Ocular Sarcoidosis Case Study
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History of Present Illness: 44 yo Male who presented with progressive decrease of visual acuity for the last year. This is in the context of progressive fatigue and poor appetite which has led to 25 pound weight loss in 12 months. He also states that, “I have swollen gland in my neck for two weeks.” Ocularly, he complained of blur, redness, and photophobia bilaterally.

POH: Glaucoma which has left him with no light perception (NLP) OD.

PMH: childhood chicken pox

Family History: Degenerative Arthritis, allergies, and non-insulin-dependent diabetes Mellitus

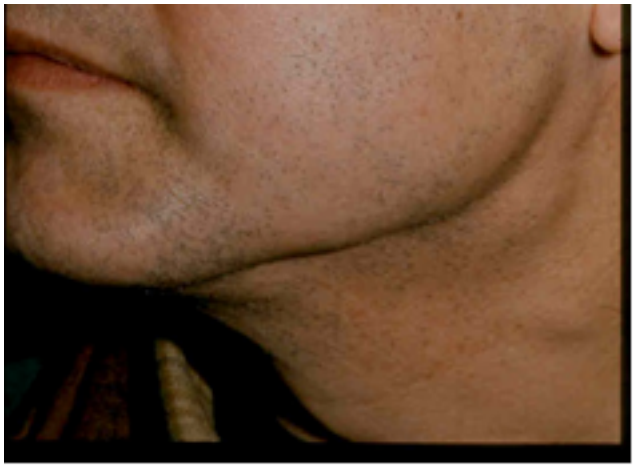
Allergies: NKDA

Meds: PF 1% q ½ hr OS, Tobradex ointment OS, Atropine qd OS

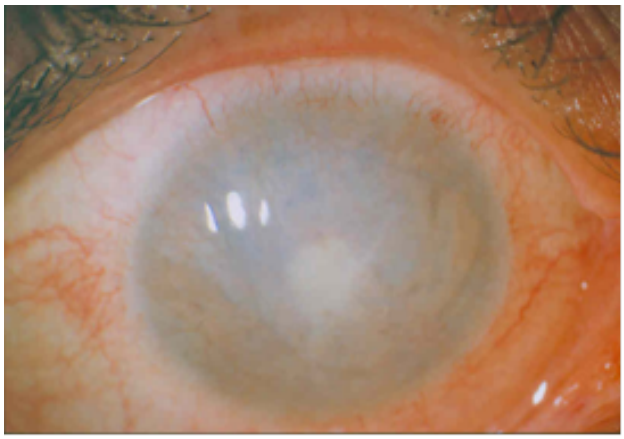
SHx: No tob, no ETOH. He is a former electrical wire-cutter. He was laid off four months ago for lack of performance on his job.

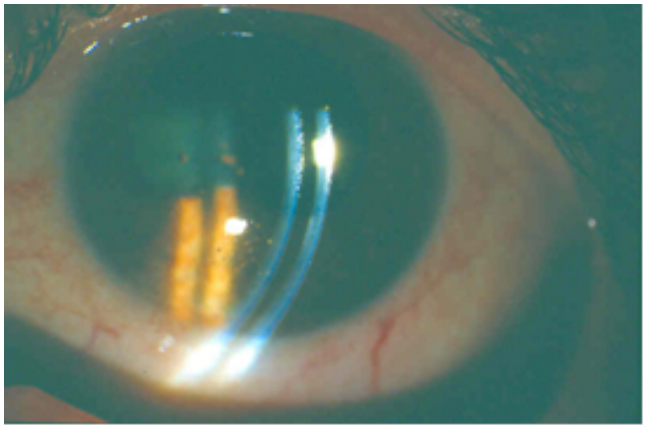
Review of systems (R.O.S.): poor appetite, 25 pound weight loss, and progressive fatigue.

Physical Exam: VA NLP OD, 20/200 OS, IOP 8 and 22 OD and OS respectively.
EOM full, CN II-XII intact, Shotty adenopathy (1.5-2 cm in diameter) Figure 1below



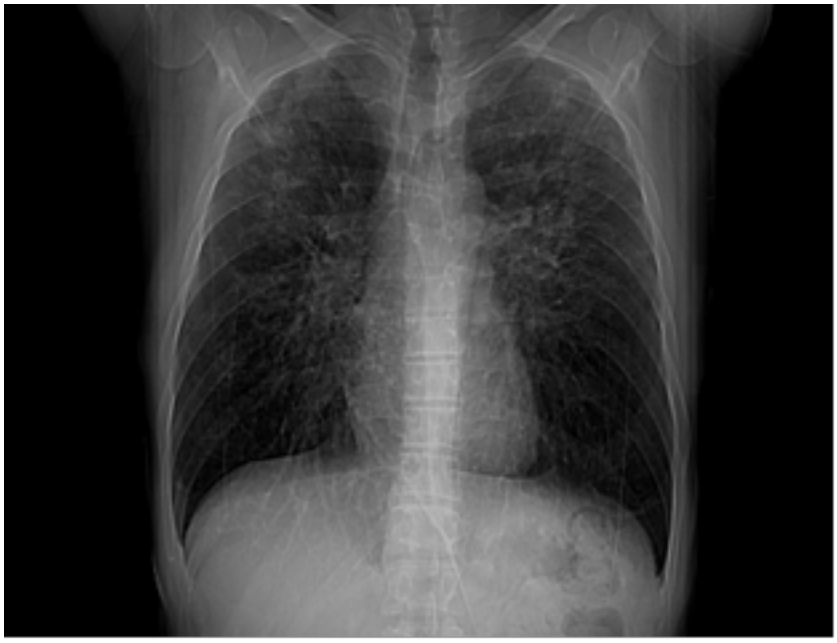
Slit Lamp exam: Anterior Segment: Pre-phthisitic OD, ½+ injected Conjunctiva, Mutton fat keratic precipitates on the cornea, 1+ cells in the anterior chamber, posterior synechiae, and 2+ PSC, 2+ NS cataract. (figure 2)



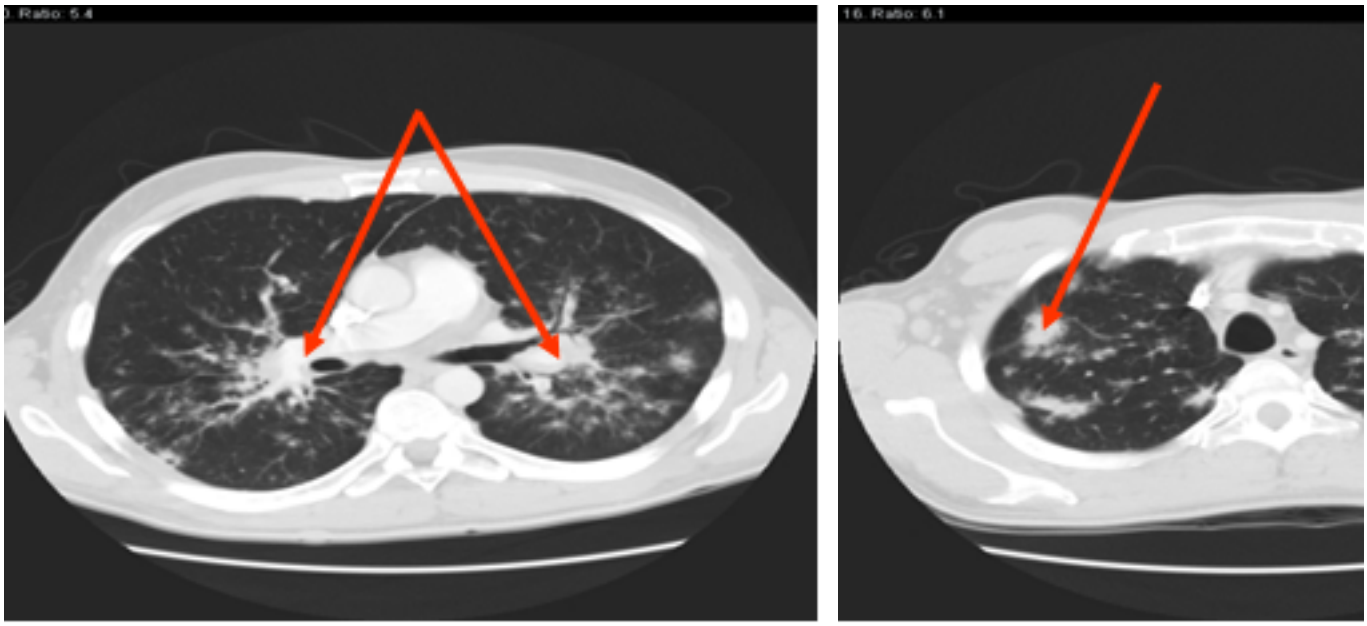


Posterior Segment: No view bilaterally.

Labs and Tests:
B scan:
OD: Funnel Retinal Detachment and Globe Irregularity
OS: Total PVD with an Attached Retina
Labs: FTA Abs, RPR, ANCA, PPD, electrolytes, CBC, LFT, RFT are all wnl. ACE 49 (8-52)
CXR: Multiple patchy airspace opacities and increased interstitial markings in the peri-hilar regions (Figure 3)

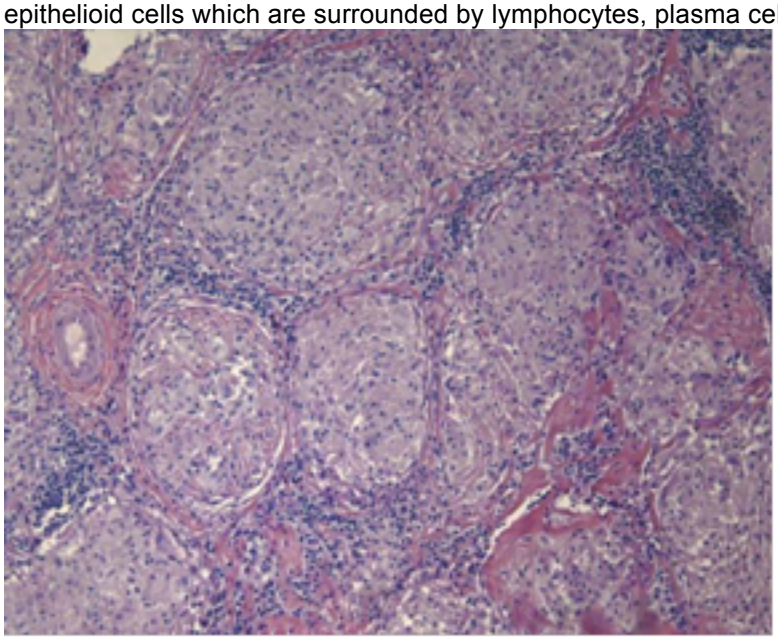


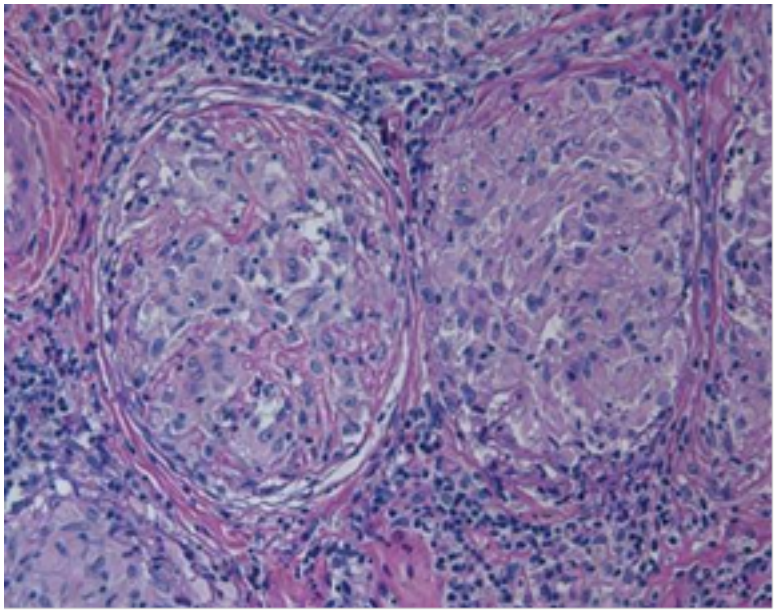
CT of Thorax: Bilateral Hilar Lymphadenopathly (A) and interstitial pulmonary infiltrates (B)



Cervical Node Needle Biopsy: Non-caseating granulomas consisting of histiocytes, and

epithelioid cells which are surrounded by lymphocytes, plasma cells, and fibroblasts. (Figure 5)



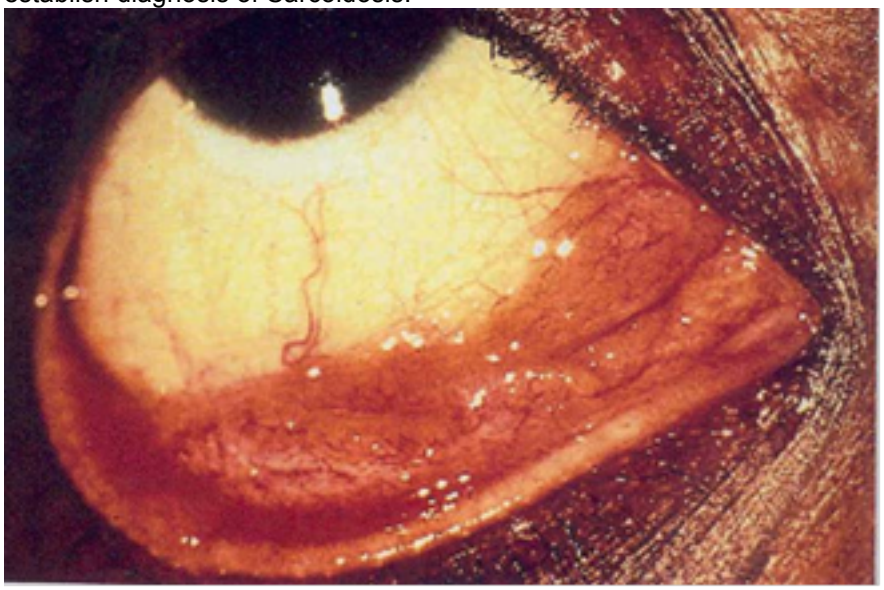


Diagnosis: Ocular Sarcoidosis

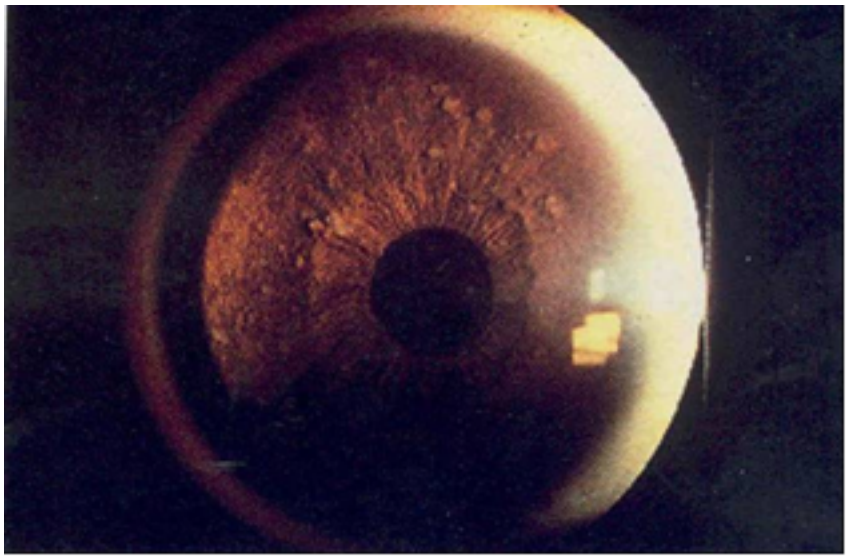
Management:
1) PF 1% q ½ hr OSà q 1hà q2h, Tobradex ointment OS, Atropine qd OS, Alphagan tid OS, Cosopt bid OS
2) Prednisone 40 mg daily with taper over 4 months? 10 mg daily, and prophylactic bactrim
3) Methotrexate 10 mg weekly with 20 mg daily Prednisone (with taper).

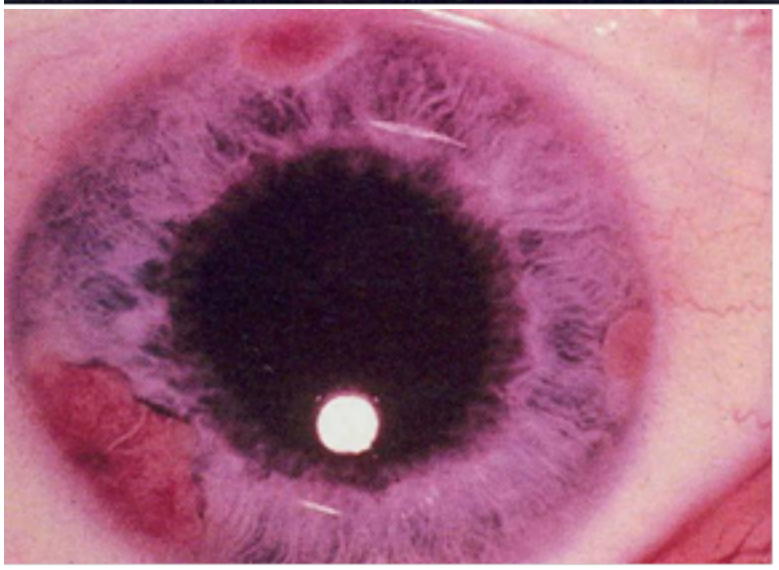
Outcome: VA: NLP OD, 20/20 OS
IOP: 14 OD, 17 OS
Ocular Sarcoid: 26-50% of all cases of sacroidosis. Sarcoidosis can involve any part of the eye and/or the orbit

Anterior Chamber: (~85% of ocular sarcoid presentations)
Conjunctiva: involved in 7-70% of ocular sarcoid. Most commonly presents as “millet-seed nodules” (Figure 6). May lead to keratoconjunctivitis Sicca. The nodules can be biopsied to establish diagnosis of Sarcoidosis.

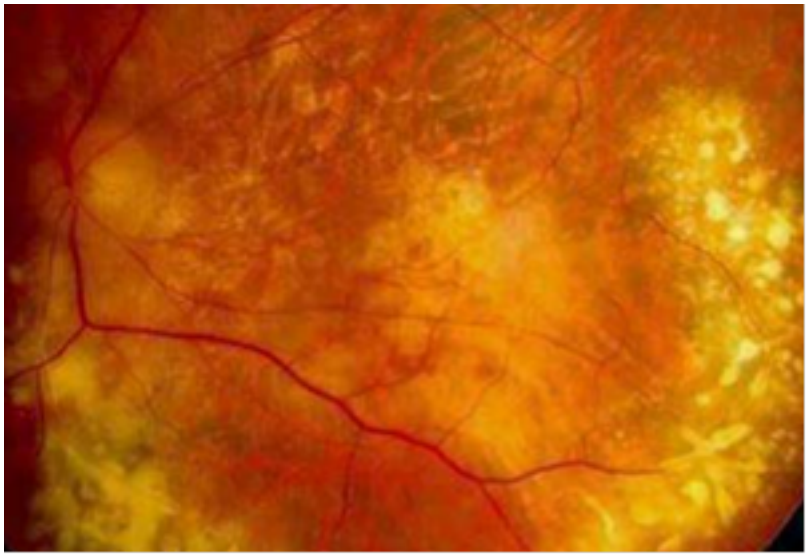


Anterior Uveitis: 20-70% of ocular manifestations of the disease. Most commonly chronic and granulomatous. Complications from the uveitis include cataract (4-35%), glaucoma (4-33%), posterior synechiae (20-26%), corneal band keratopathy (4.5-11%), and Iris Nodules (12.5%). Iris nodules can either be busacca (Figure 7A) or koeppe which are granulomas attached to the iris, or true iris nodules (Figure 7B). New iris nodules signify acute inflammatory episode of ocular Sarcoidosis.

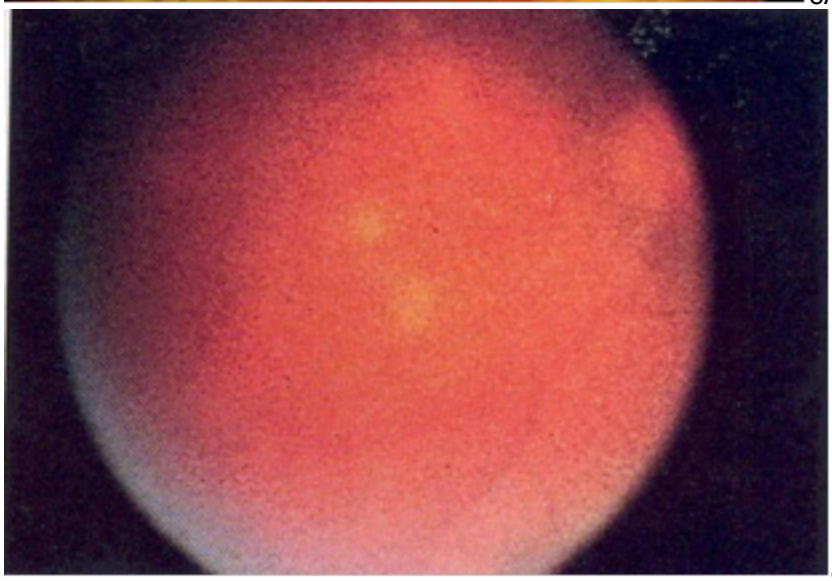




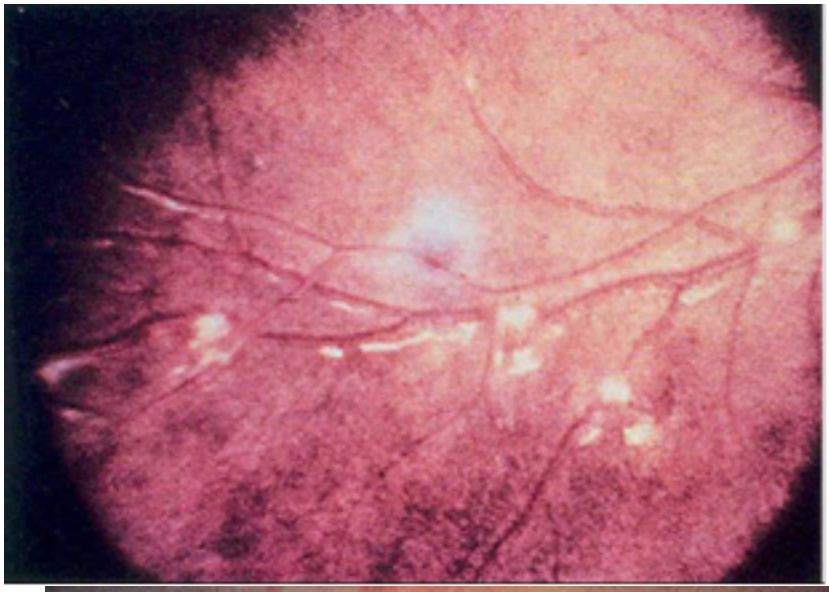
Posterior Uveitis: involved in 25% of ocular cases of Sarcoidosis. Most commonly involved are: periphlebitis: “candle-wax dripping” (Figure 8A); vitritis (3-62%) (Figure 8 B); intermediate Uveitis (16-38%); panuveitis (9-30%); posterior uveitis (12%); exudative RDà pthisis; retinal vasculitis (9-34%) (Figure 8C), CME (19-72); optic nerve (7.4-34%) (Figure 8D)

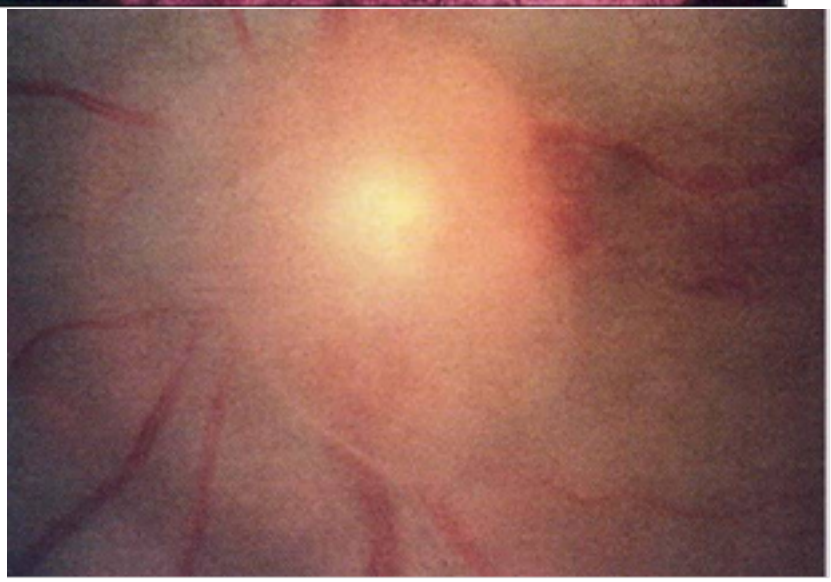


8A



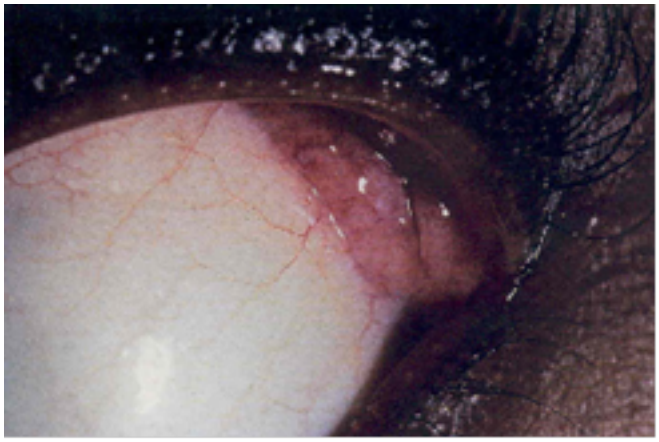
8B





8C 8D

Orbit: Any part of the orbit can be involved. Most commnly affected are the Lacrimal glands (7% to 69%) (Figure 9). Involvement of the lacrimal glands can lead to diplopia. The lacrimal gland can be biopsied to establish diagnosis. Involvement of the extra-ocular muscles can lead to diplopia and/or painful external ophthalmoplegia.



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. (Figure 5 above).

Other helpful tests:
1) Chest X-ray: Lung involvement in Sarcoidosis. Can guide treatment and indicative of prognosis.
2) Angiotensin converting enzyme (ACE) is produced by epithelioid cells and might serve a surrogate marker for granuloma load.
3) Also, granuloma monocytes hydroxulate 25-hydroxy-vitamin D to 1,25-D? 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.

Treatment: depending 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.