(SLE)
6. Antibody to small nuclear ribonucleoprotein (anti-RNP)(SLE)
7. Anti neutrophil cytoplasmic antibodies (ANCA) c-ANCA in WG 96%.
8. Anti CCP
9. Urine Analysis
10. HBsAg
11. FTA –ABS
12. CXR
13. Sinus CT

LOCAL INVESTIGATIONS

- Corneal scraping/culture (to rule out primary or secondary infection).
- Conjunctival biopsy in suspected autoimmune associated PUK

TREATMENT

Treatment goals:

- Prevent superinfection.
- Promoting re-epithelialization.
- Control of inflammation

Prevent superinfection:

Prophylactic topical antibiotics - prevent secondary infections (staph in MGD- Pseudomonas)

Promoting re-epithelialization

LUBRICATION

- Diluting effect of inflammatory cytokines in preocular tear film
(Many rheumatoid patients have KCS as a manifest of secondary Sjogren syndrome)

- **Melting stops or slows down if epithelium made to heal**
- **How? By Lubricants ,Patching , Bandage Contact lens, punctal occlusion, possibly tarsorrhaphy.**

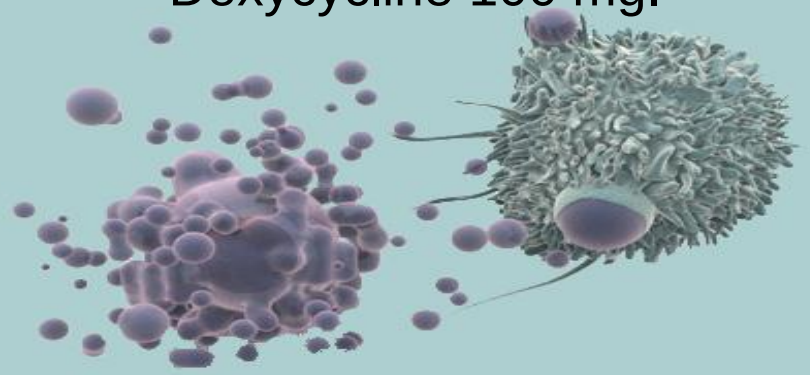
Control of inflammation

1-Topical Collagenase Inhibitors

- Sodium citrate 10 %
- Acetylcysteine solution 20 %
- Medroxy progesterone 1%

2- Systemic Collagenase inhibitors

- Tetracycline250 mg QID.
- Doxycycline 100 mg.

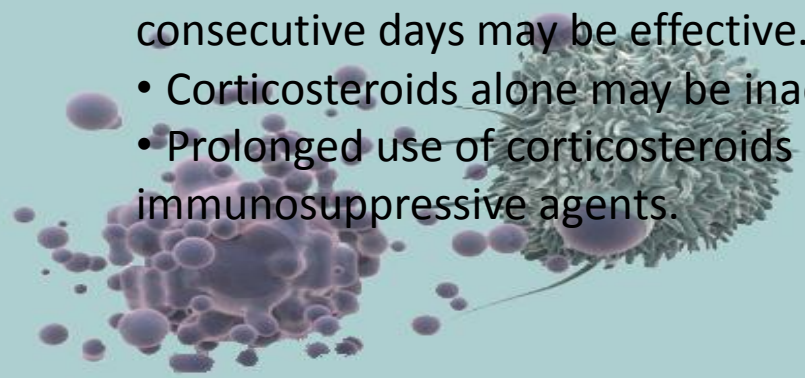


3- Topical corticosteroids

- initial therapy in milder cases (typically when not associated with a systemic CVD)
- Useful in mild cases of RA-associated PUK
- May be harmful in a subset of vasculitic PUK because they inhibit new collagen production.
- ***Not effective in WG, microscopic polyangiitis, Churg–Strauss syndrome, and PAN.***
- In these cases, corticosteroids may promote progression and even enhance perforation and thus must be used judiciously.

4- Systemic corticosteroids

- Oral prednisone ***1 mg/kg/day*** - very commonly used for the management of more severe cases of PUK.
- If there is progression, pulsed ***methylprednisolone (0.5-1.0 g)*** for 3 consecutive days may be effective.
- Corticosteroids alone may be inadequate to control the progressive ocular disease process
- Prolonged use of corticosteroids - significant ***systemic side-effects*** necessitating alternative steroid-sparing immunosuppressive agents.



5- Systemic immunosuppressants

Indications :

1. potentially lethal systemic vasculitic syndromes such as PAN, RA, SLE, Sjogren's syndrome, WG, allergic angiitis of Churg-Strauss,GCA
2. necrotizing scleritis and vasculitis confirmed by histopathologic analysis of ocular tissue.
3. Bilateral and /or progressive Mooren's ulcer.
4. Disease progression despite local conjunctival resection and tectonic procedures (e.g., tissue adhesive)

- In *idiopathic PUK,* Studies suggest that ***cyclosporin A*** with dosages in the range of 2.5mg/kg/day may be a reasonable initial choice,
(especially if nephrotoxicity is not a concern)
- In cases of PUK as with *WG , necrotizing scleritis or Severe or rapidly progressive PUK associated with other CVDs:* First-line therapy is Cytotoxic immunosuppressives, such as ***cyclophosphamide*** (2 mg/kg/day), ***together with corticosteroids (oral or pulse intravenous***).

- **Maintenance:** ***oral or SC methotrexate*** (7.5 -12.5 mg /week)

Surgical Treatment

- Conjunctival resection

• Temporarily remove the limbal source of local cellular mediators and collagenases important in progression of the disease process

• May be of great diagnostic help

- Patch graft

• When a perforation is too large for tissue adhesive to seal the leak, some type of patch graft will be necessary.

• This may range from a small tapered plug of corneal tissue to a penetrating keratoplasty

- Tissue adhesive *cyanoacrylate glue*

- Amniotic Membrane transplantation- preserve the integrity of the globe

- Conjunctival flaps

 control corneal melting in difficult to manage microbial keratitis,

 but best avoided in immune mediated disease

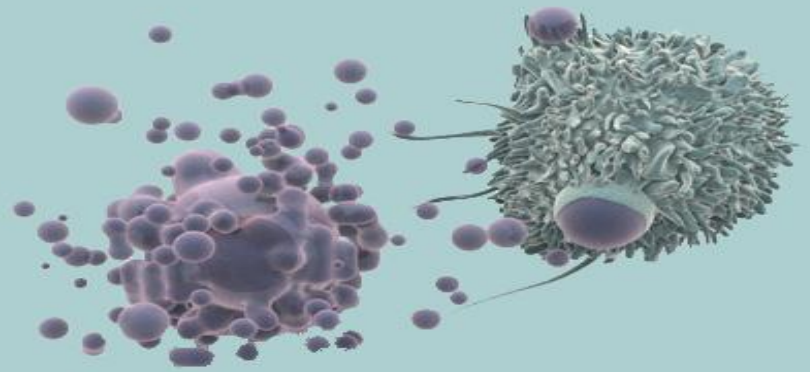
 Bringing conjunctival vasculature closer to area of corneal disease accelerate Melting

- Rehabilitative surgical therapy

• Initial ***lamellar tectonic grafting*** followed by ***central penetrating keratoplasty***

prognosis

- Many patients with mild or moderate PUK may ***maintain good vision*** if the inflammatory process is rapidly controlled.
- The prognosis is more guarded when PUK is with a systemic CVD.
- Significant visual loss and ocular morbidity may develop with corneal perforation.
- Both ocular and systemic prognosis is more guarded when there is concomitant scleritis, especially necrotizing scleritis.



CASE:

A 63- year-old woman presents with a red, very painful right eye for several weeks

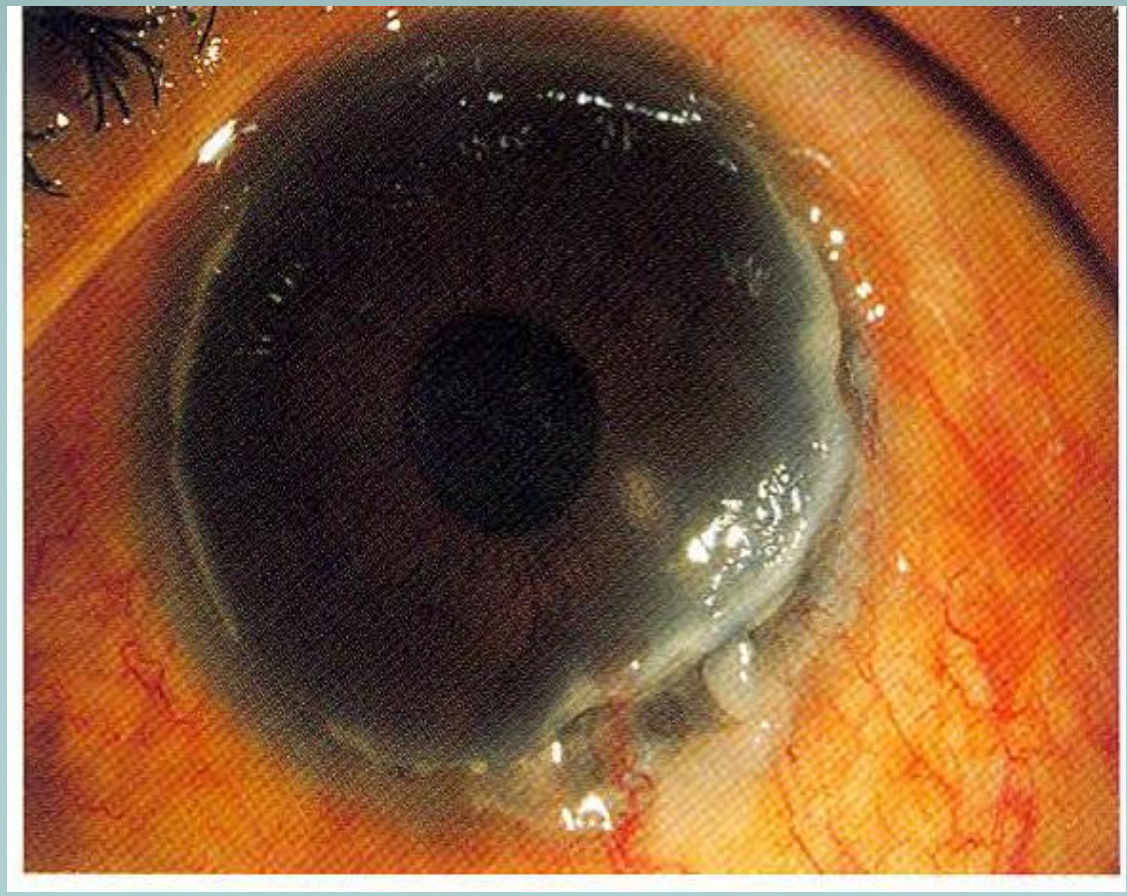
Examination discloses an ulcerative, circumferential marginal keratitis with a leading, undermined edge and early neovascularization. Which one of the following regarding the condition is **false**?

a. There is dysregulation in both cellular and humoral immunity.

b. A milder, less painful variant may be seen in young African American men.

c. Medical management might include oral prednisone, cyclophosphamide or methotrexate.

d. An evaluation for connective tissue disease is mandatory.

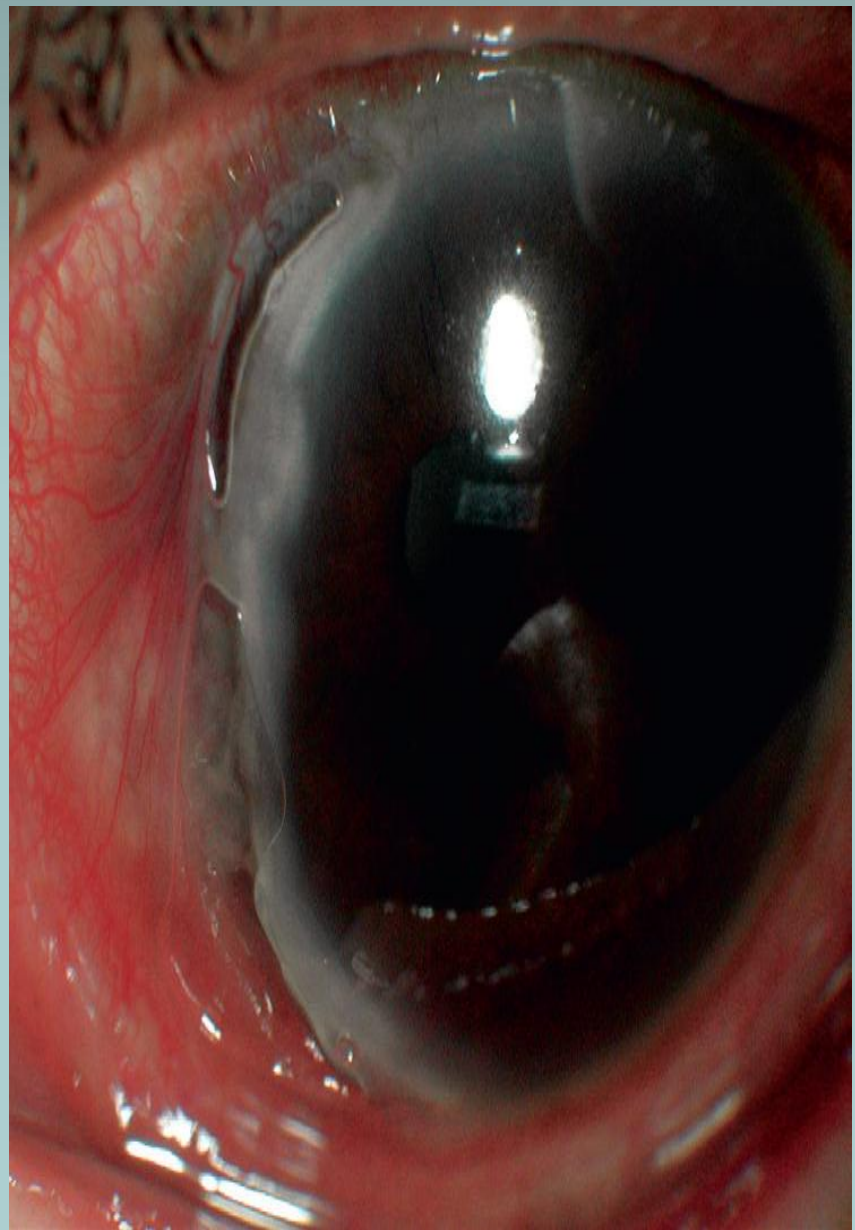


Mooren's ulcer

- *rare, degenerative, superficial ,idiopathic disease*

- **strictly a PUK, with no associated scleritis.**

- **Mooren's ulcer can occur in all age groups, but the vast majority present b/w *40 and 70 years*.**
- **Disease can occur in either sex, but *men* outnumber women.**
- ***Interpalpabral limbus* is involved most often, followed by the inferior and then the superior limbus.**
- **occurs in complete absence of any diagnosable systemic disorder**



SYMPTOMS:

- severe and persistent chronic pain.
- Redness Photophobia Lacrimation
- Decrease in visual acuity due to associated iritis, central corneal involvement and irregular astigmatism.

- rapidly progressive, ulcerative keratitis that begin peripherally and progresses circumferentially , then centripetally
- single or multi-centric crescent shaped peripheral corneal ulcer with a characteristic steep, infiltrated, undermined, or overhanging edges.

Mooren ulcer types

Limited type(typical or benign Mooren's ulcer)

- More common (75%), benign, and unilateral
- mild to moderate symptoms.
- generally responds well to medical and surgical treatment.
- occur in older patients.

Atypical or malignant Mooren's ulcer

- bilateral although both eyes may not be affected simultaneously
- relatively more pain and generally a poor response to therapy.
- younger patients more common in African-American
- progresses relentlessly and is more likely to result in corneal perforation.
- may be associated with coexistent parasitemia.

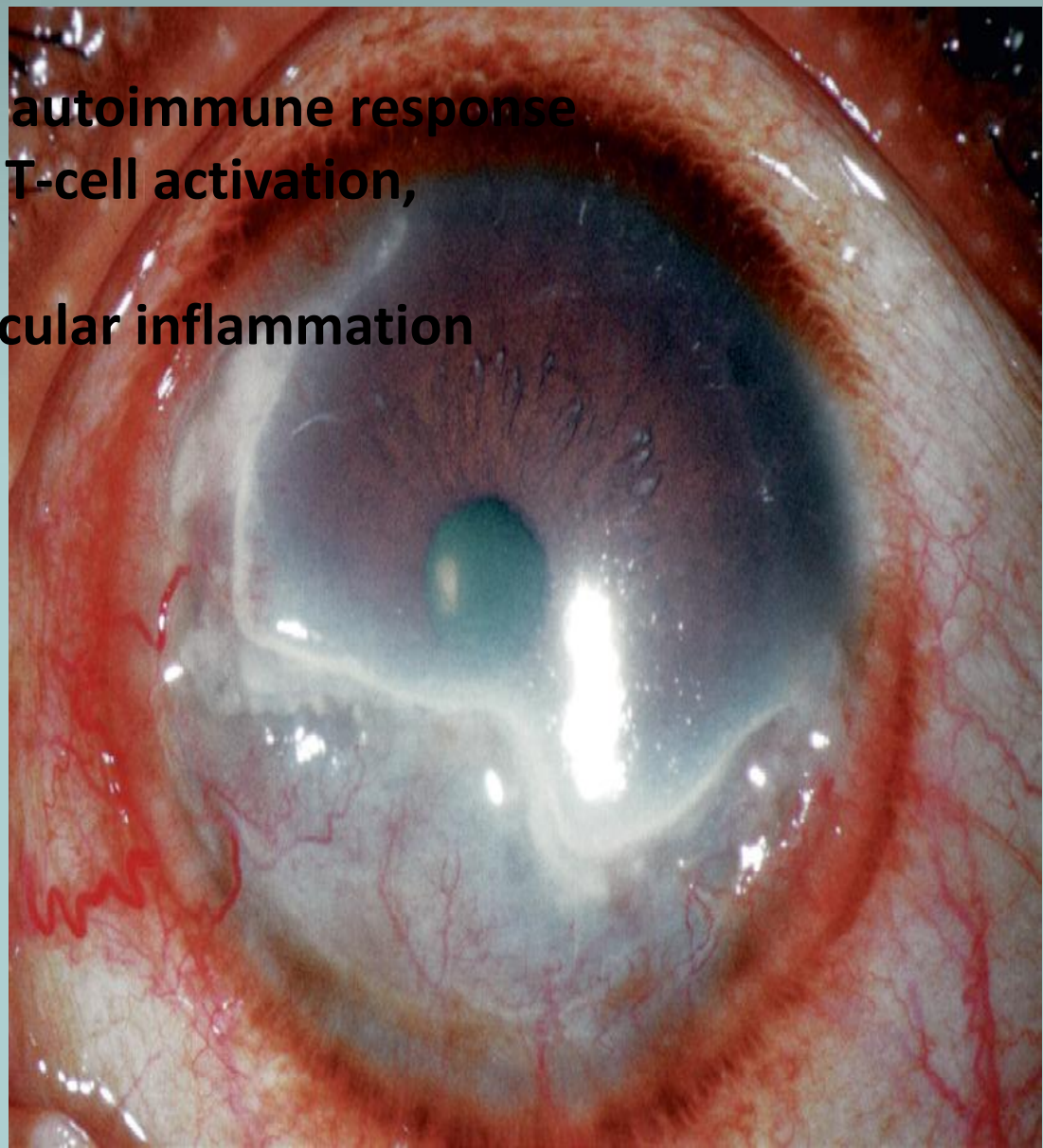
Pathogenesis

- Trauma or infection alter normal corneal antigen autoimmune response
- Altered antigen taken up by macrophages(APC), T-cell activation, differentiation, and proliferation.
- Lymphocytes return to the conjunctival vessels ocular inflammation

Healing: Corneal epithelialization and vascularization associated with scarring and thinning.

Advanced cases: most of the cornea is lost leaving behind a central island surrounded by area of grossly thinned, scarred, and vascularized tissue.

Corneal perforation is uncommon in type 1.



Management

- stepwise approach to management is recommended

- Unilateral, mild-moderate cases.

1.Topical steroids hourly and low-frequency antibiotics.

2. Conjunctival resection:

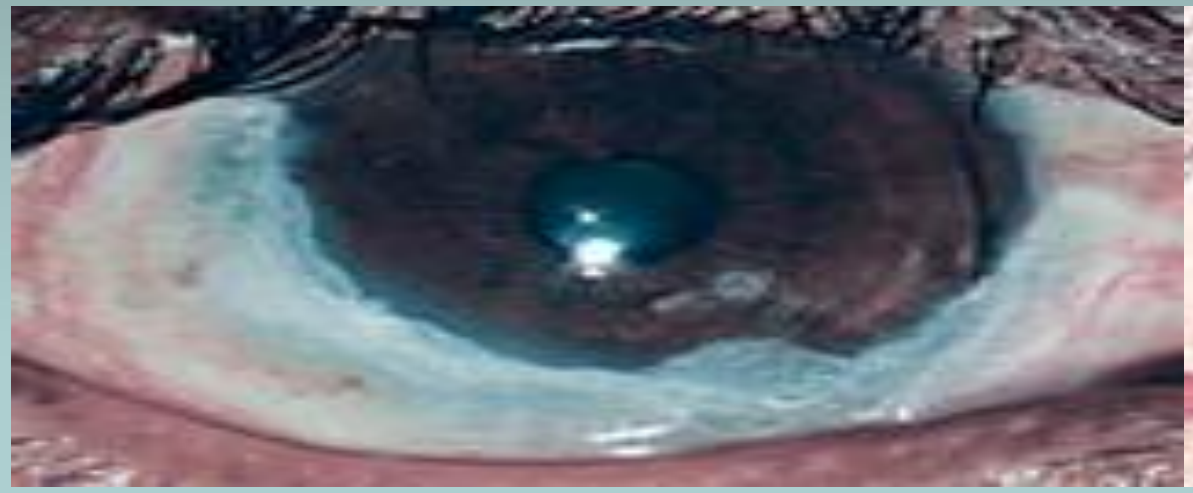
Conjunctiva is resected 4 mm back from the limbus and 2 mm beyond the circumferential margins.

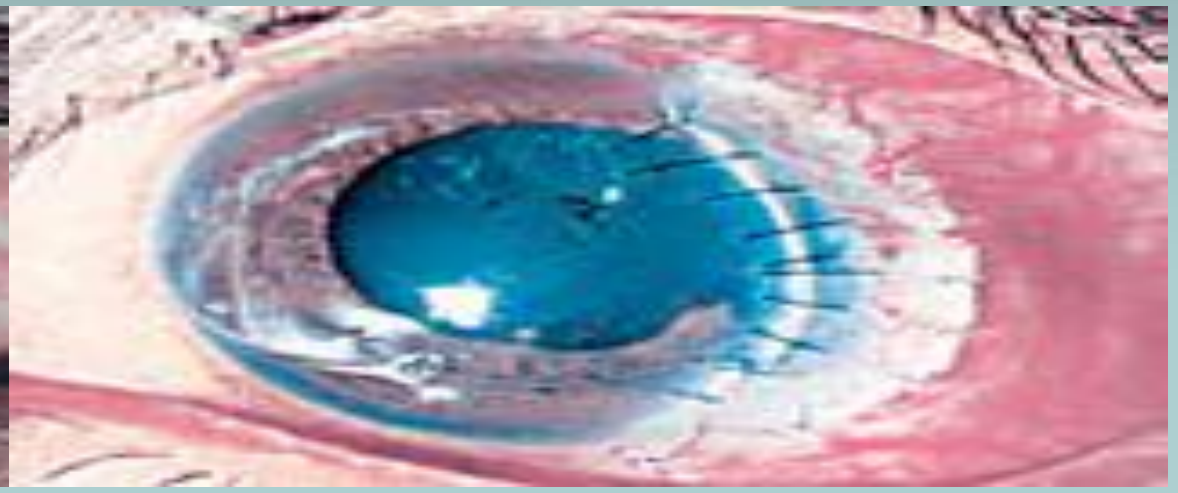
3. Topical acetylcysteine 10% and L-cysteine (0.2)topical.

4- Cyclosporine up to 2%.

In case of Perforation

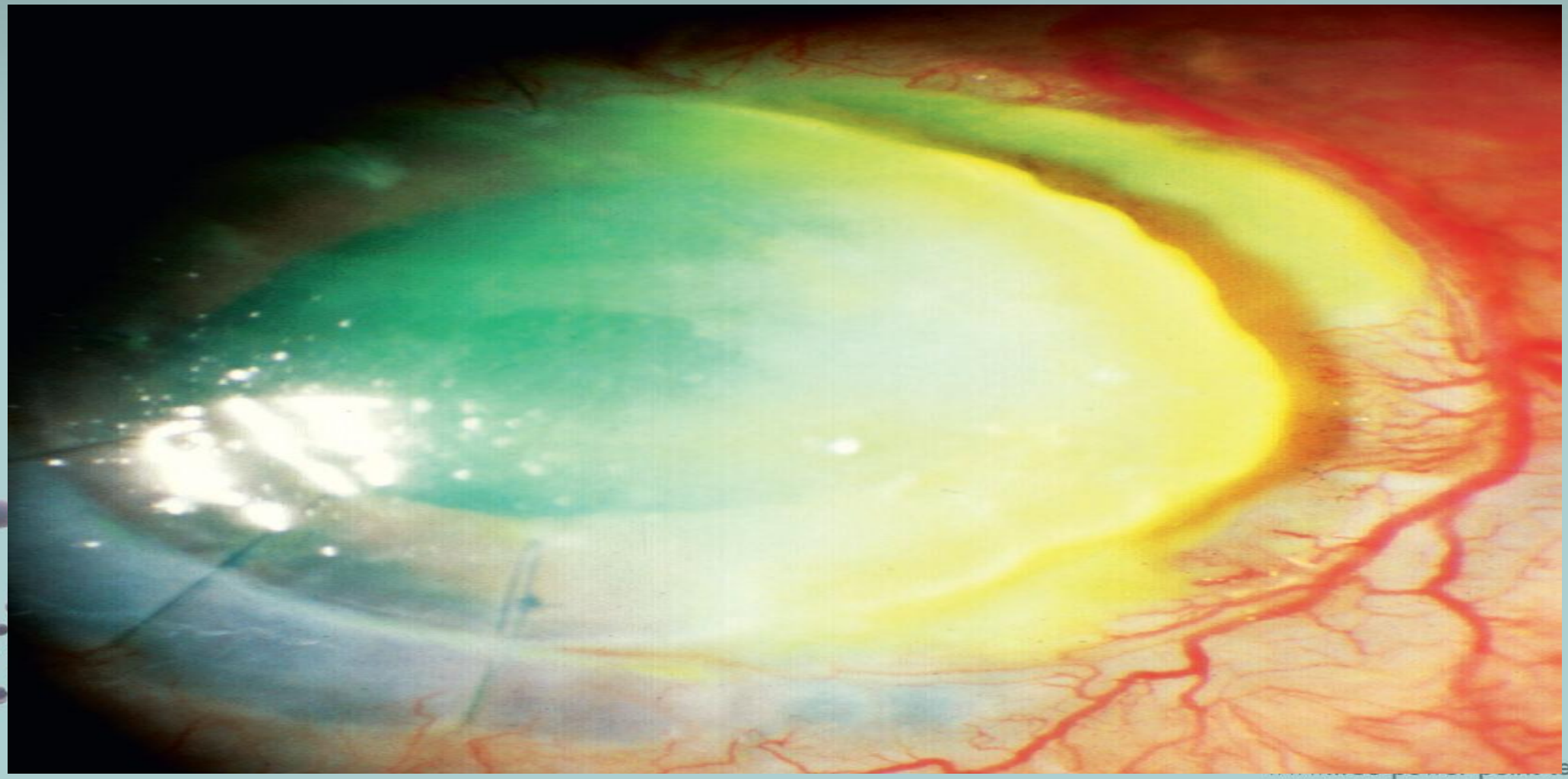
- Ulcer debridement with ***cyanoacrylate adhesive*** and ***bandage contact lenses***.
- ***Lamellar keratoplasty*** with ***IV methotrexate***– halt the process





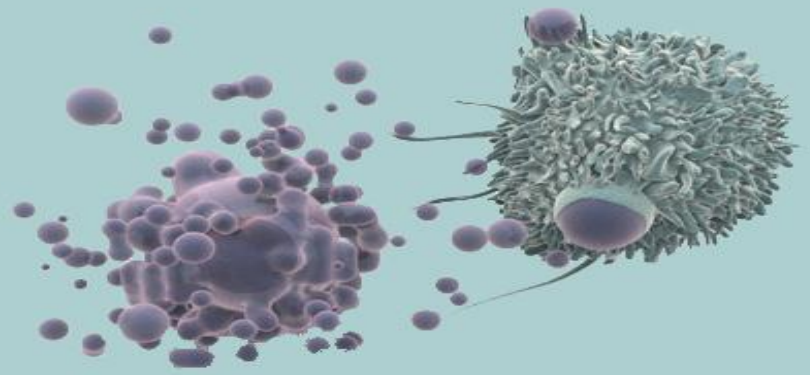
Bilateral or progressive Mooren's ulcer that didn’t respond therapeutic steroids and conjunctival resection will require systemic cytotoxic chemotherapy

When penetrating keratoplasty is done!!



PUK vs Mooren

PUK	MOOREN
SYSTEMIC IMMUNE DISEASE	NO SYSTEMIC DISEASE
Discomfort unless associated with scleritis	Severe pain
females	males
Scleritis may happen	No scleritis
Guarded prognosis	better

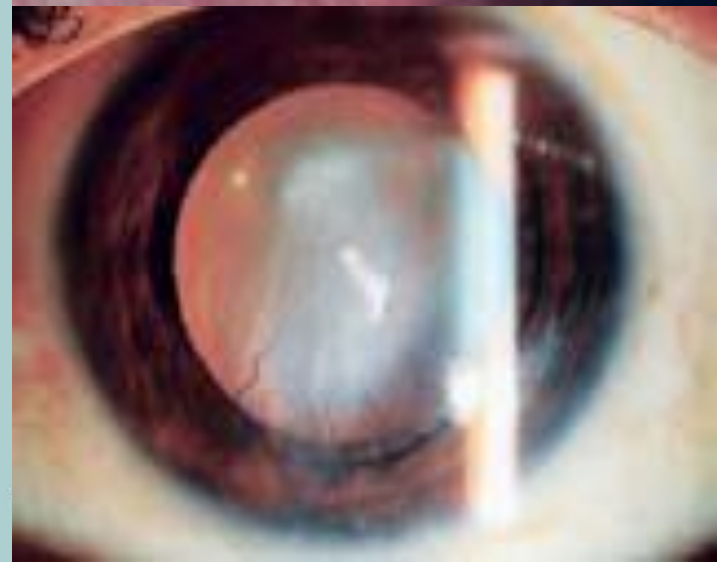


The diagnosis should be PUK from the beginning; then, if the aggressive systemic evaluation is negative and the adjacent sclera is not involved, it's appropriate to hang the label Mooren's ulcer on this patient “dr. Foster said” “Because PUK If not treated, the patient will die,"

Interstitial Keratitis (IK)

Definition:

Inflammation of corneal stoma without primary involvement of epithelium or endothelium.



Etiology

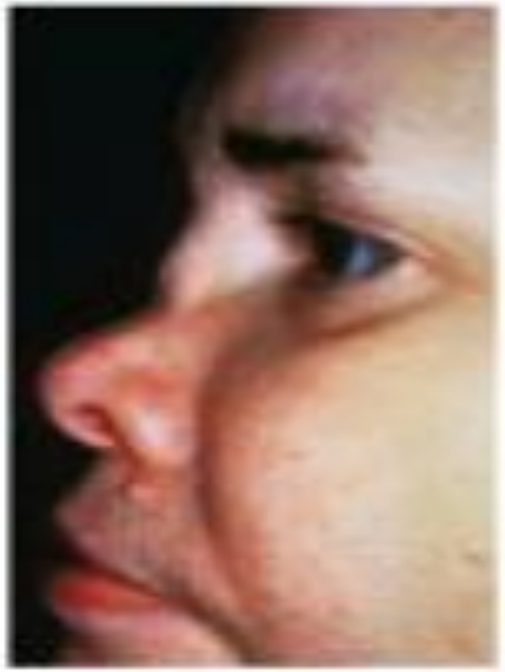
Infectious:

- Bacteria: syphilis (congenital 90% M.C cause of bilateral IK- or acquired), TB (unilateral, sectoral), borrelia, leprosy (but not in USA).
- Virus: HSV,VZV , EBV.
- Parasitic: onchocerciasis, leishmania, malaria, African sleeping sickness (taenia cruzi)

Non-infectious

Phlyctenular keratitis, Cogan's syndrome, sarcoidosis, lymphoma, or contact lens-related.

Frontal bossing;
Saddle nose

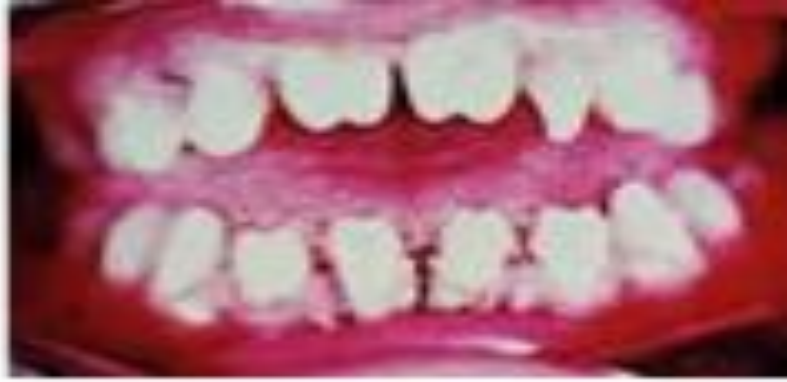


Saddle nose

Interstitial keratitis

Rhagades

Perforated hard palate



Hutchinson teeth

Mulberry molar

Saber shins

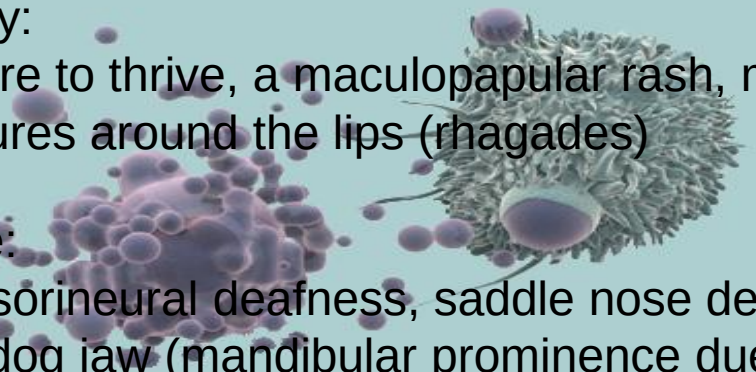
Syphilitic interstitial keratitis

Congenital syphilis

- Interstitial keratitis is a delayed immune-mediated response, not a manifestation of active infection. Onset is at around 5–20 years of age.
The interstitial keratitis is usually bilateral in 80%, although usually not simultaneous.
- can be initiated by minor corneal trauma.

Systemic findings:

Early:
failure to thrive, a maculopapular rash, mucosal ulcers, characteristic fissures around the lips (rhagades)

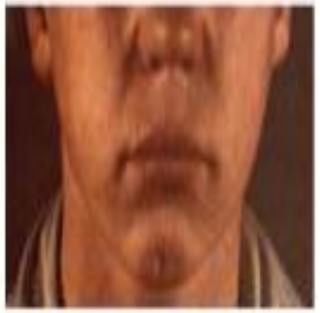


Late:
sensorineural deafness, saddle nose deformity , joint swelling,sabre tibiae, bulldog jaw (mandibular prominence due to maxillary underdevelopment), Hutchinson teeth. Hutchinson's triad refers to IK, deafness, and notched teeth).

Frontal bossing; Saddle nose

Interstitial keratitis

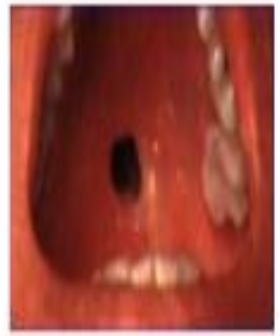
Hutchinson teeth



Rhagades

Mulberry molar

Saddle nose



Perforated hard palate

Saber shins

Ocular history and findings

Initial **symptoms** are pain, watering, photophobia, and redness.

Signs

○ Profoundly decreased VA is typical in the **active stage**.

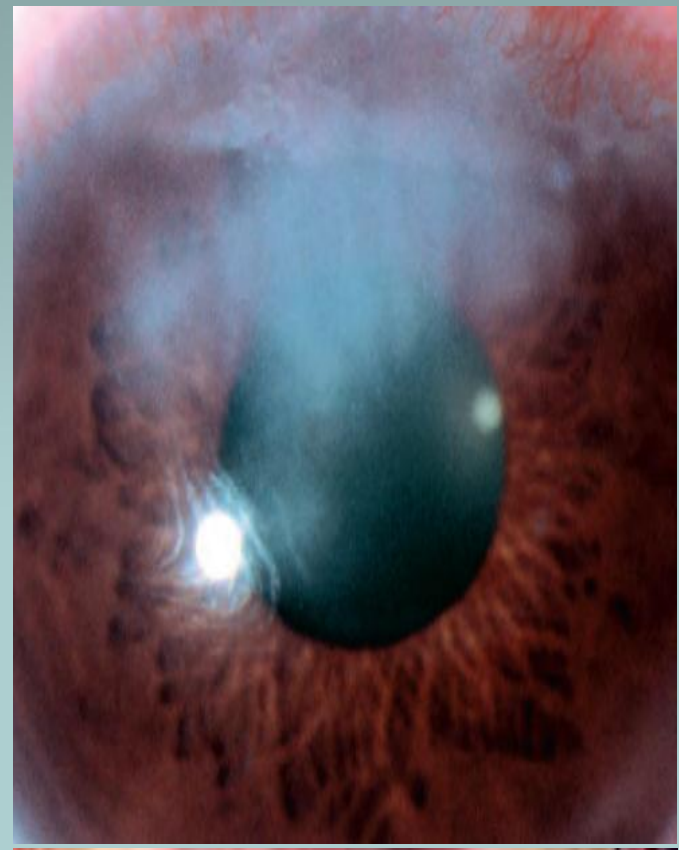
○ Limbitis associated with deep stromal vascularization, with cellular infiltration and clouding that may obscure the still-perfused vessels to give the characteristic pinkish ‘salmon patch’ appearance.

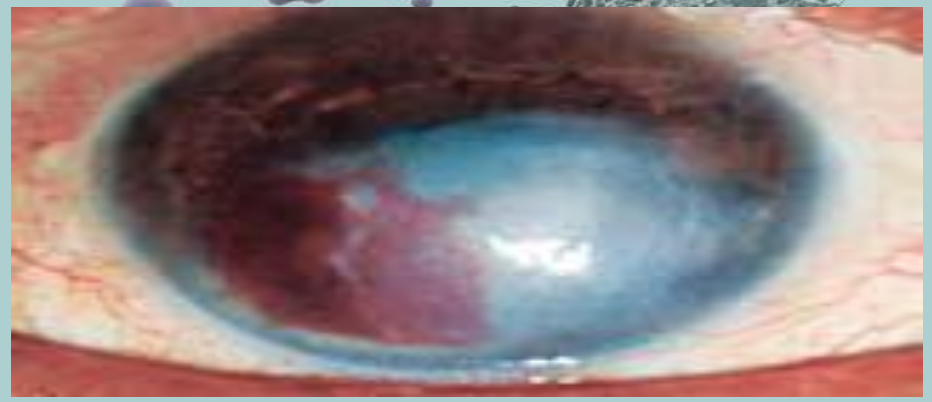
○ Granulomatous anterior uveitis.

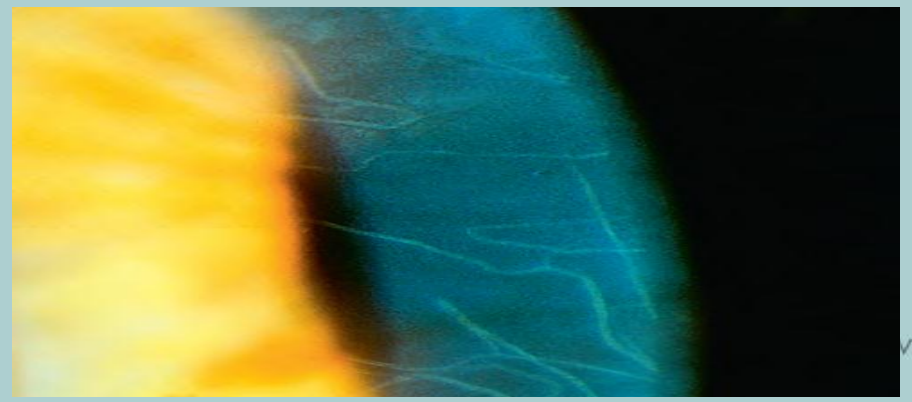
○ After several months the cornea begins to clear and the vessels become non-perfused (‘ghost vessels’)

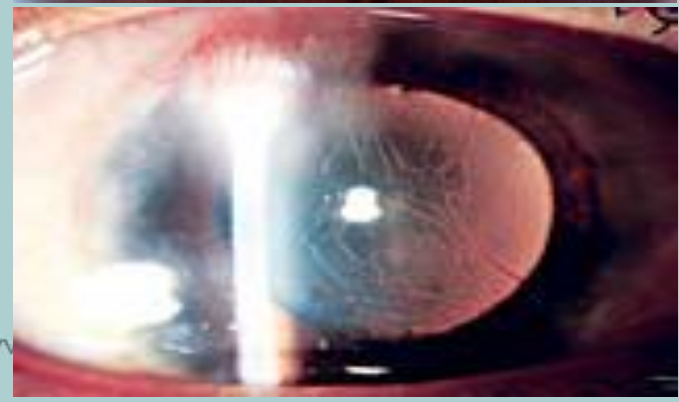
○ If the cornea later becomes inflamed, the vessels may re-fill with blood and, rarely, bleed into the stroma.

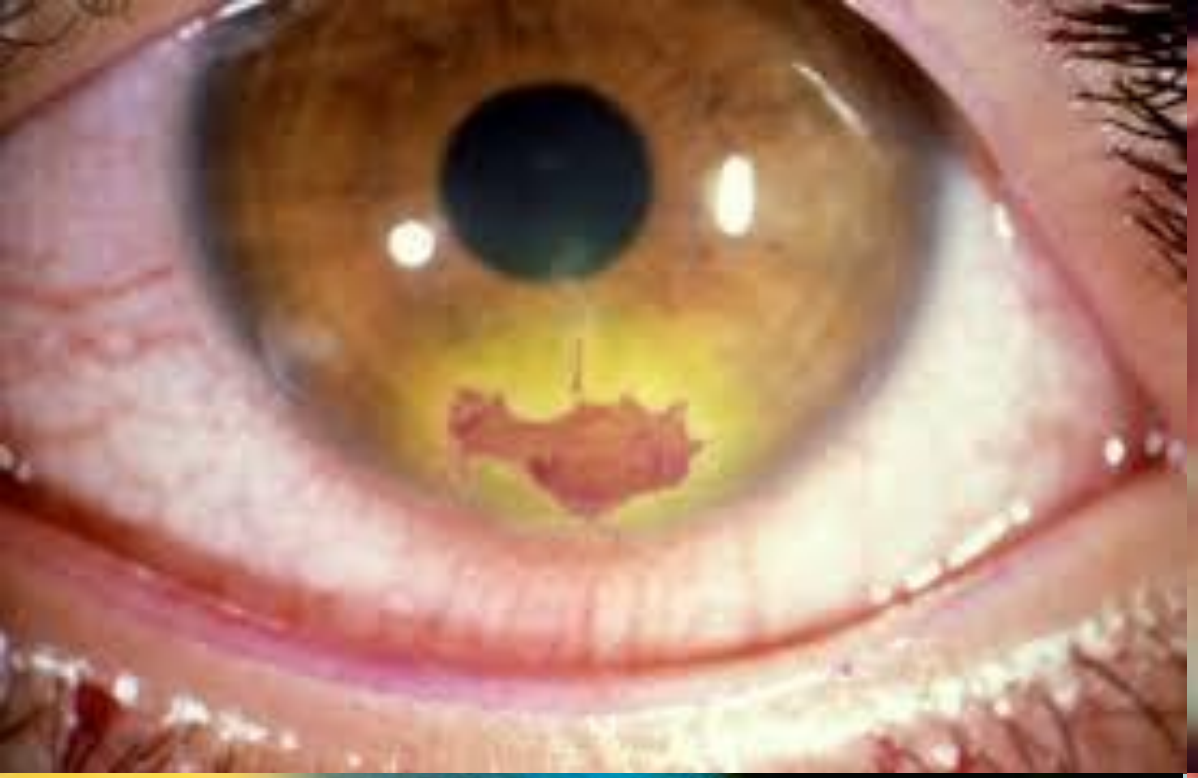
○ The healed stage is characterized by ghost vessels, feathery deep stromal scarring and sometimes thinning, astigmatism and band keratopathy.

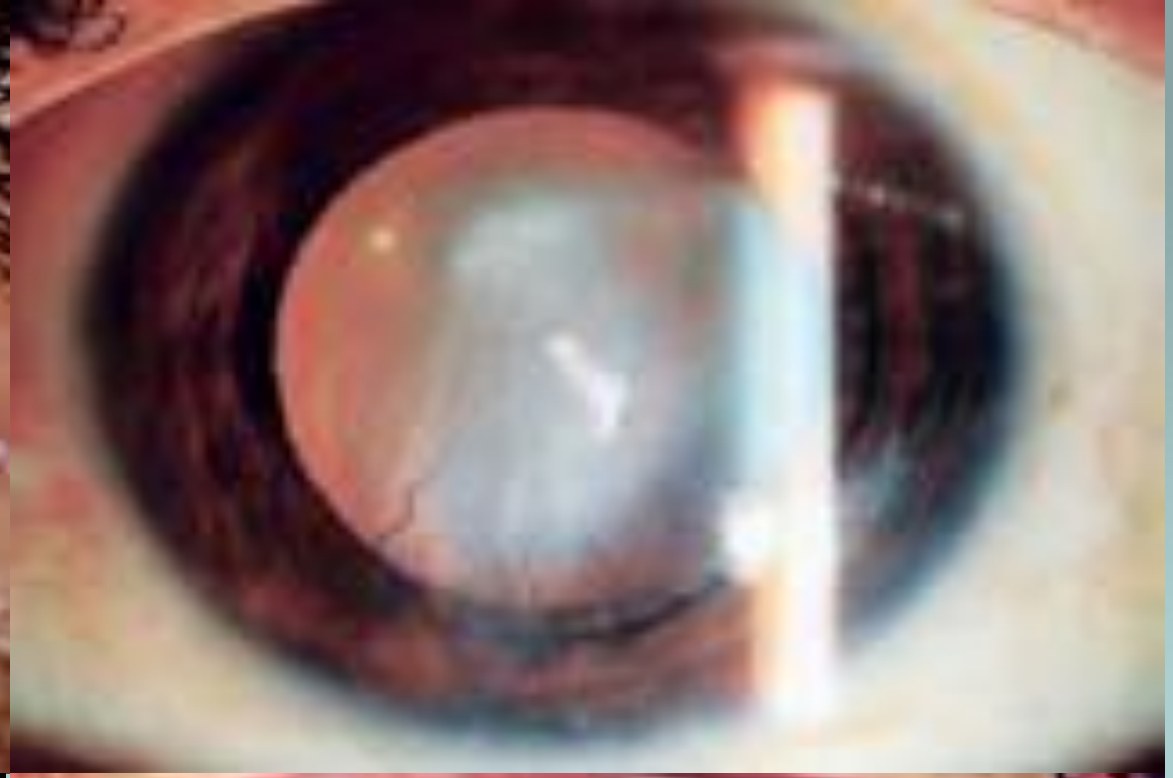


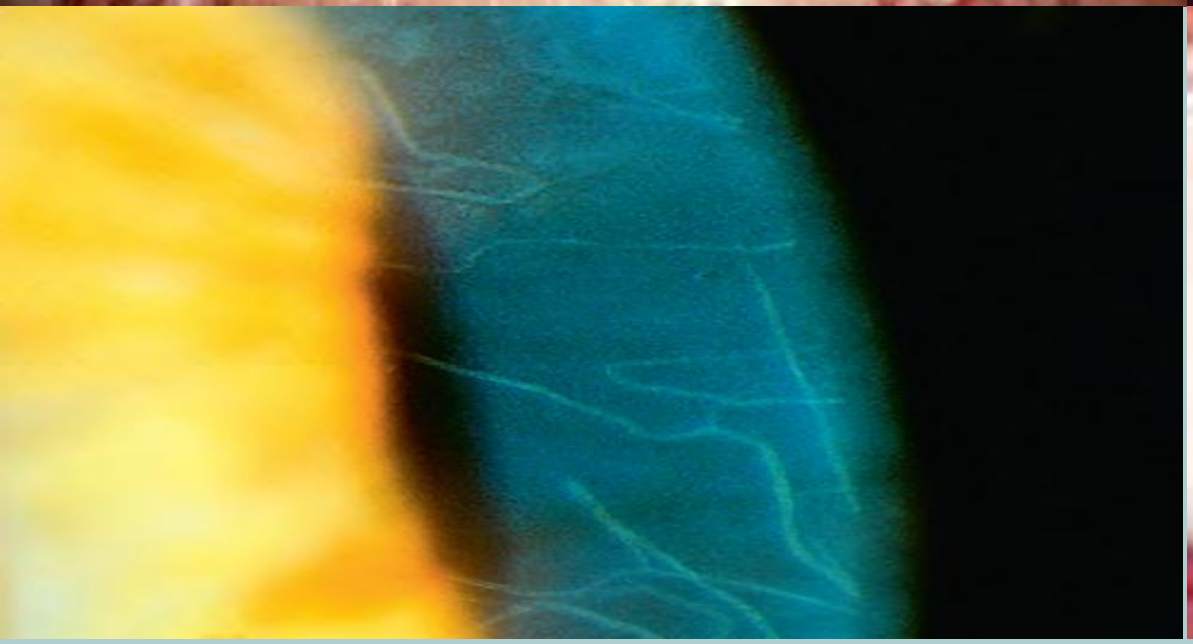


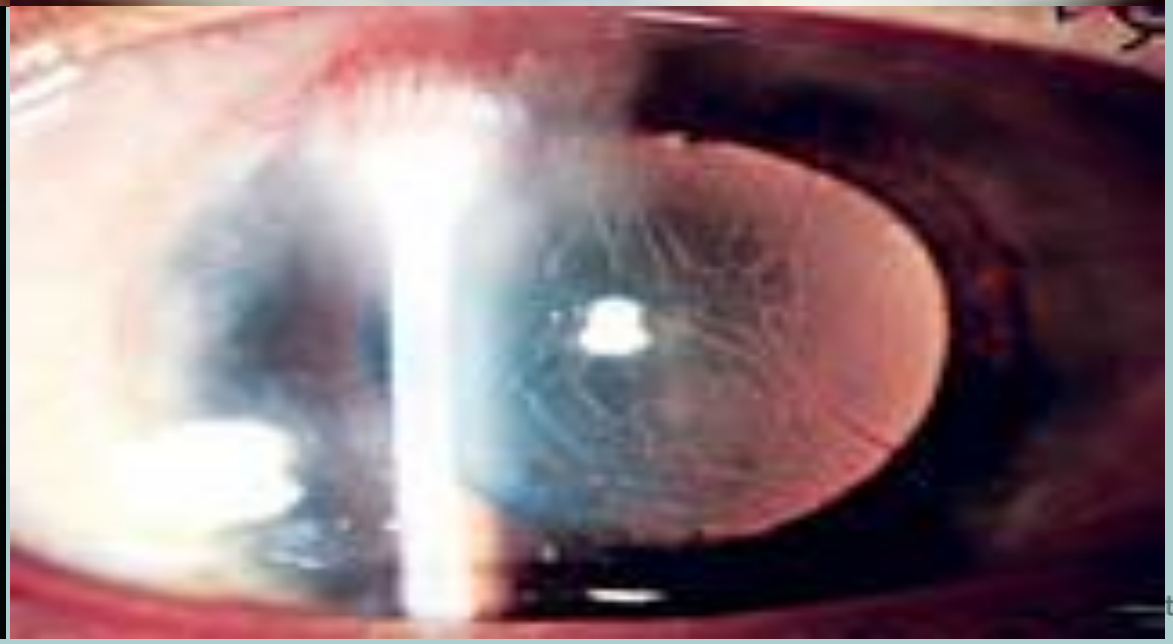








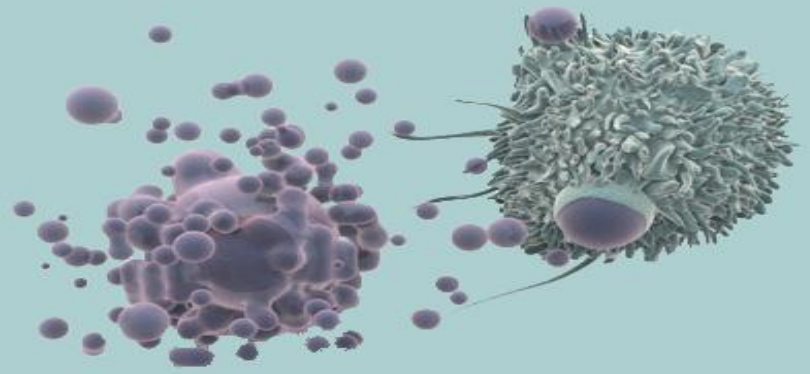


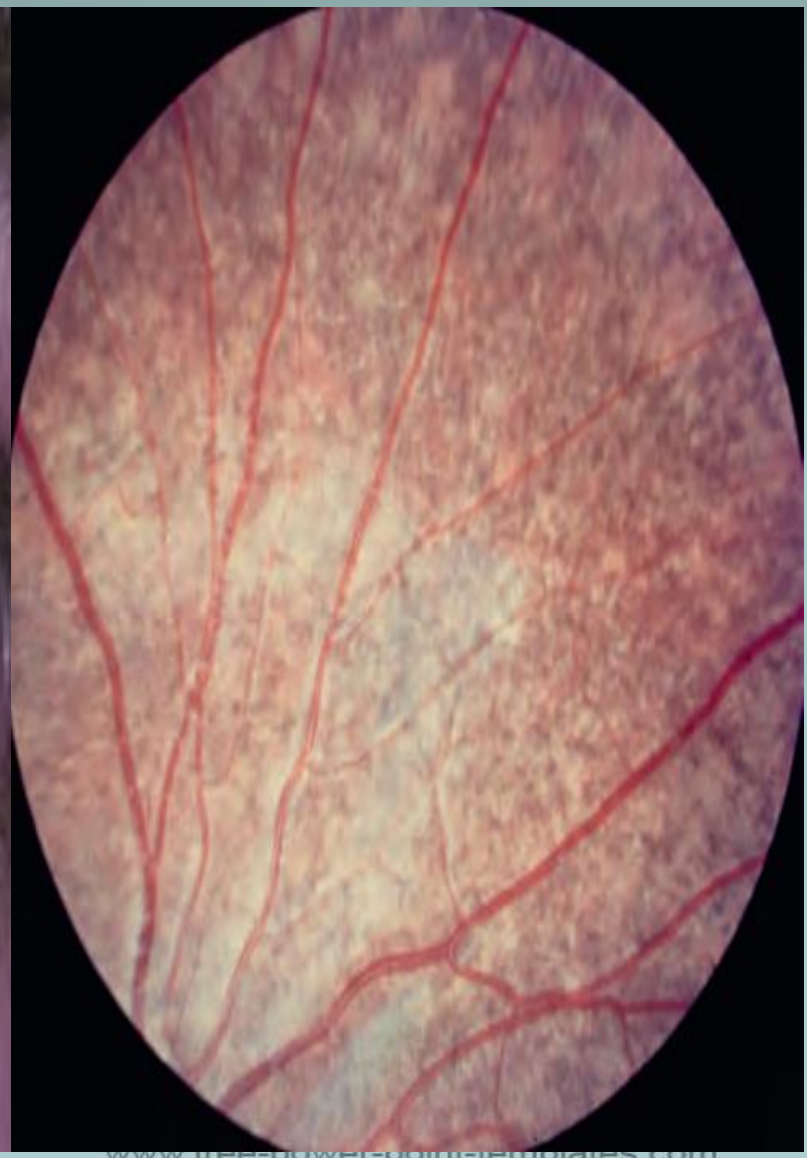


AQUIRED SYPHILIS:

- Interstitial keratitis is rare and unilateral.
- Usually after 10 years from getting the disease (chancre), in neurosyphilis stage. although it can occur as part of primary infection.

Other ocular features including anterior uveitis, dislocated/subluxated lens, cataract, optic atrophy, salt and pepper pigmentary retinopathy and Argyll Robertson pupils are more common than IK.





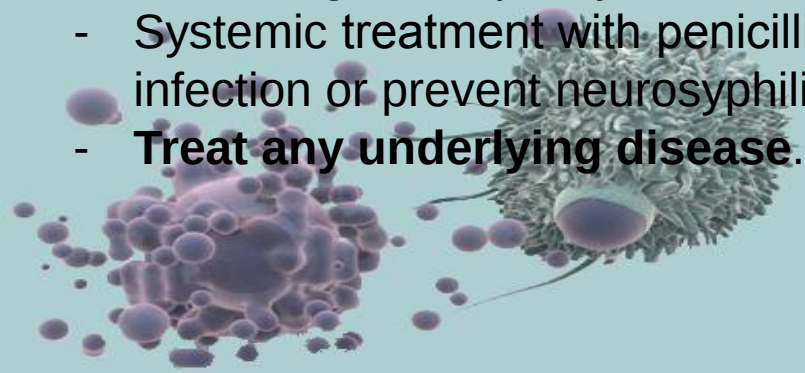
Investigations

- (VDRL) and rapid plasma reagin tests are positive in active disease while the *Treponema*-specific antibody test
- (FTA-ABS) remains positive even after treatment and more specific.
- PPD

Treatment

1. **Acute disease**:
- Intense topical steroids (e.g. dexamethasone 0.1%/prednisolone1% 6–8 times/day) can reduce the inflammation, limit opacification, and improve vision. Tapering steroids may be needed for several months to keep the inflammation under control.
- cycloplegics.

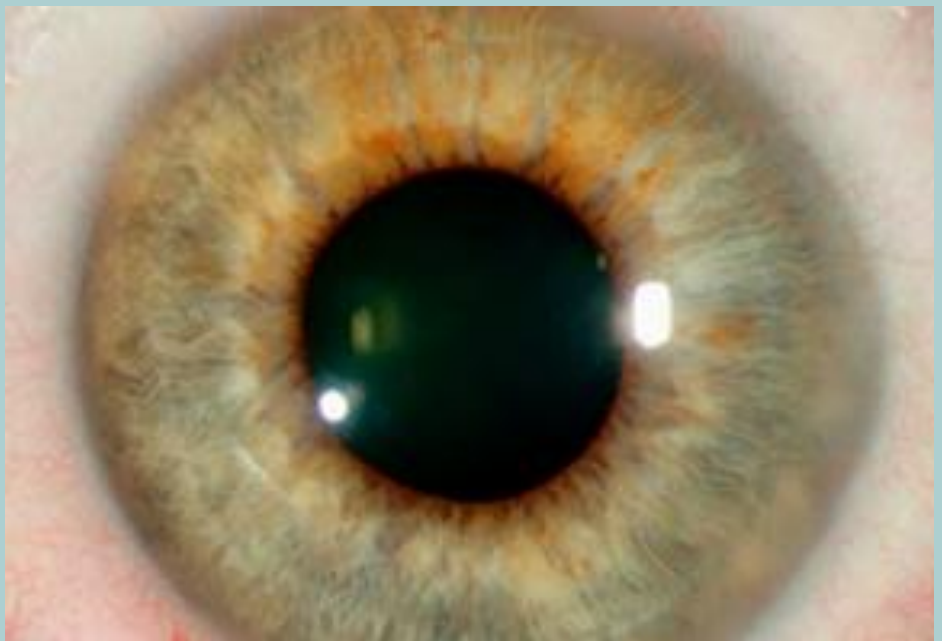
2. **Old inactive disease** with central scarring:
- Penetrating keratoplasty can be considered if minimal amblyopia is present.
- Systemic treatment with penicillin doesn't treat interstitial keratitis but is used to treat systemic infection or prevent neurosyphilis.
- **Treat any underlying disease**.

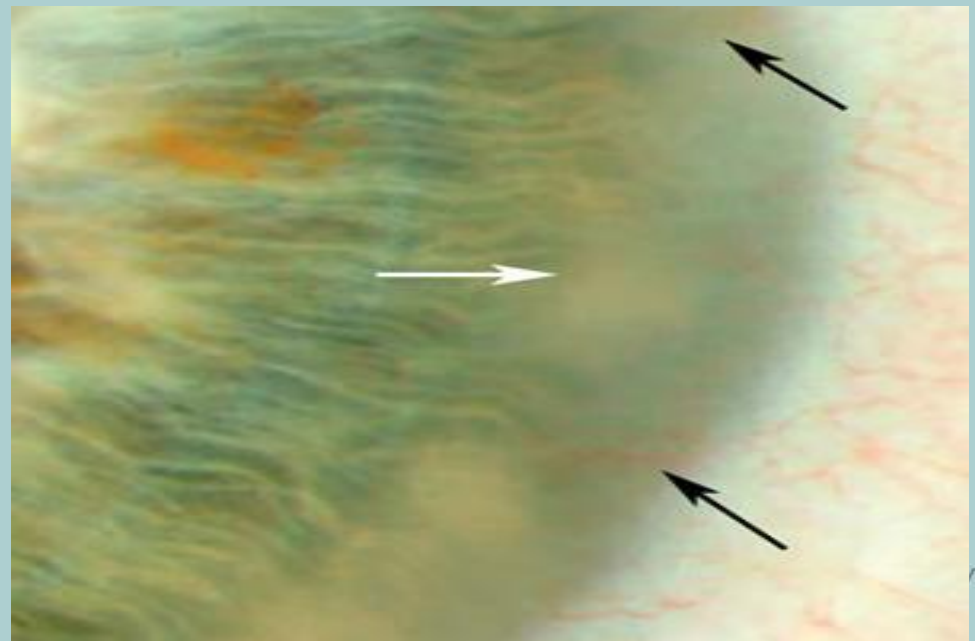


case

42-year-old Caucasian female complains of an annoying red left eye for the past two months with recent onset of vertigo and otalgia.

Slit lamp examination: Limbal edema and stromal opacity were noted as well as several fine vascular loops extending into the mid-stroma of the cornea. There is mild vascular injection, especially inferiorly and at the episclera covered by the lower lid, associated with tenderness of the globe.





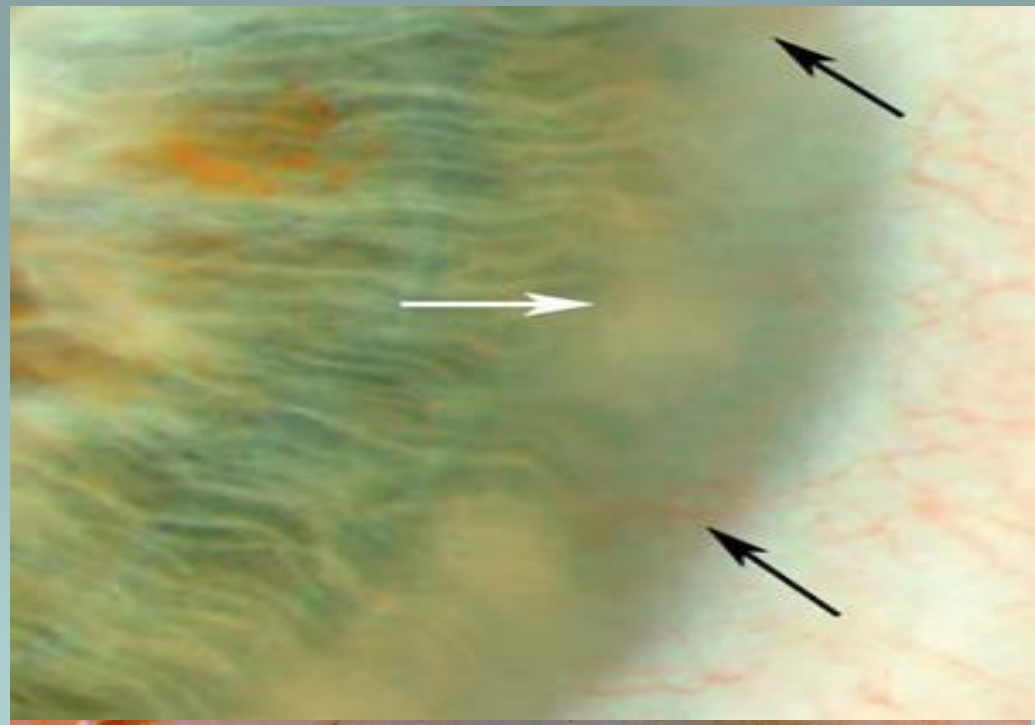
Cogan’s syndrome

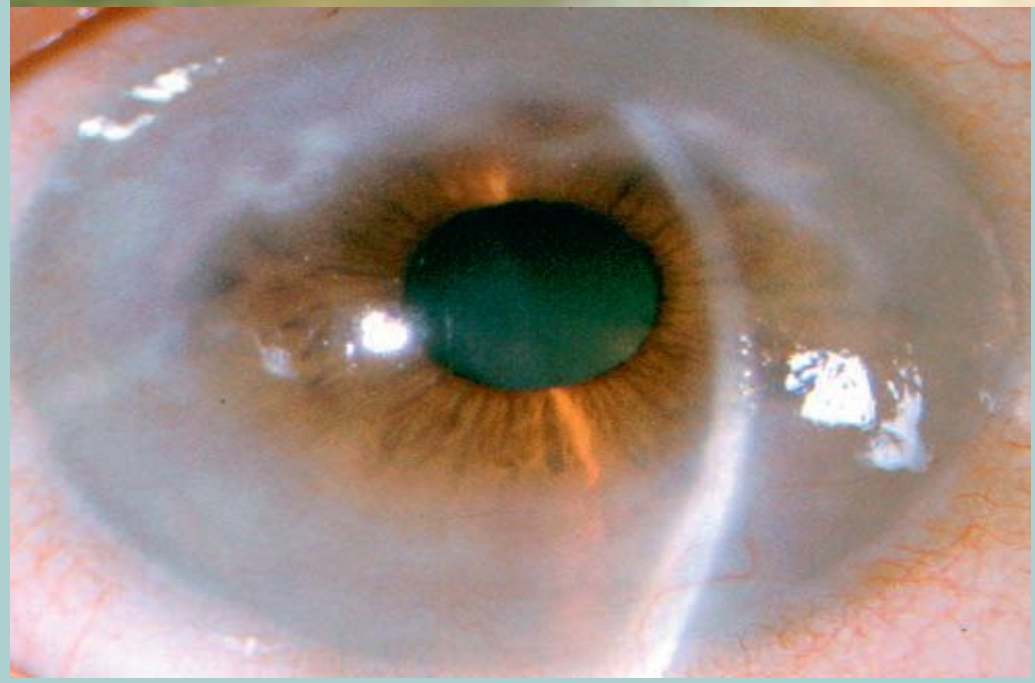
- a rare systemic autoimmune vasculitis characterized by interstitial keratitis, vertigo, tinnitis, and hearing loss developing within months of each other.
- vestibuloauditory dysfunction remain the hallmarks of Cogan’s syndrome.
- The etiology is unknown, but is believed to involve autoimmunity to the inner ear and cornea.

- Affects young adults, F=M; children can also be affected. Systemic features occur in 30% and may include multisystem vasculitis that can be life-threatening,

- Ocular symptoms non specific Redness, pain, photophobia and blurred vision.

- The interstitial keratitis is bilateral and starts as peripheral and superficial nummular lesions before developing deeper multifocal stromal nodules and later vascularization





The diagnosis is one of exclusion and based on positive ocular and otological features

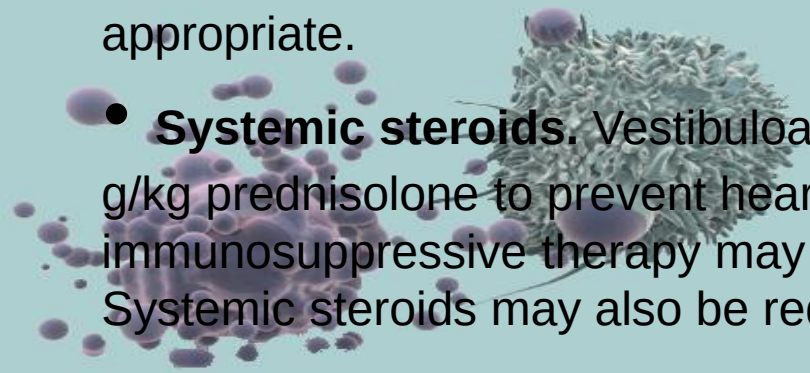
Investigations

○ Erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) may be elevated; elevated white cell count.
○ Antibodies to inner ear antigens may be detectable.
○ MRI may show inner ear and other abnormalities.

Treatment

- **Topical steroids** for keratitis, with additional measures as appropriate.

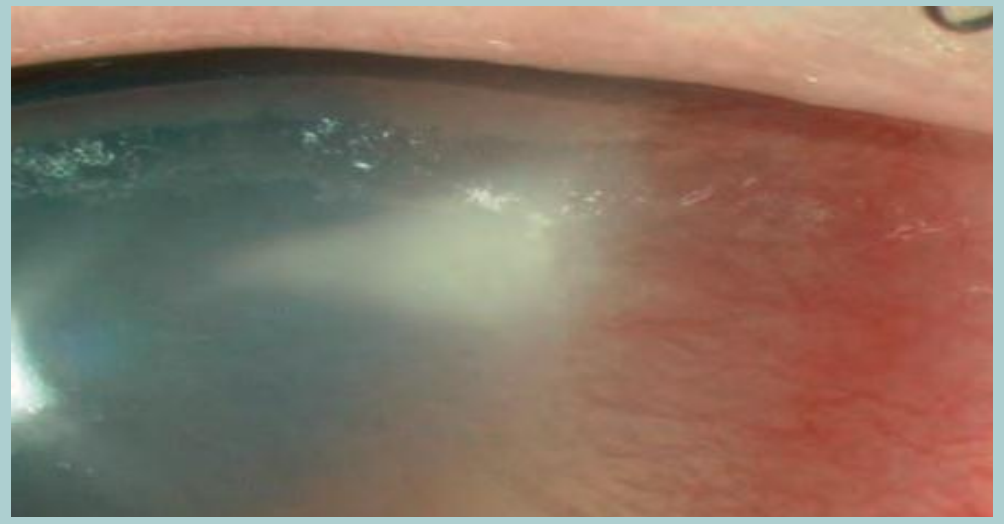
- **Systemic steroids.** Vestibuloauditory symptoms require immediate treatment with 1–2 g/kg prednisolone to prevent hearing loss.
immunosuppressive therapy may also be required.
Systemic steroids may also be required for scleritis or retinal vasculitis



case

A 12-year-old female complains of blurry vision, foreign body sensation, and photophobia in her right eye. While she has had chronic problems for years in both eyes, she has noted the acute onset and progressive worsening of the symptoms in the right eye for one week.

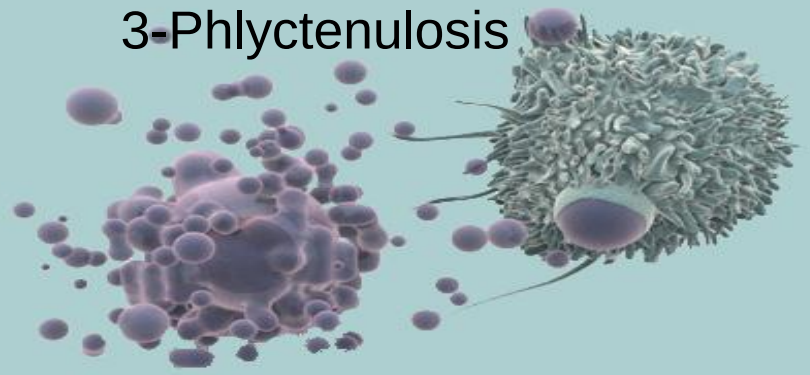
Past Ocular History: Since the age of 6, the patient has struggled with meibomian gland dysfunction and chronic staphylococcal blepharoconjunctivitis, as well as recurrent chalazia.



Staphylococcal hypersensitivity disorders

- There are a variety of disorders that occur in patients with coexisting blepharitis or H/O recurrent chalazia and styes.
- The exact pathophysiology: not known but type IV hypersensitivity reaction to staphylococcal antigens is believed to be responsible.

These disorders include:
1- Staphylococcal hypersensitivity syndrome
2- Marginal keratitis
3-Phlyctenulosis

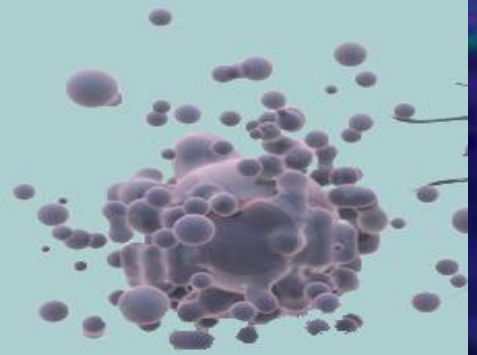


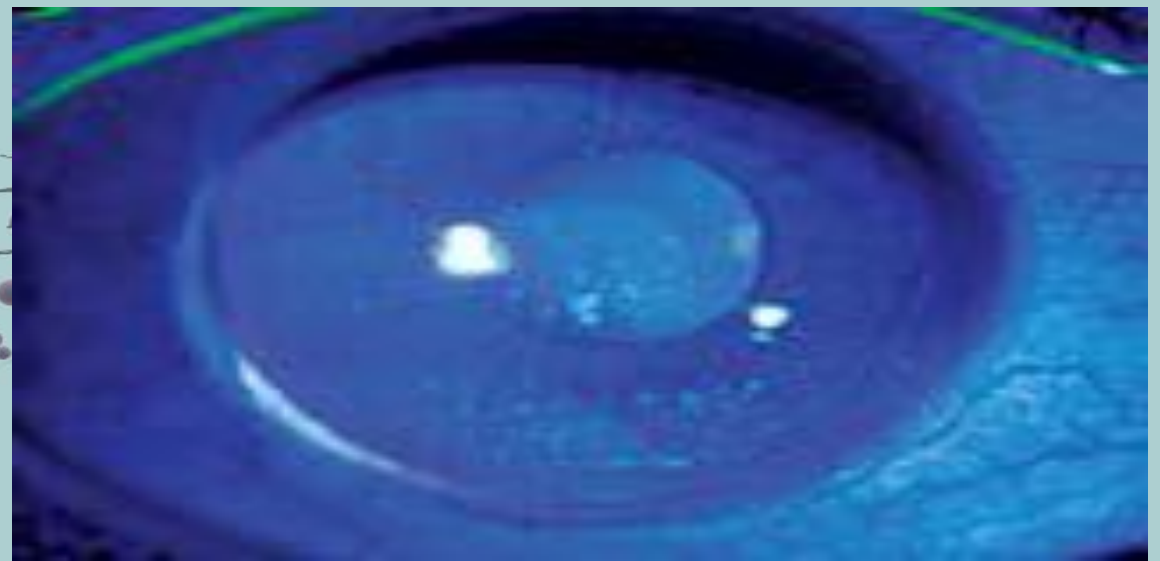
Staphylococcal hypersensitivity syndrome

- conjunctival injection and a punctate epithelial keratopathy that predominantly affects the inferior cornea and Conjunctiva. Occasionally a diffuse keratopathy can occur.
- The condition can be bilateral, asymmetrical, or unilateral. The extent of corneal involvement can be significant despite only mild eyelid disease.
- Symptoms range from mild irritation to foreign body sensation, watering, and photophobia.

Treatment

Treating the blepharitis together with a short course of topical steroids depending on the degree of inflammation.



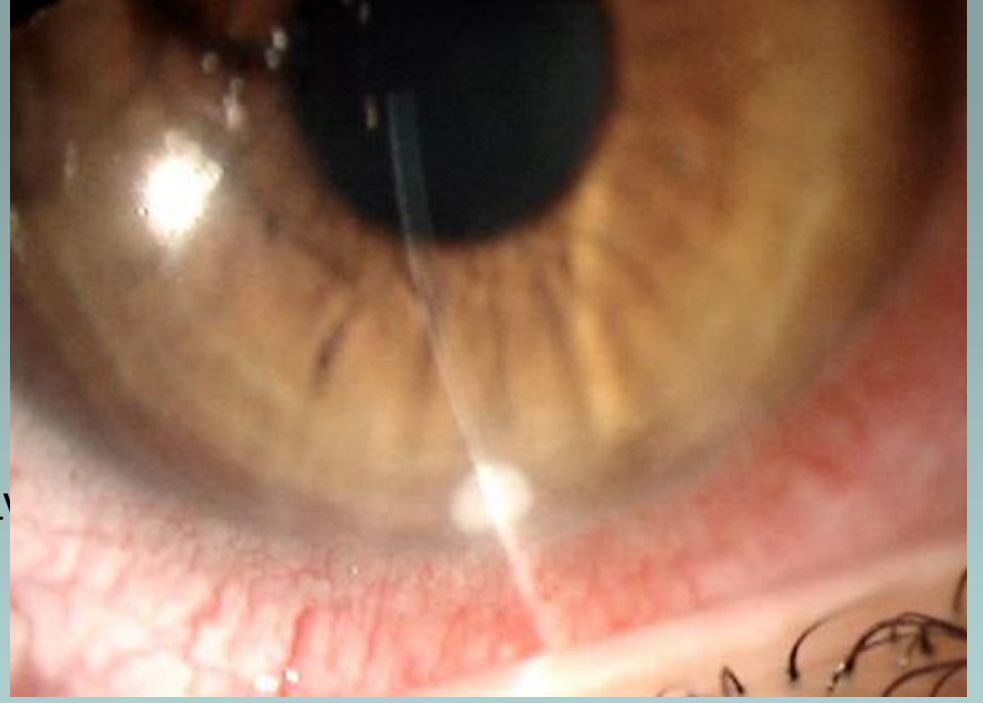


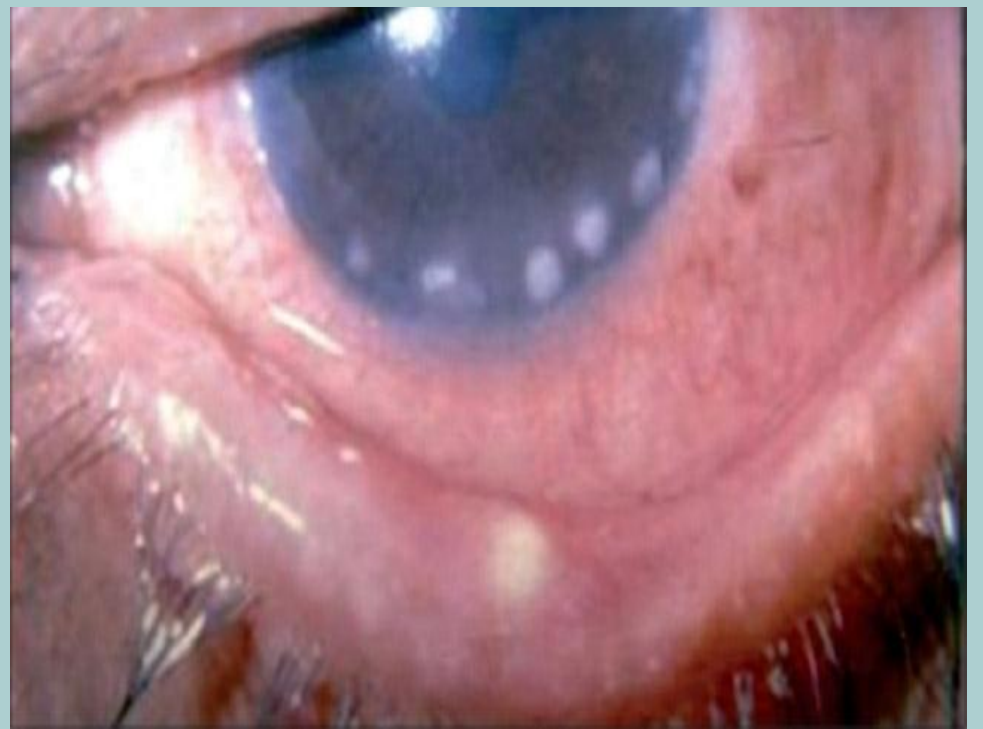
Marginal keratitis

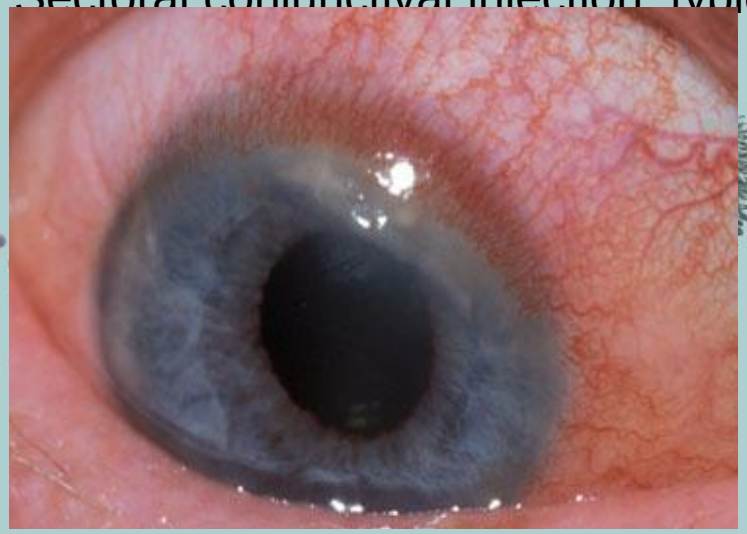
Symptoms
are mild irritation, watering, redness, and photophobia

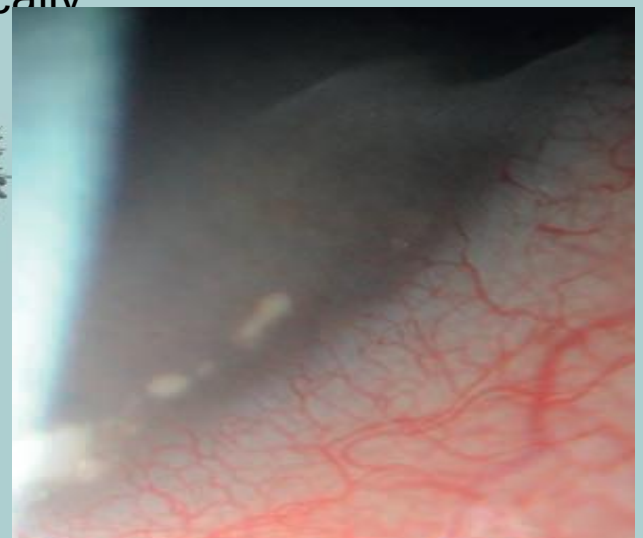
This classically presents as a grey-white anterior stromal sterile corneal infiltrate. It commonly occurs on the peripheral cornea, leav a 1 mm clear area between the infiltrates and the limbus

Initially the epithelium is intact but it eventually breaks down after prolonged inflammation and the ulcer spreads circumferentially with small blood vessels growing towards the infiltrate.
Sectoral conjunctival injection, typically

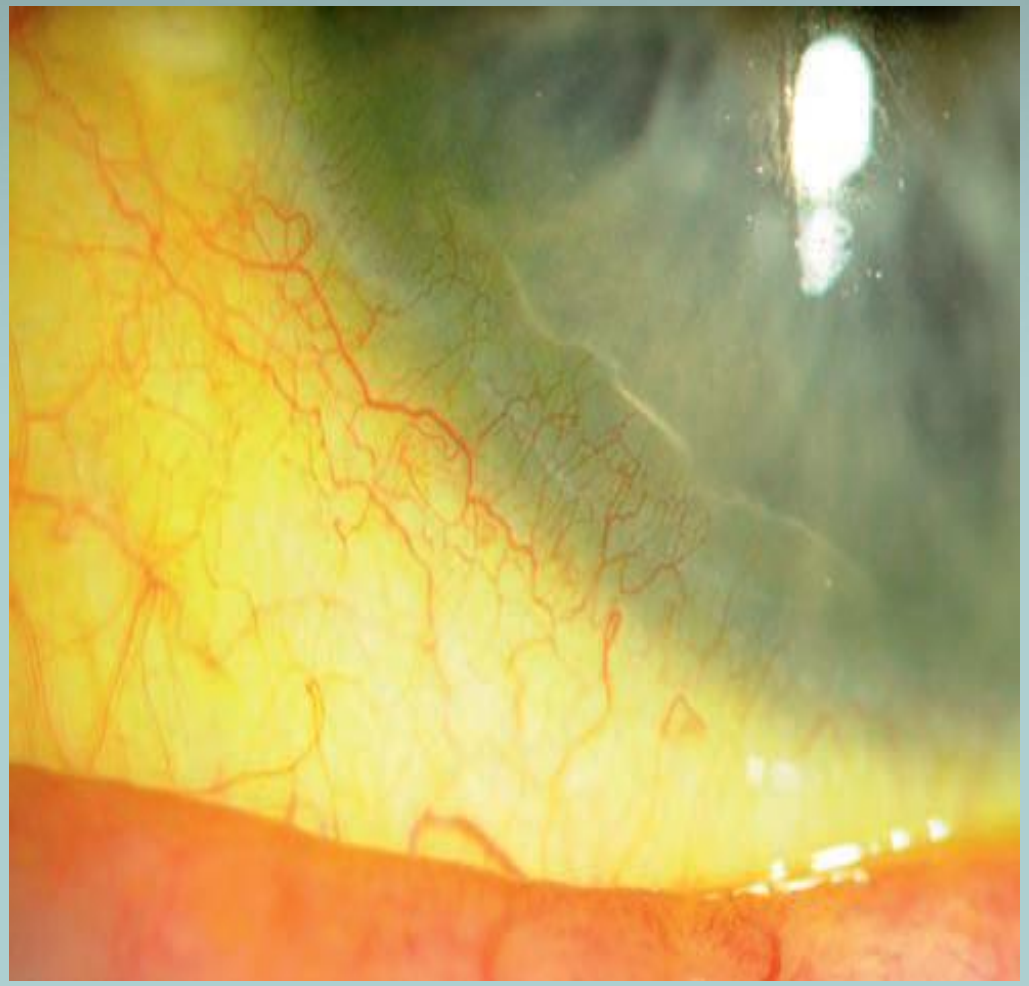








Treatment



Without treatment, resolution generally occurs in 1–4 weeks, depending on severity. Occasionally there is residual superficial scarring and slight thinning with mild pannus.

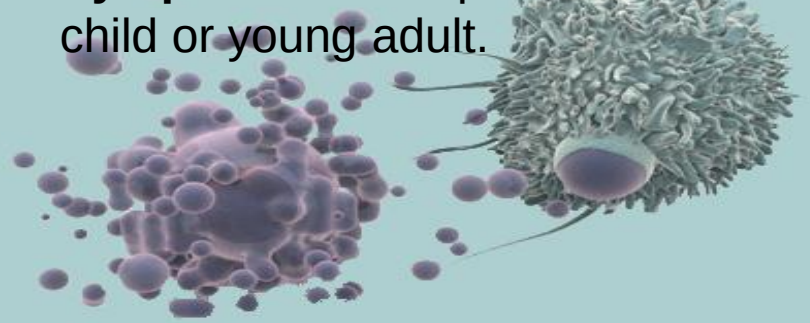
- A weak topical steroid such as fluorometholone or prednisolone 0.5% is instilled four times daily for 1–2 weeks, combined with a topical antibiotic.
- An oral tetracycline course (erythromycin in children, breastfeeding and pregnancy) may be required for **recurrent** disease.
- Blepharitis is treated as necessary.

Phlyctenulosis

- usually a self-limiting disease but may rarely be severe.
- Most cases in developed countries.

Etiology :
occurs commonly in children and young adults as a result of type IV hypersensitivity to microbial antigens (most commonly *Staph. aureus* but also *Mycobacterium tuberculosis* in endemic areas).

Symptoms. Photophobia, lacrimation and blepharospasm, often in a child or young adult.



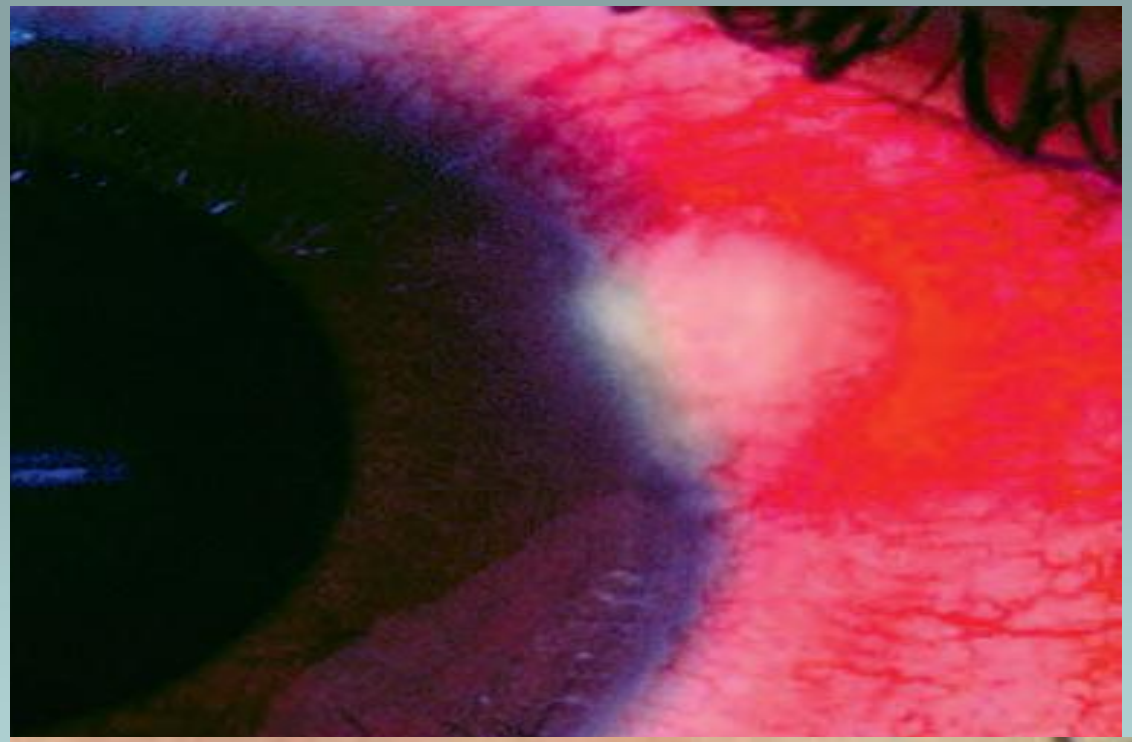
Signs

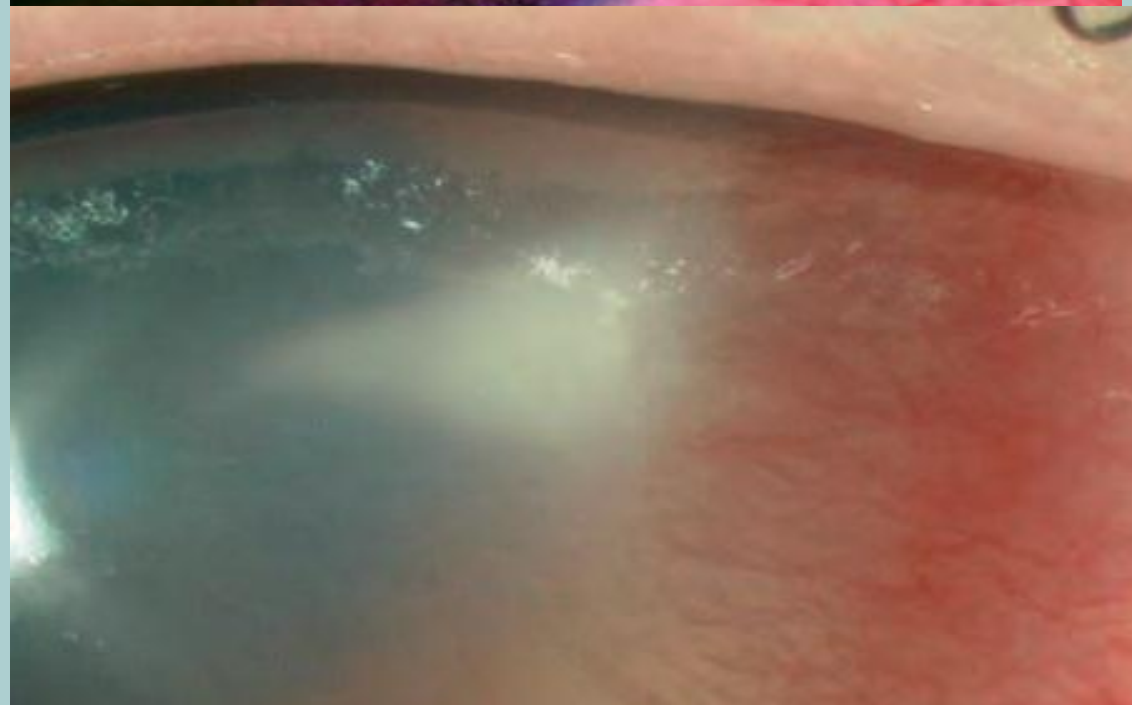
○ A small white limbal or conjunctival nodule associated with intense local hyperaemia.
○ A limbal phlycten may extend onto the cornea.
○ Spontaneous resolution usually occurs within 2–3 weeks; a healed lesion often leaves a triangular limbal-based scar associated with superficial vascularization and thinning but occasionally severe thinning and even perforation can ensue.
○ Very large necrotizing or multiple (miliary) lesions may occur.

Investigation for tuberculosis is generally indicated only in endemic areas or in the presence of specific risk factors

Treatment

This is as for marginal keratitis.





These topics have been covered:

1- TYPE II: ocular cicatricial pemphigoid
2- TYPE III: Stevens-Johnson syndrome – peripheral ulcerative keratitis & Mooren ulcer.
3- TYPE IV: interstitial keratitis and Cogan syndrome -
staph hypersensitivity disorders : staph hypersensitivity syndrome, marginal keratitis, phlyctenulosis.

Thanks for listening