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UVEITIS CASES #3: DIAGNOSIS AND TREATMENT

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COVID Vaccine #2

HAPPY 2021 Y'ALL!

Thanksgiving 2020

Pre-COVID! Those were the DAYS!

Best COVID vacation EVER!

VON's VISION Children's Event 2020

WHAT IS UVEITIS?

Eye inflammation which affects the uvea, which is the middle and vascular layer of the eye.

There are more than 30 varieties.

Pattern recognition!

Symptoms include:

- Redness, pain, photophobia, blurred vision, floaters, decreased vision, headache, no symptoms

WHY DOES THE LOCATION OF THE INFLAMMATION MATTER?

Can help determine etiology, focus the work-up, and determine treatment

Anterior (most common, 2/3):

- Iritis
- Keratitis
- Scleritis

Intermediate:

- Cells in vitreous, snowbanks or snowballs, +/- CME

Posterior:

- Vasculitis, Retinitis/Choroiditis
- Vitritis
- CME

Panuveitis:

- All or several of the above

WHAT DO I LOOK FOR IN REFERRAL NOTES?

Labs are greatly appreciated and makes initial referral exam much more productive for patient, though NOT REQUIRED.

In referral exam note:

- Granulomatous vs non-granulomatous (KP)
- Always check IOP, consider OCT RNFL esp if history of steroid use
- Dilate that day (break synechiae, help symptoms, examine retina and vitreous)
- If decreased VA, OCT macula
- If posterior, fundus photo/Optos/dilated fundus exam

HELPFUL DIAGNOSTIC TESTING IN UVEITIS

VA

IOP

Optos/DFE

media quality

retinal and optic nerve appearance

OCT macula and RNFL

Fluorescein Angiogram

Therapeutic trial with certain meds can be diagnostic

LAB TESTING

Non-Granulomatous

- HLA-B27 (especially if severe, hypopyon, plasmoid)
- ANA
- RF
- ESR, CRP
- ACE, lysozyme
- If pedi, Urine testing, Cr

Granulomatous

- T. pallidum IgG/IgM, +/- RPR, toxoplasma
- ACE, (lysozyme)
- Quantiferon Gold/PPD
- CXR and PPD or gamma interferon
- Ask about cold sores, shingles in the past
- What about HSV, VZV Ab testing?

LISTEN TO THE HISTORY

The history is VERY important.

Signs and symptoms:

- Redness
- Pain vs no pain, headache?
- Photophobia
- blurred vision, metamorphosia
- floaters?

Timing: Sudden? Slow onset? How long ago? After fever/malaise? After starting a medication?

Other parts of body involved? Arthritis, rash (on what body part, half of face and scalp?), kidney issues, sinus infections

CASE 1:

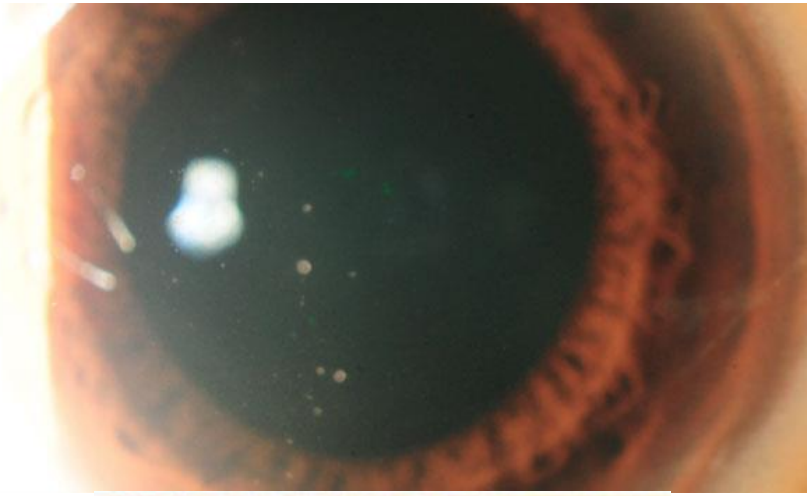
65 year old man with long history of Posner-Schlossman syndrome AKA glaucomatocyclitic crisis of the left eye.

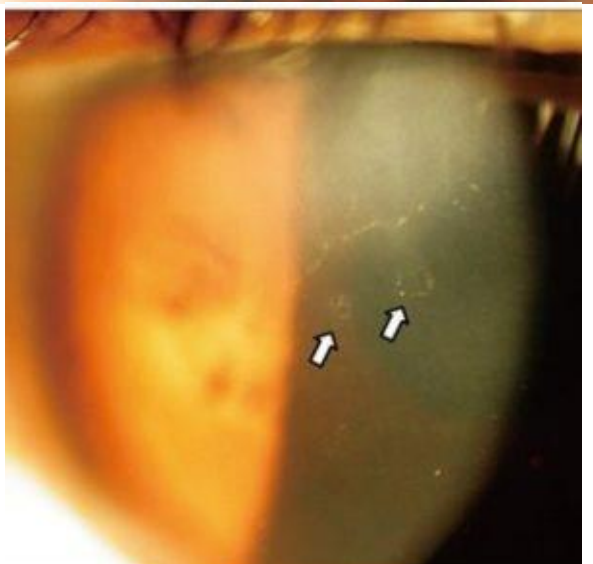
By the time I met him, his left optic nerve was cupped to 0.95, very abnormal visual field OS, 20/400 VA OS

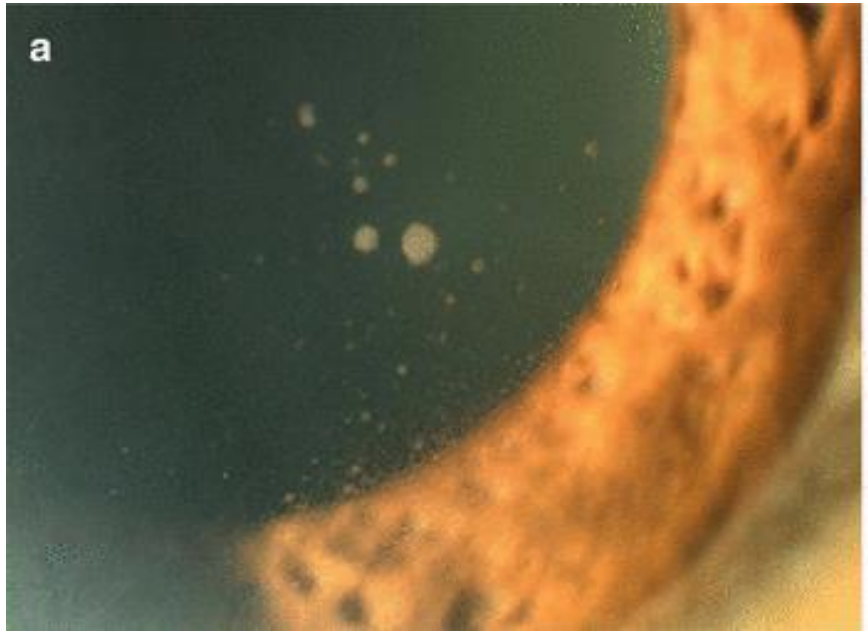
He reports IOPs in 50-60 range which he would treat with glaucoma drops and some PF drops

Mild pain, blurred vision and haloes, with flare ups. He would have about 1 flare up per month for the past 25-30 years.

CMV IRITIS







CMV IRITIS

Cytomegalovirus (CMV) is part of the Herpesviridae family of DNA viruses, which also includes herpes simplex virus (HSV), varicella-zoster virus (VZV), and Epstein-Barr virus

Can cause keratitis, anterior uveitis, scleritis, and retinitis. Herpetic anterior uveitis commonly causes endotheliitis, stromal and epithelial keratitis, and iris stromal atrophy, as well as an increase in IOP, likely due to trabeculitis-related impairment of aqueous outflow, iris atrophy or iris heterochromia.

CMV more commonly in immune deficient patients (CMV Retinitis) but also in immunocompetent patient. CMV has been recognized as a rare cause of persistent anterior uveitis accompanied by severe increase in IOP

CMV IRITIS

Recent studies have demonstrated a high rate of CMV PCR positivity in the anterior chamber taps of patients with PSS, and nearly all reported PSS patients improve with the addition of oral valganciclovir to their antiglaucoma medications. Thus, it is important to maintain a high index of suspicion for CMV anterior uveitis—and to consider PCR testing—in all cases of suspected PSS.

Induction regimen of valganciclovir (900 mg twice daily) is used; and as the disease becomes quiescent, the drug can be maintained at a once-daily dosage of 900 mg.

Rare side effects include bone marrow suppression and renal toxicity. For patients on chronic treatment thus check CBC with Diff and Cr every 2-3 months.

Cytomegalovirus

Hypertensive iridocyclitis

E C Kim, T P Margolis

A new ocular presentation of cytomegalovirus?

The single most common diagnosis assigned to patients with uveitis is "idiopathic," and in a recent large epidemiological study 48% of new cases of uveitis were assigned this diagnosis.[1] However, with the development of new diagnostic technologies, the discovery of novel ocular pathogens, and the recognition that established ocular pathogens can present in previously unrecognised ways, fewer patients are now being diagnosed with "idiopathic" uveitis. Examples of novel ocular pathogens include *Bartonella henselae*,[2] *Borrelia burgdorfi*,[3] West Nile virus,[4] *Tropheryma whipplii*,[5] microsporidia,[6] *Baylisascaris procyonis*,[7] and the leptospires.[8] Examples of established ocular pathogens that can present in previously unrecognised ways include atypical presentations of toxoplasmosis,[9 10] herpesviruses as the cause of the acute retinal necrosis syndrome[11 12] and some cases of Posner-Schlossman syndrome,[13] and varicella zoster virus as the cause of progressive outer retinal necrosis.[14] In addition, two independent groups recently reported evidence linking rubella virus with Fuchs' heterochromic iridocyclitis.[15 16] In the current issue of the *BJO* (p 846), de Shryver *et al* present evidence in support of cytomegalovirus (CMV) as a cause of hypertensive iritis in immune competent individuals.[17]

CMV is an extremely common human pathogen, infecting about 80% of the adult population.[18] Following primary infection CMV establishes a lifelong latent infection in myeloid and dendritic cell progenitors.[19 20] Like the other herpesviruses, latent infection with CMV is characterised by a low level of viral gene transcription. However, in immune competent patients this chronic latent infection is usually kept in check by a well established immune response, but recent studies indicate that in about a third of latently infected patients the infection is inefficiently controlled.[21] For many years, it has been recognised that CMV can cause ocular disease in immune compromised individuals including neonates, transplant recipients, and patients with HIV/AIDS with low CD4 cell counts. CMV retinitis is slowly progressive, characterised by white infiltrates and retinal haemorrhage, with progression that often follows the retinal vasculature. There may be an accompanying vitritis, and iritis. The iritis is characterised by fine stellate keratic precipitates distributed diffusely over the corneal endothelium.

> **There is compelling evidence to rethink the established paradigms about CMV ocular disease**

Over the past 5 years, several groups have published case reports linking CMV to hypertensive iridocyclitis in four immune competent patients, a concept that challenges the current paradigm of CMV mediated ocular disease.[22–25] The cumulative evidence for CMV as a causative agent in these cases included polymerase chain reaction (PCR) detection of viral DNA in the aqueous humour, local CMV specific antibody production, and clinical response to ganciclovir but not to aciclovir. However, all three lines of evidence were not obtained for any single clinical case reported. In the current issue of the *BJO*, de Schryver *et al* present more complete evidence linking CMV to hypertensive iridocyclitis in immune competent patients. In this case series, the authors present five cases of chronic or recurrent hypertensive iritis in immune competent individuals which previously would have been labelled "idiopathic," but which the authors provide credible evidence in support of CMV as the causative agent. CMV DNA was detected from the aqueous in five of five patients, but not from appropriate negative controls. A CMV specific antibody response was detected in the aqueous in four of four patients tested and therapy with ganciclovir, foscarnet, or valganciclovir led to resolution of inflammation in all five cases. Although one might be concerned that any of the above findings alone might represent a false positive result, the combination of all three lines of evidence provides compelling evidence to rethink the established paradigms about CMV ocular disease.

This challenge to established dogma arrives at a time when there is increased recognition that CMV can be a cause of clinical disease in immune competent individuals. It is fairly well known that CMV is the most common cause of heterophile negative mononucleosis characterised by fever, malaise, liver function abnormalities, and an atypical lymphocytosis,[26 27] but there are also a number of case series implicating CMV as a causative agent of meningitis, colitis, hepatitis, dermatitis, haemolytic anaemia, thrombocytopenia, and pneumonia in immune competent individuals.[28–30] Furthermore, there is growing evidence of "subclinical" reactivation of latent CMV especially in the elderly[31 32] and in patients with atopic disease,[33] and that chronic CMV immunological challenge leads to immune dysregulation, including altered cytokine profiles, chronic cell mediated inflammation, and reduced T cell diversity.[34 35] Is it possible that such alterations in immune surveillance and response could put a patient at even greater risk for developing non-infectious forms of uveitis?

An important point that de Shryver *et al* did not raise in their paper is the possibility that the CMV detected in the eyes of some, or all, of their patients might have been a consequence of local immunosuppressive therapy rather than the primary cause of their ocular inflammation. This possibility needs to be considered given that latent CMV is present in monocytes that transit through ocular tissues, especially in eyes with inflammation, and that this latent state is continuously, but inefficiently, regulated by the immune system.[36] A recent case report by Saidel *et al* serves as an illustrative example of how local immune suppression can give rise to ocular CMV disease. In this report the authors described a case of CMV retinitis that developed in an immune competent diabetic patient following an intravitreal injection of triamcinolone acetonide for macular oedema.[37]

With the growing awareness that CMV may cause hypertensive iridocyclitis and with the increased use of diagnostic testing of aqueous from eyes with uveitis, it is likely that more cases of CMV hypertensive iridocyclitis will soon be described. As this occurs the dual challenge to our profession will be to keep an open mind as well as be sharply critical of the evidence presented. We look forward to further studies and discussion on the topic of hypertensive iridocyclitis and the role that CMV may have in the pathogenesis of this condition.

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CASE 2:

32 year old African American man with blurred vision and floaters in both eyes for the past year.

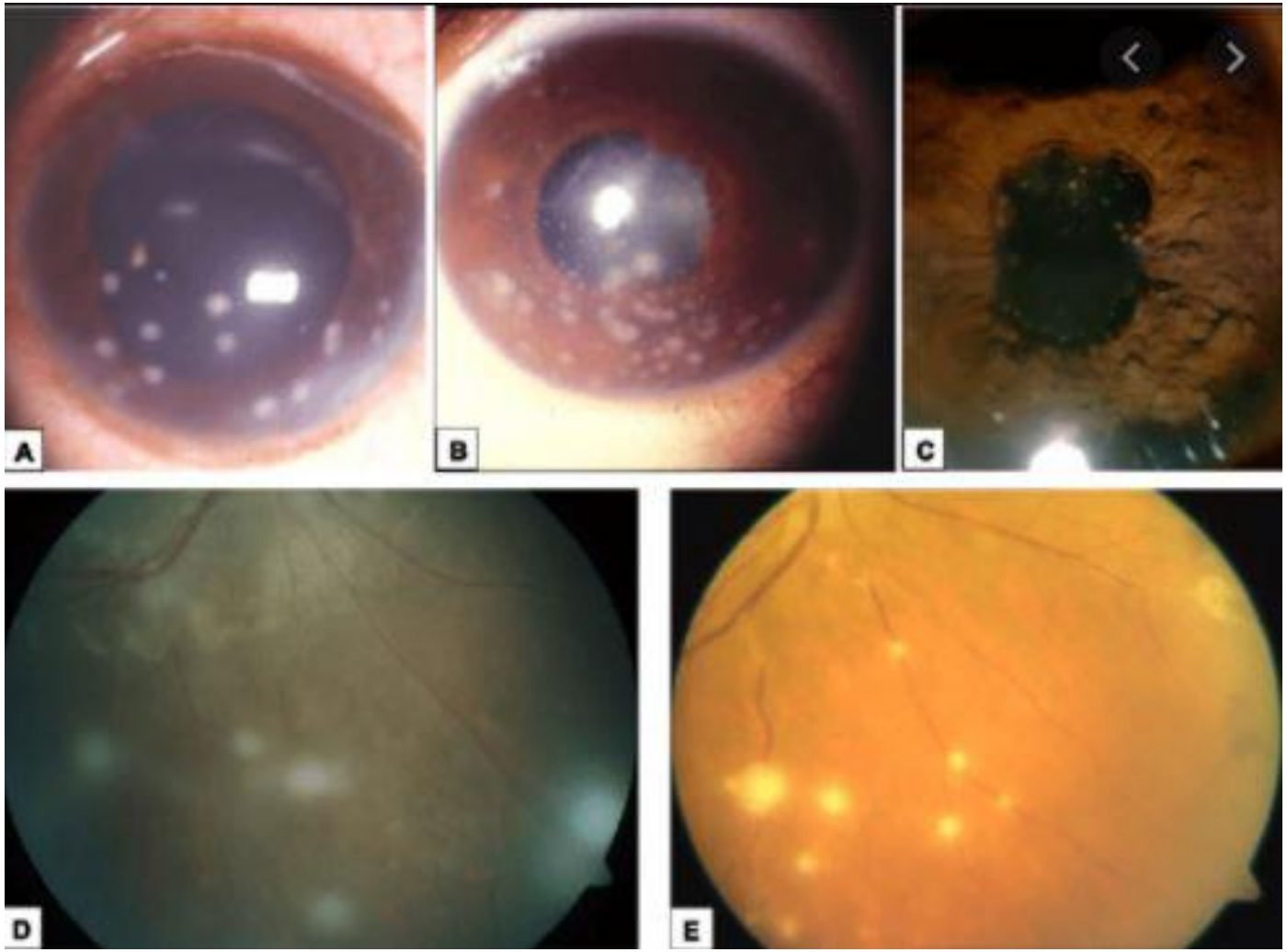
Slit lamp exam shows bilateral granulomatous severe iritis, mutton-fat KP, severe posterior synechiae and pupillary membranes OU.

Poor view of the retina due to posterior synechiae and pupillary membranes however appears normal.

He has noticed difficulty with breathing while playing basketball, which he's played for years and has never had problem like this before.

Has been stressed lately as his wife is expecting their first baby.

SARCOIDOSIS WITH UVEITIS



SARCOIDOSIS WITH UVEITIS: DIAGNOSIS

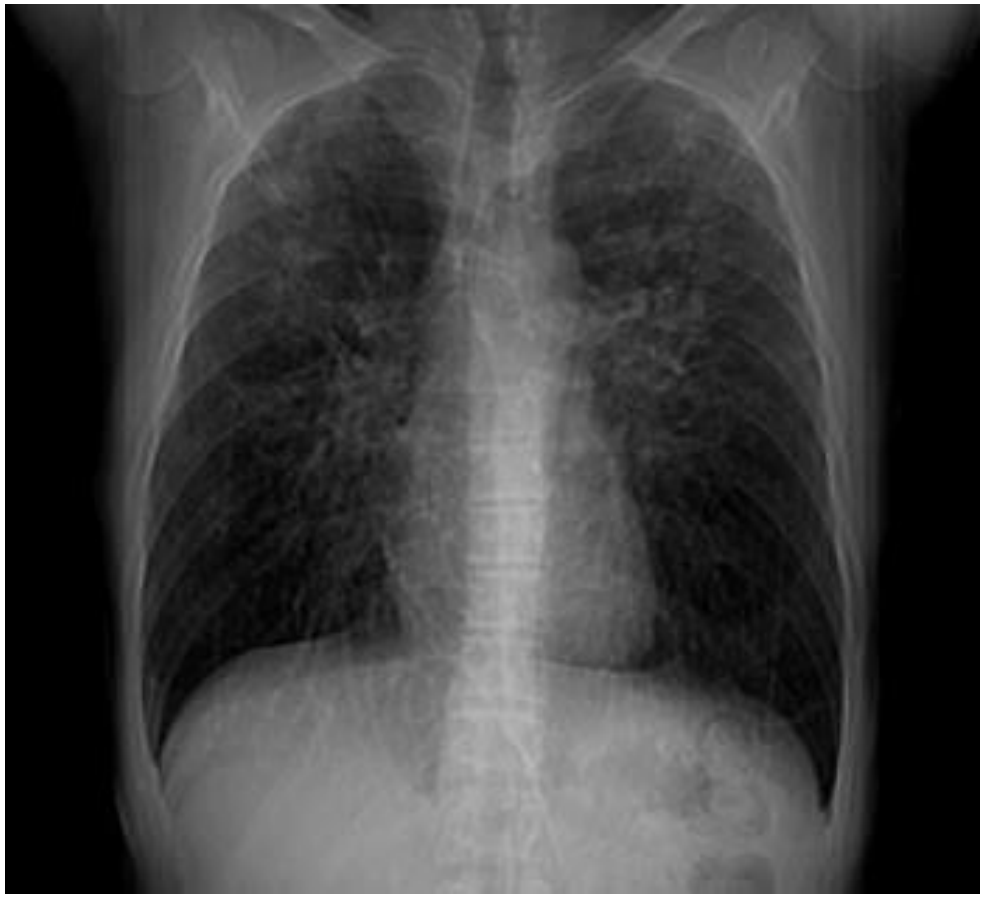
Conjunctiva: involved in 7-70% of ocular sarcoid. Most commonly presents as “millet-seed nodules” May lead to keratoconjunctivitis Sicca. The nodules can be biopsied to establish diagnosis of Sarcoidosis.

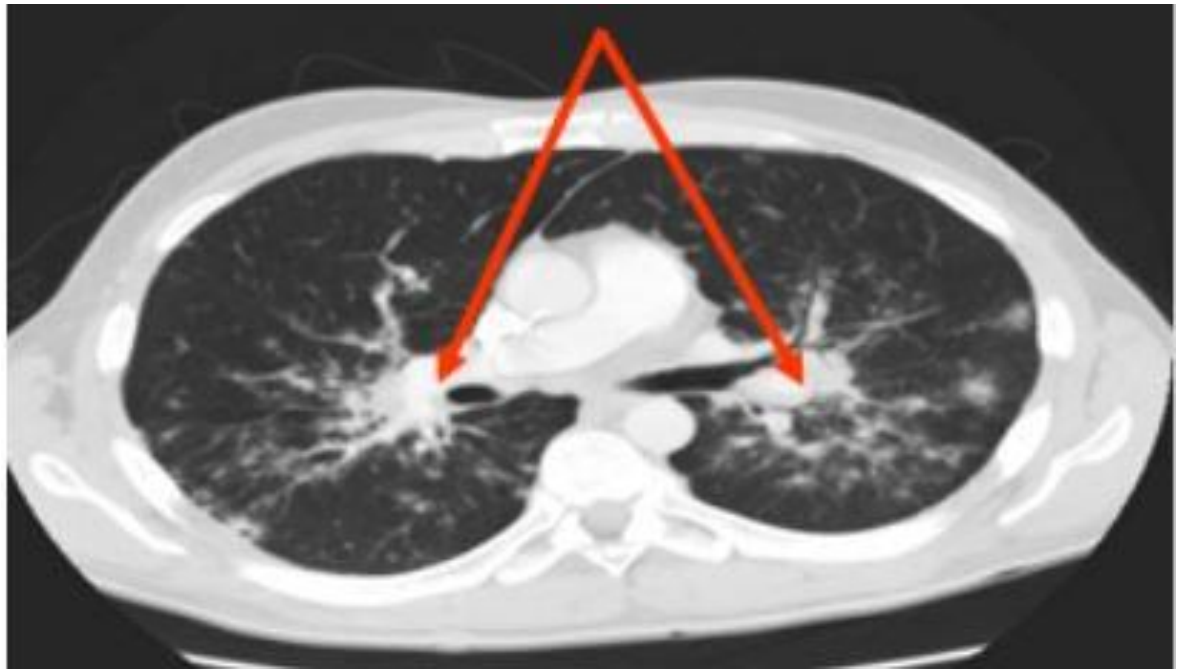
Anterior Uveitis: Anterior Chamber: (~85% of ocular sarcoid presentations). Most commonly chronic and granulomatous.

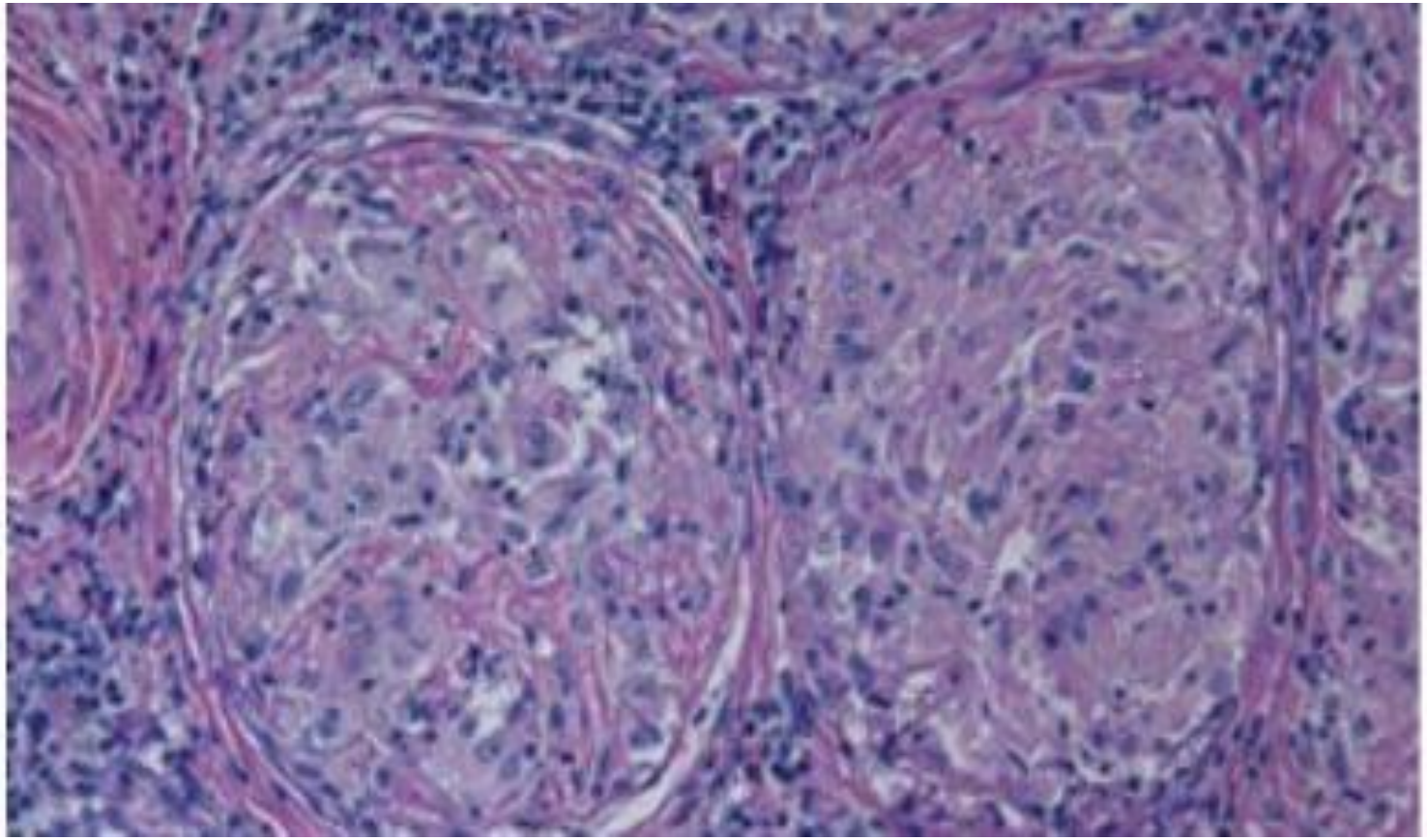
Complications from the uveitis include cataract, glaucoma, posterior synechiae, corneal band keratopathy, and Iris Nodules. Iris nodules can either be busacca or koeppe which are granulomas attached to the iris, or true iris nodules. New iris nodules signify acute inflammatory episode of ocular Sarcoidosis.

Posterior Uveitis: involved in 25% of ocular cases of Sarcoidosis. Most commonly involved are: periphlebitis: “candle-wax dripping”, vitritis, intermediate uveitis, panuveitis, posterior uveitis, exudative RD, pthisis; retinal vasculitis, CME, optic nerve edema.

SARCOIDOSIS WITH UVEITIS







SARCOIDOSIS WITH UVEITIS

Diagnosis is based on histological evidence of non-caseating granulomas consisting of histiocytes, epithelioid cells, and multinucleated giant cells which are surrounded by lymphocytes, plasma cells, and fibroblasts. (Figure 5 above).

SARCOIDOSIS WITH UVEITIS: DIAGNOSIS

Diagnosis is based on histological evidence of non-caseating granulomas consisting of histiocytes, epithelioid cells, and multinucleated giant cells which are surrounded by lymphocytes, plasma cells, and fibroblasts

Other helpful tests:

1) Chest X-ray: Lung involvement in Sarcoidosis.

2) Angiotensin converting enzyme (ACE) is produced by epithelioid cells and might serve a surrogate marker for granuloma load.

3) Hypercalcemia and hypercalciurea.

4) Anergy: to many antigens without increase in opportunistic infections due to compartmentalization of T helper cells and lack of delayed hypersensitivity reaction.

5) CT scan: superior to chest X-ray in identifying hilar lymph node involvement as well as pulmonary infiltrates.

SARCOIDOSIS WITH UVEITIS: TREATMENT

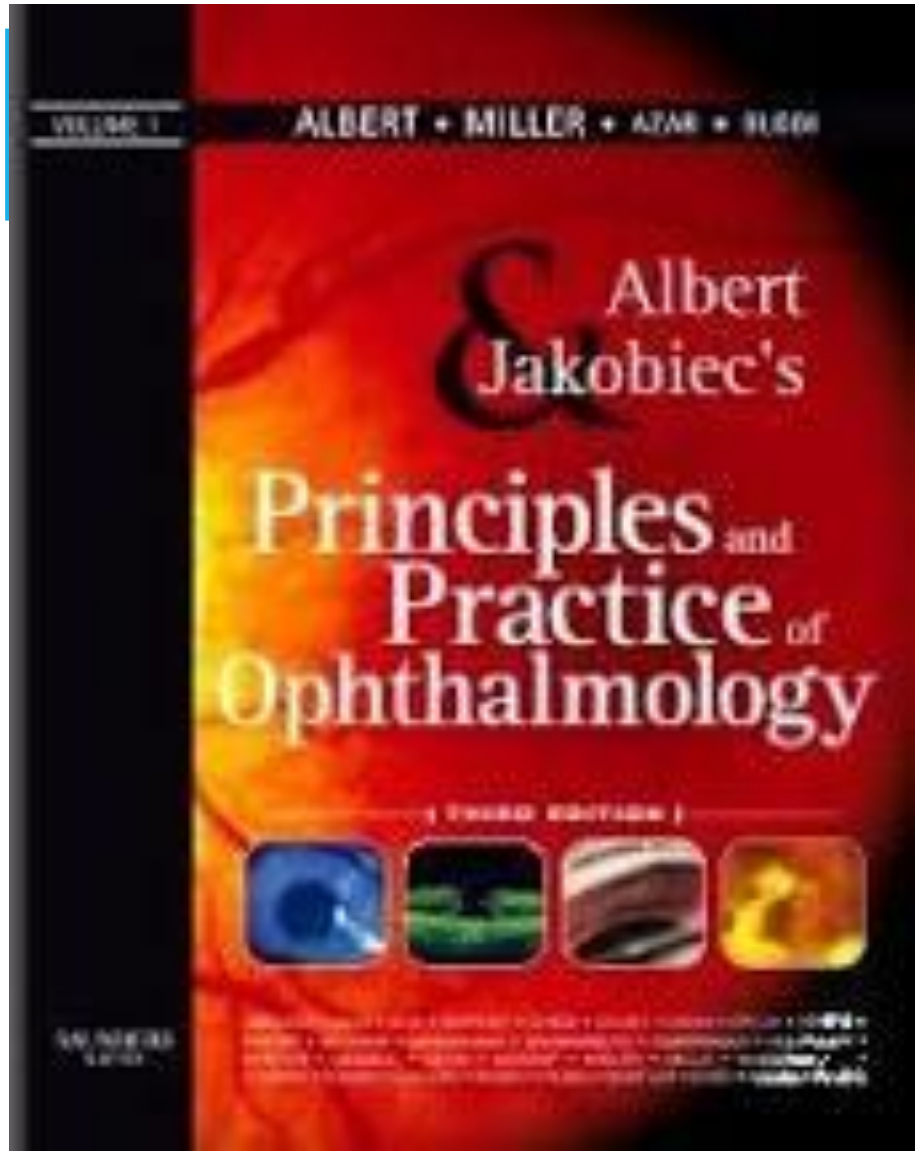
Depends on the presentation and severity of the disease.

If mild anterior uveitis then topical steroids and cycloplegics might be all that is necessary.

Systemic corticosteroids might be necessary in cases of non-responsive anterior uveitis; posterior uveitis; neovascularization symptomatic orbital disease; or optic nerve compromise.

If refractory to corticosteroids, oral NSAIDs can be added.

Finally, If inflammation persists, or in cases of steroid dependency; or significant side effects, then immunomodulatory therapy should be instated namely methotrexate, azathioprine, cyclosporine A, TNF-alpha inhibitors like Humira.



CASE 3:

4 year old girl with right eye non-granulomatous iritis of the right eye for 6 months. It improves with prednisolone drops but never quiets down completely.

The inflammation was found on eye exam for glasses so no symptoms of blurred vision or eye redness.

Has dealt with a sore a swollen knee which started when she was 3.

On exam, BCVA 20/20 OU, IOPs 24 OD, 12 OS

1+ cells OD, no cells OS, no lens changes OU.

No cells in vitreous, no CME OU, c/d 0.2 OU.

JUVENILE IDIOPATHIC ARTHRITIS (JIA) UVEITIS

JIA is the most common rheumatologic disease of childhood, up to 70% of arthritic disease in childhood.

Systemic: High grade fever, multiple extra-articular manifestations, Uveitis is rare.

Polyarticular: Five or more joints involved within 3 months of onset of disease, Uveitis is uncommon.

Pauciarticular: Less than five joints involved within 3 months of onset of disease, Highest association with uveitis.

Pauciarticular JIA, the most common subtype, comprises 40%-60% of all JIA cases. It is defined as the involvement of less than 5 joints during the first three months of disease. [2] Systemic symptoms, like fever or rash, are mild if present. The knees are the most common joints involved, but the small bones of the hands or feet may also be affected. Patients with the pauciarticular subtype are those at highest risk of developing uveitis.

JIA UVEITIS

Clinical Presentation:

The classic presentation is an asymptomatic, bilateral, non-granulomatous iridocyclitis in a white eye.

The arthritis typically precedes the onset of uveitis.

The latency between onset of arthritis and detection of uveitis is around two years.

However, the wide range of reported delay of onset of ocular inflammation may be due to the difficulty of determining the onset of an asymptomatic disease, as well as the occurrence of uveitis in a patient who is preverbal.

Band keratopathy commonly seen, glaucoma can be caused by posterior synechiae, peripheral anterior synechiae, and steroid-induced glaucoma.

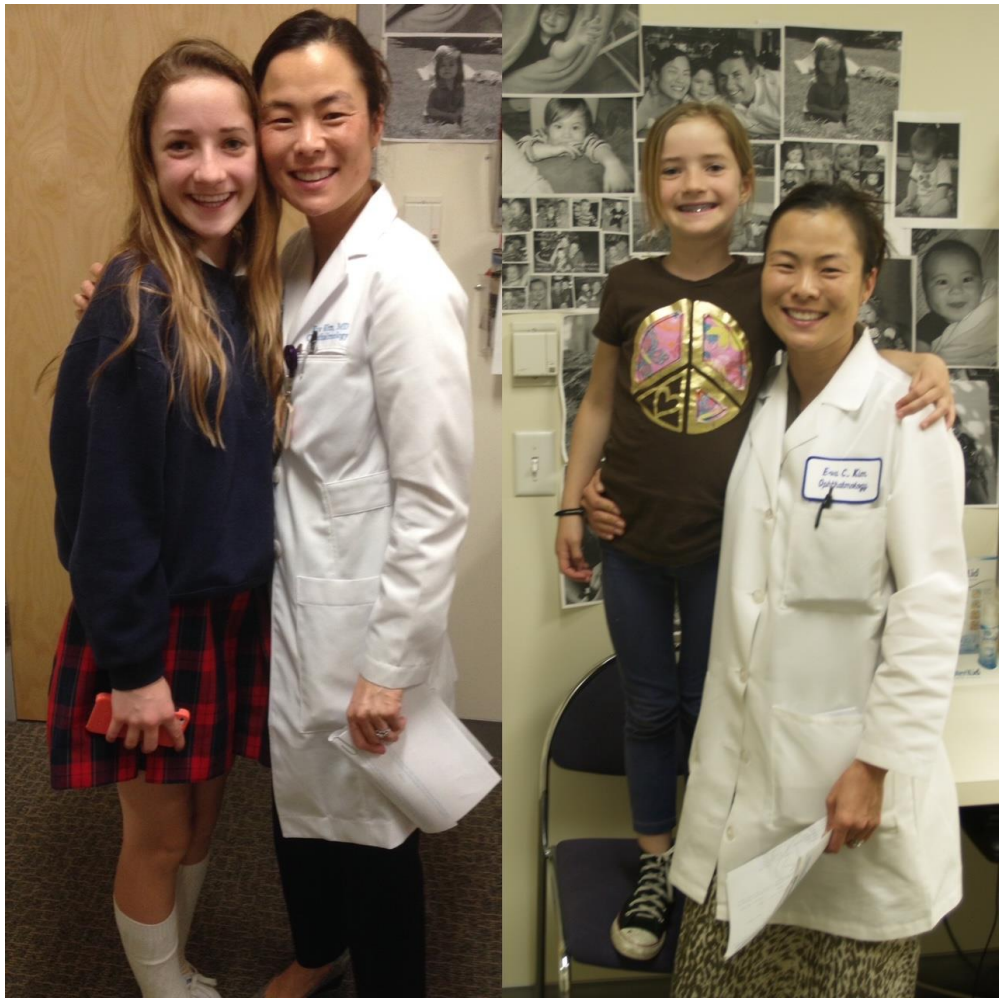
. In review of our JIA patients, cataracts were found in 71%, band keratopathy in 66%, and glaucoma in 30%. Cystoid macular edema developed in 37%

JIA UVEITIS: TREATMENT

TREATMENT:

- Knowing that patient was not able to quiet down on prednisolone alone for past 6 months and also has steroid-induced ocular hypertension, I decided to start a steroid-sparing immune-modulation treatment (IMT).
- 1/3 controllable with topical steroid, 2/3 require IMT
- Started on oral prednisone 20 mg per day
- Started on METHOTREXATE 15-25 mg per week (systemic NSAIDs, Azathioprine, Humira/Infliximab (Remicade))
- Tapered off prednisone over few months
- On Methotrexate for 6 years
- Once off methotrexate, never needed to re-start. Remission.

JIA UVEITIS



CASE 4:

28 year old man with red eyes, blurred vision, floaters, and photophobia

Both eyes affected

Exam showed bilateral granulomatous KP and 3+ cells OU, a couple iris granulomas OU

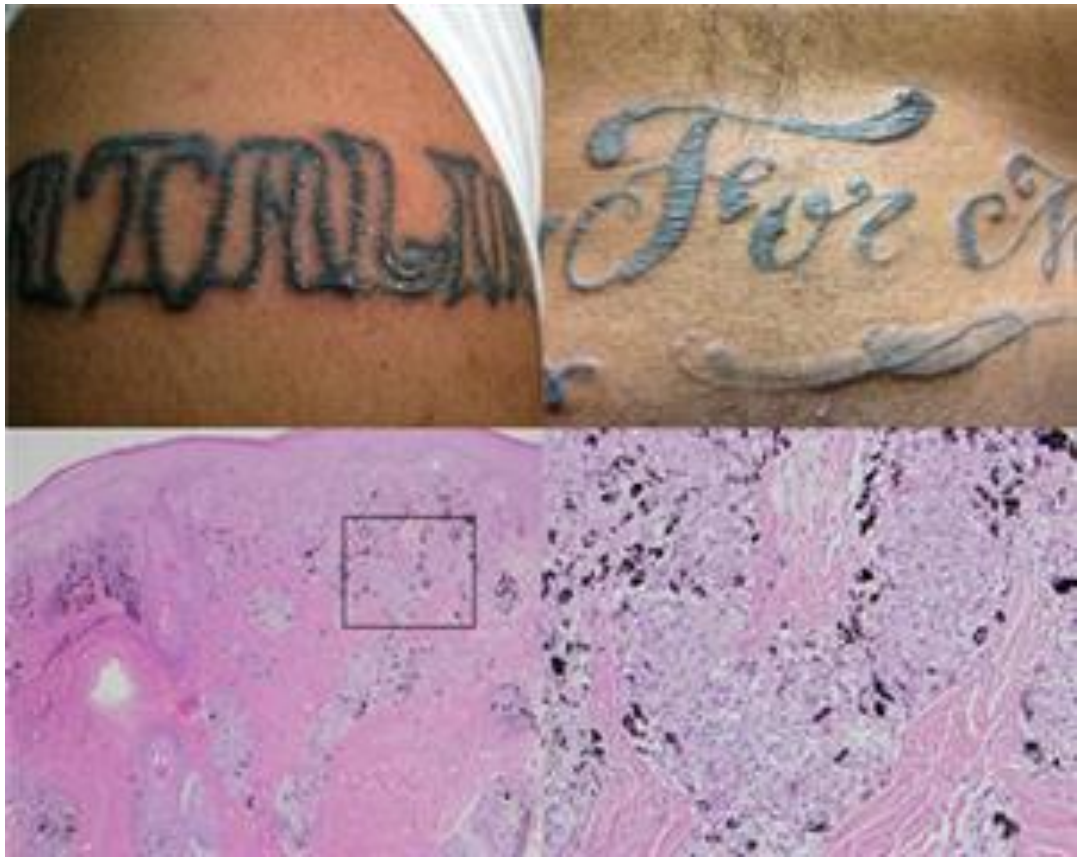
1+ vitreous haze with tiny granulomas in vitreous

Mild disc edema OU

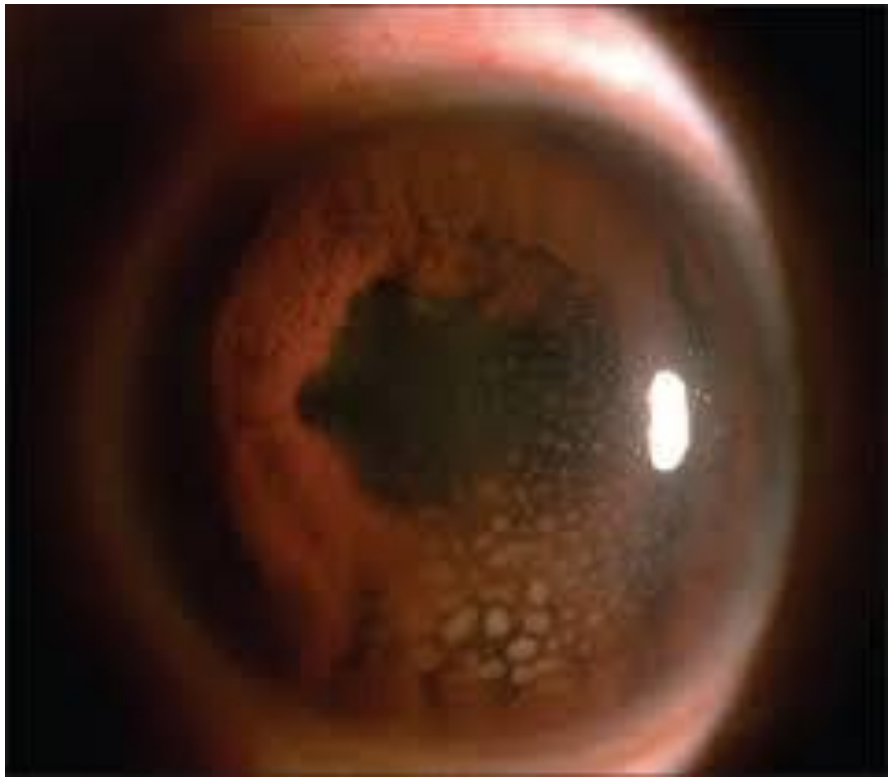
Few peripheral white Dalen-Fuchs like nodules OU

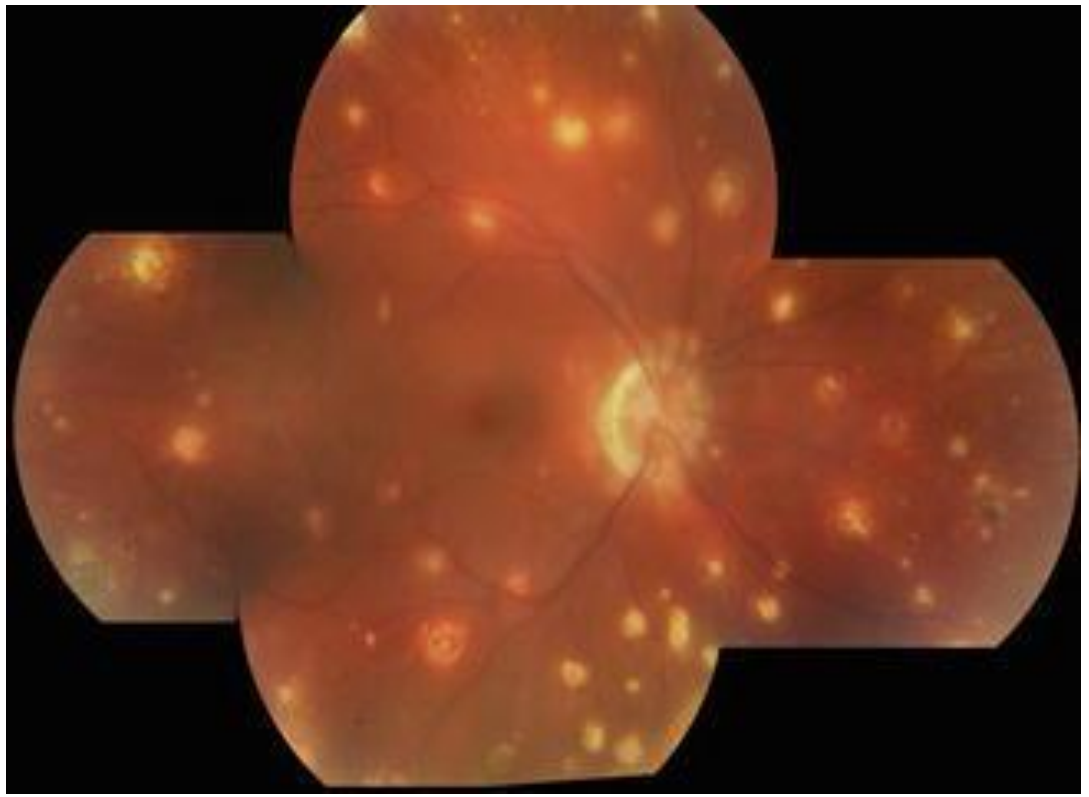
I noticed along both arms...

TATTOO-INDUCED UVEITIS



TATTOO-INDUCED UVEITIS





TATTOO-INDUCED UVEITIS

Raised and indurated skin, seen only with black tattoo ink

Can be non-granulomatous or granulomatous uveitis

Can be recurrent or chronic persistent disease

Uveitis starts aroud 6 months after the tattoo placed

Tattoo biopsy can reveal noncaseating granulomas with histiocytes surrounding black tattoo pigment in the dermis.

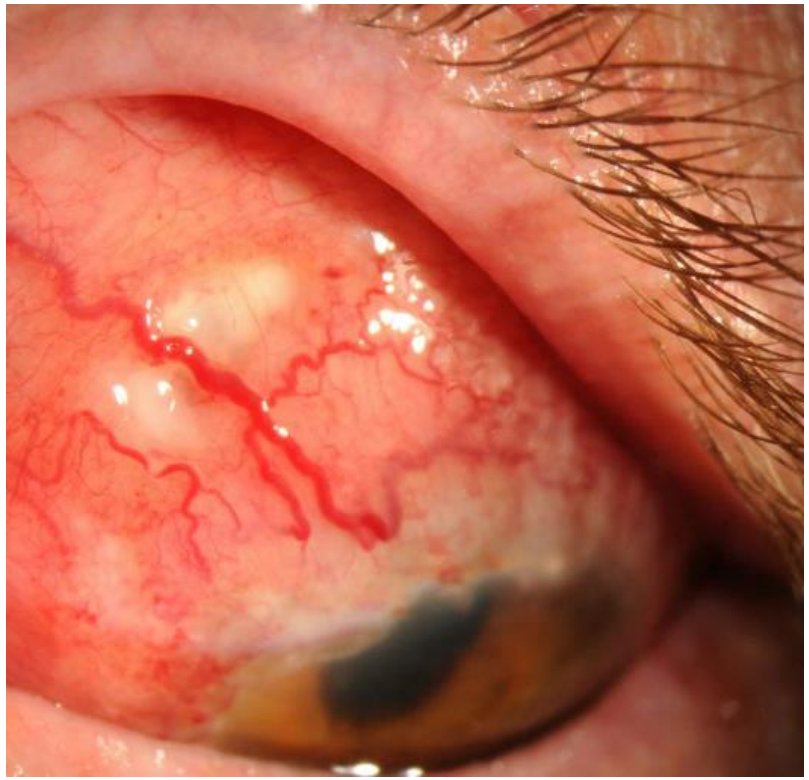
Vision-threatening complications of intraocular inflammation, include iris bombe, pupillary membrane, cystoid macular edema and glaucoma.

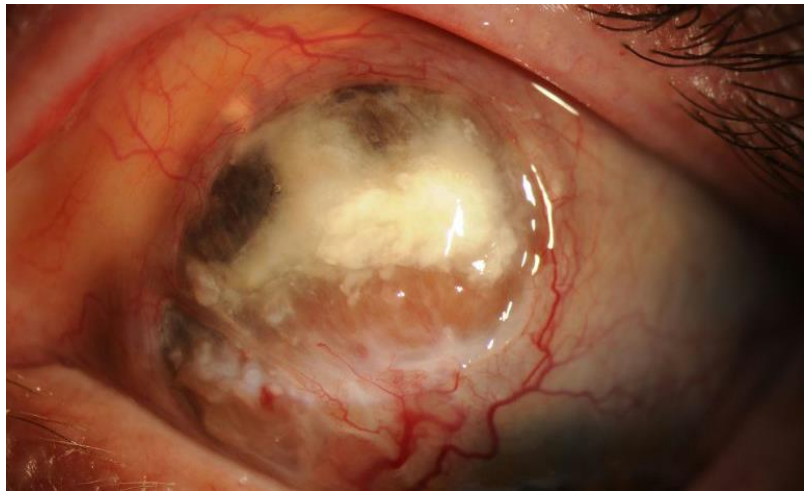
Treat with topical steroid, systemic steroid, and sometimes IMT.

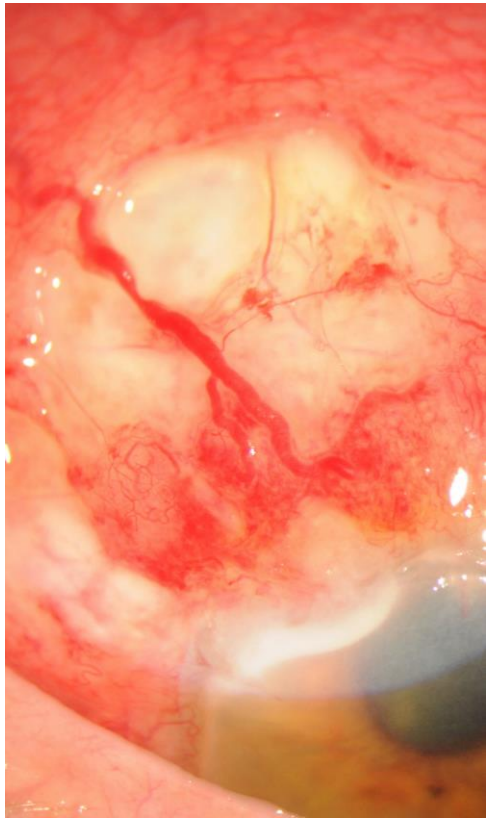
Removing tattoo does not help

CASE 5:

70 year old woman with severe 3+ injection of superior sclera of the right eye, severe TTP, almost appeared as an ulcer with corneal peripheral ulcerative keratitis







SCLERITIS DUE TO GRANULOMATOSIS WITH POLYANGIITIS (FORMERLY WEGENER'S GRANULOMATOSIS)

Rare, less than 200K cases per year in US

Vasculitis or Blood vessel inflammation in nose, sinuses, throat, lungs, kidneys, eyes

Can be fatal (lung and/or kidney failure)

Often ages 40-65

Symptoms: sinus infections, nose bleeds, cough +/- blood, SOB, fever, joint pain, numbness limbs, fingers, toes, weight loss, blood in urine, skin sires, eye redness/scleritis/iritis, hearing problems

Saddling of bridge of nose, DVT, kidney damage, hearing loss

DIAGNOSIS

Blood testing: ESR, CRP, C-ANCA, P-ANCA, CBC (for anemia), Cr

Urine testing: to look for blood or high protein levels

Imaging Studies: CXR, CT of Chest/abdomen/pelvis

Biopsy of affected tissue/organ

TREATMENT

Steroids such as Prednisone, Solumedrol (infusion, oral, topical)

Cyclophosphamide, Azathioprine, methotrexate, mycophenolate mofetil, Rituximab, plasmapheresis

Work with PCP and Rheumatologist

CASE 6

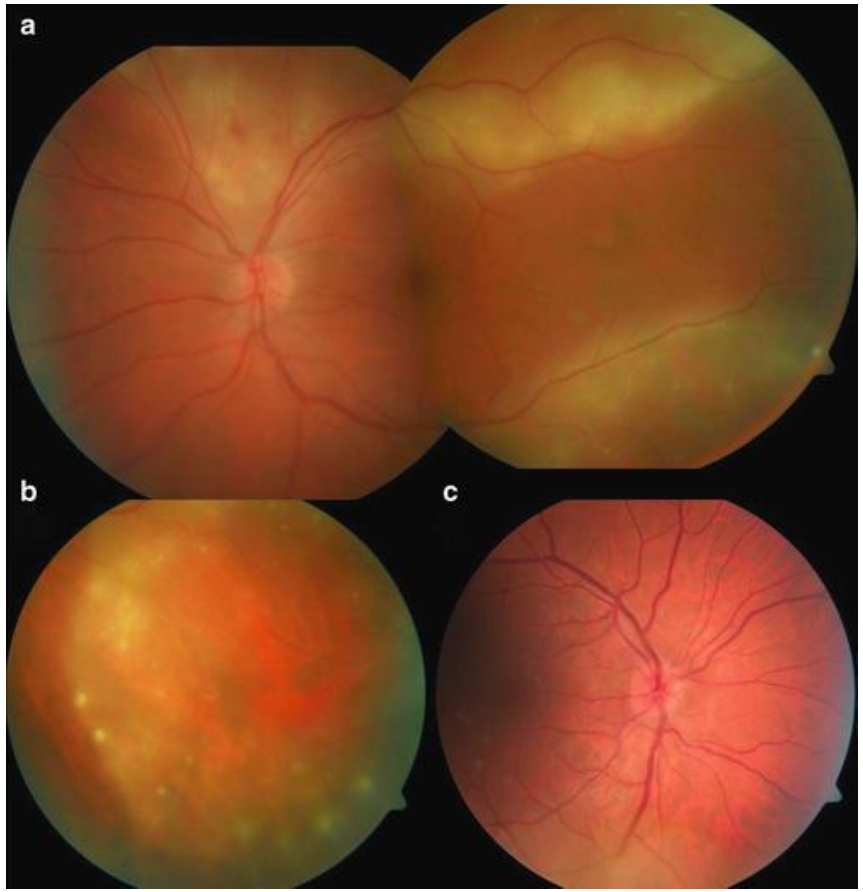
49 year old man with blurred vision and floaters for the past 3-4 weeks

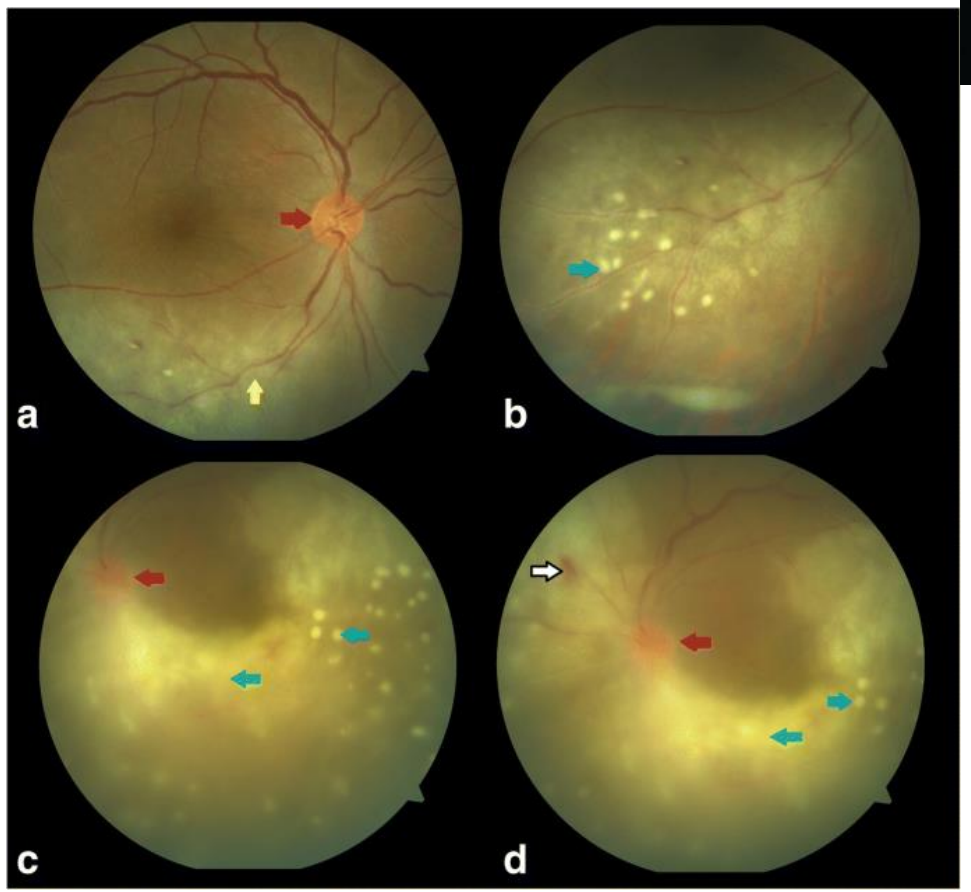
Eyes have been red on and off, very photosensitive

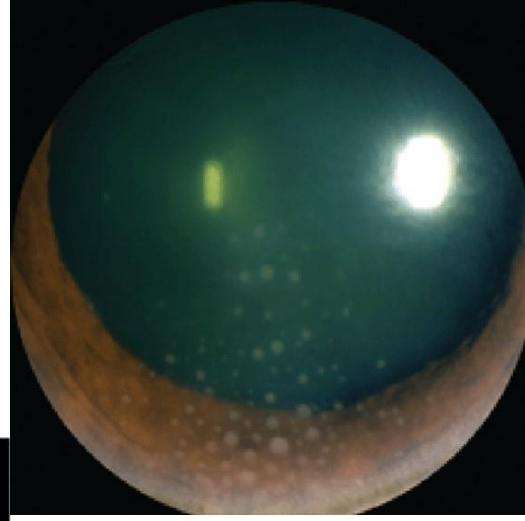
Feeling well overall, maybe some fatigue

SLE: Bilateral granulomatous panuveitis, granulomatous KP, poor view of the retina due to vitreous cells and granulomas

SYPHILIS PANUVEITIS







SYPHILIS PANUVEITIS: TREATMENT

Since bilateral granulomatous panuveitis with chorioretinitis, very suspicious for syphilis

+RPR, + FTA-ABS (need both non-trep and trep test)

Infectious Disease referral

Neurosyphilis, LP done, tested for HIV (was HIV+), always check HIV

Penicillin G 18-24 million units per day (3-4 million units IV every 4 hours or continuous infusion for 10-14 days): curable!

Topical prednisolone drops helpful for anterior uveitis, frequent follow up.

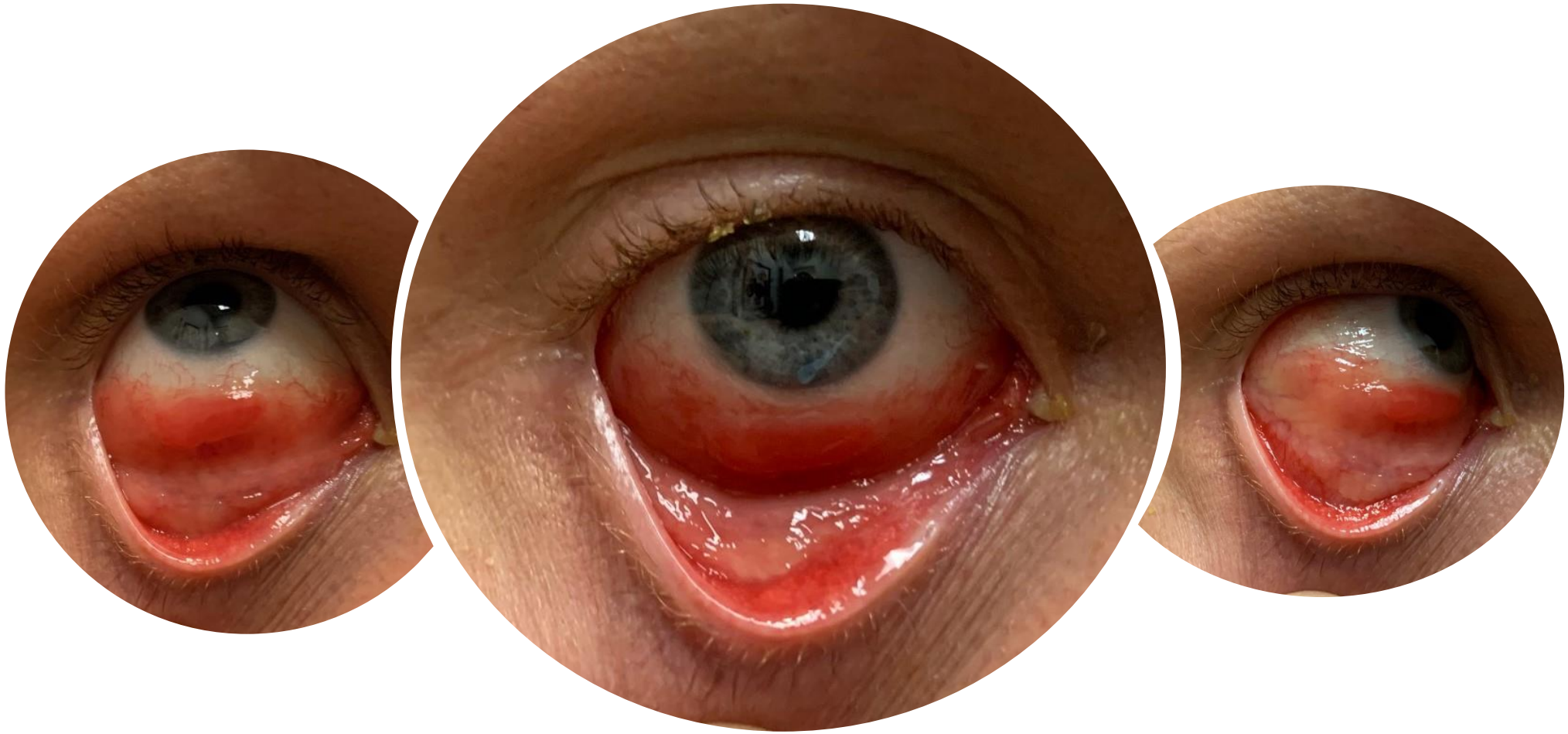
CASE 7:

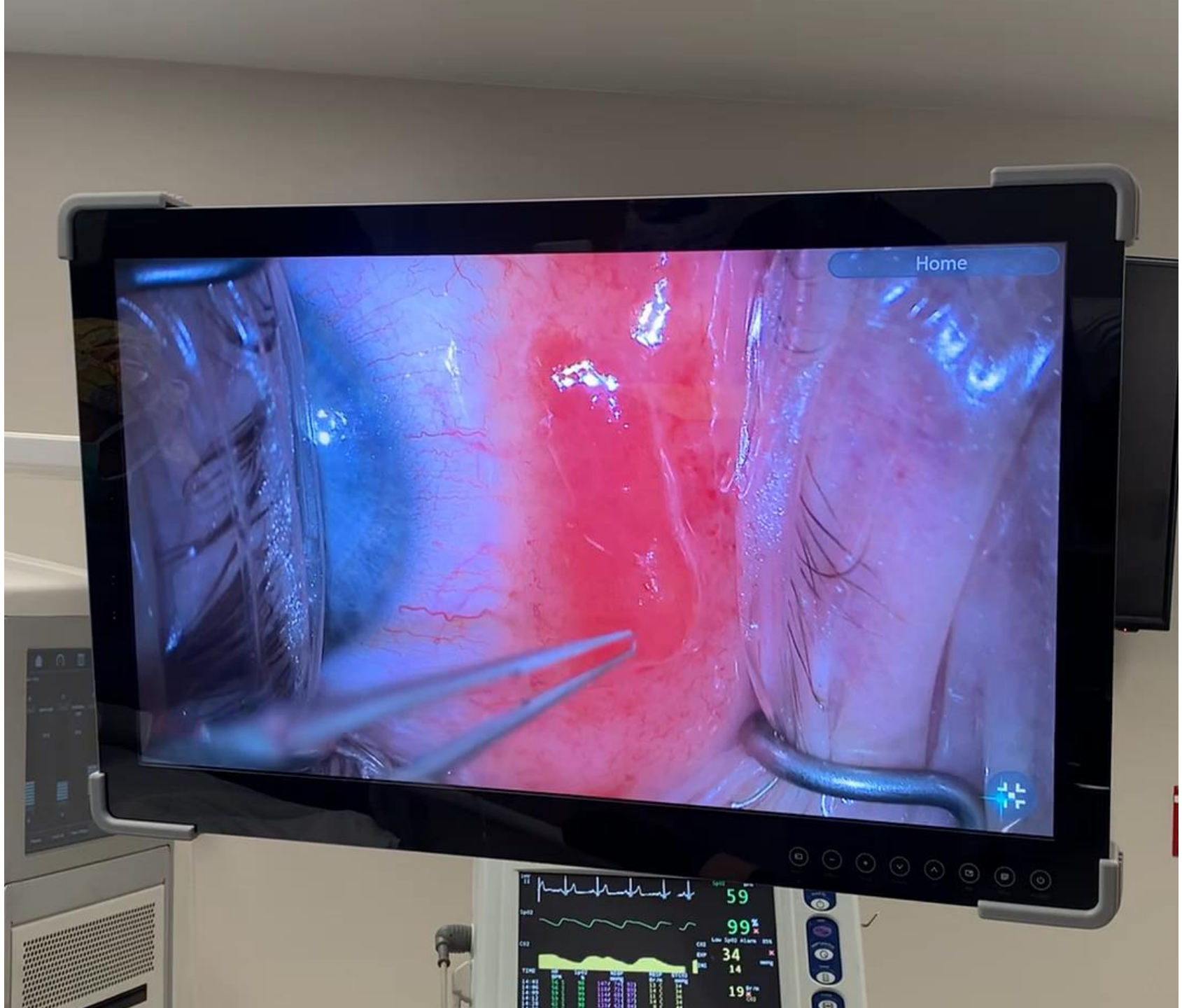
33 year old man referred by his optometrist for scleritis OD

He did not have any pain, only noticed that his right eye looked swollen or smaller than his left eye. No visual changes, no discomfort.

He did not even notice that his right eye had a reddish area inferiorly.

Poor historian so unable to tell me how long his eye was affected.

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CONJUNCTIVAL LYMPHOMA

Biopsy Results:

1. Right eye conjunctival biopsy: (pathology)

Atypical lymphoid population consistent with LOW-GRADE B-CELL LYMPHOMA (NON_HODGKIN LYMPHOMA)

2. Right eye conjunctival biopsy: (flow cytometry)

Monoclonal B-cells consistent with a B-CELL LYMPHOPROLIFERATIVE DISORDER

CONJUNCTIVAL LYMPHOMA

Ocular surface tumor that usually appears as a painless, salmon-pink, “fleshy” patch that is smooth or follicular.

Patients often have minimal symptoms, leading to an average delay of eight months between clinical onset and diagnosis.

Clinical complaints include conjunctival redness, irritation, and excessive tear production. Patients may also present with a palpable mass, ptosis, or diplopia.

CONJUNCTIVAL LYMPHOMA

Differential diagnosis

benign reactive lymphoid hyperplasia, benign ocular surface tumors (papilloma, pyogenic granuloma), malignant tumors (squamous cell carcinoma), scleritis, episcleritis, ectopic lacrimal gland, presence of a foreign body, amyloid deposition, and chronic follicular conjunctivitis.

Biopsy

Biopsy then send fresh tissue for flow cytometry and gene rearrangement studies in addition to formalin-fixed tissue analysis. Histopathologic evaluation and immunohistochemical studies are necessary to establish the diagnosis (flow cytometry).

Staging

Although conjunctival lymphoma typically presents with a localized lesion, a minority of patients have disseminated disease at presentation. Proper staging evaluation includes a complete history and physical examination, complete blood count, metabolic panel, serum lactate dehydrogenase levels, serum protein electrophoresis and $beta_2$-microglobulin levels, CT neck, chest, abdomen, and pelvis, MRI of the orbit and brain), and +/-bone marrow biopsy.

CONJUNCTIVAL LYMPHOMA

The mainstay of treatment for localized disease is external-beam radiotherapy (EBRT). The complete remission rate is in excess of 90 percent for MALT lymphoma, with excellent long-term local control in the majority of patients. Potential complications of EBRT include dry eye, keratitis, cataract formation, optic neuropathy, and retinopathy.

For systemic disease, Rituximab has shown efficacy in the treatment of conjunctival MALT lymphoma, with response rates between 50 to 87 percent. However, median time to recurrence was less than one year. Rituximab can also be injected into the conjunctival lesion.

5-year survival of 93%. Because nearly 20 percent of patients with ocular adnexal lymphoma eventually progress to disseminated disease, follow-up is essential and should be continued indefinitely. Follow-up is recommended every three months for the first year and every six months afterward.