GRAND ROUNDS

May 14, 2009

Beatrice Whitaker M.D.

HPI

- 23 BM+ presented to ER with complaint of blurry vision, pain, redness and swelling of left eye x 1 day. Reported history of a LUL chalazion x 3 weeks for which he had received erythromycin ointment to use BID from and hot compresses. He was to follow up for possible excision but decided to try home remedies including warm milk and epsom salt for the last 2 weeks without relief.
- ROS: +headache, pain on left side of face, nasal stuffiness, no fever, chills, night sweats, no recent dental work or tooth pain, sinus infection or trauma
- PMH/PSH: removal of tonsils/adenoids as a child
- POH: wore eyeglasses at age 3, no ocular sx or trauma

HPI continued….

- FH: no glaucoma/blindness
- Soc: denies x 3
- All: NKDA
- Meds: none

Physical Exam

- NVasc: od 20/20 os 20/50
- EOM: full od, see photo os
- CVF: Full ou
- Pupils:3-2 ou no apd ou
- Tpen: 9,10/13,13
- Color Plates: od 14/14 os 14/14

Physical Exam

- SLE:
- LLA: WNL OD, +3 edema/erythema, warmth, tenderness OS
- C/S: w/q od, 2+inj,+3 chemosis with superior heaping of conj os
- K: clear ou
- AC: d/q ou no c/f ou
- I/P: r,r,I ou
- L: clear ou

- DFE:
- V:clear ou
- D: s/p 0.3 od, blurred margins with disc edema 0.3 os
- M: flat ou
- V: wnl od, tortuous vessels near disc os
- P: no rd, heme, cws or dbh ou
- HEENT: +Proptosis OS, +preauricular node Left, sensation intact V1, V2, V3 distribution B/L



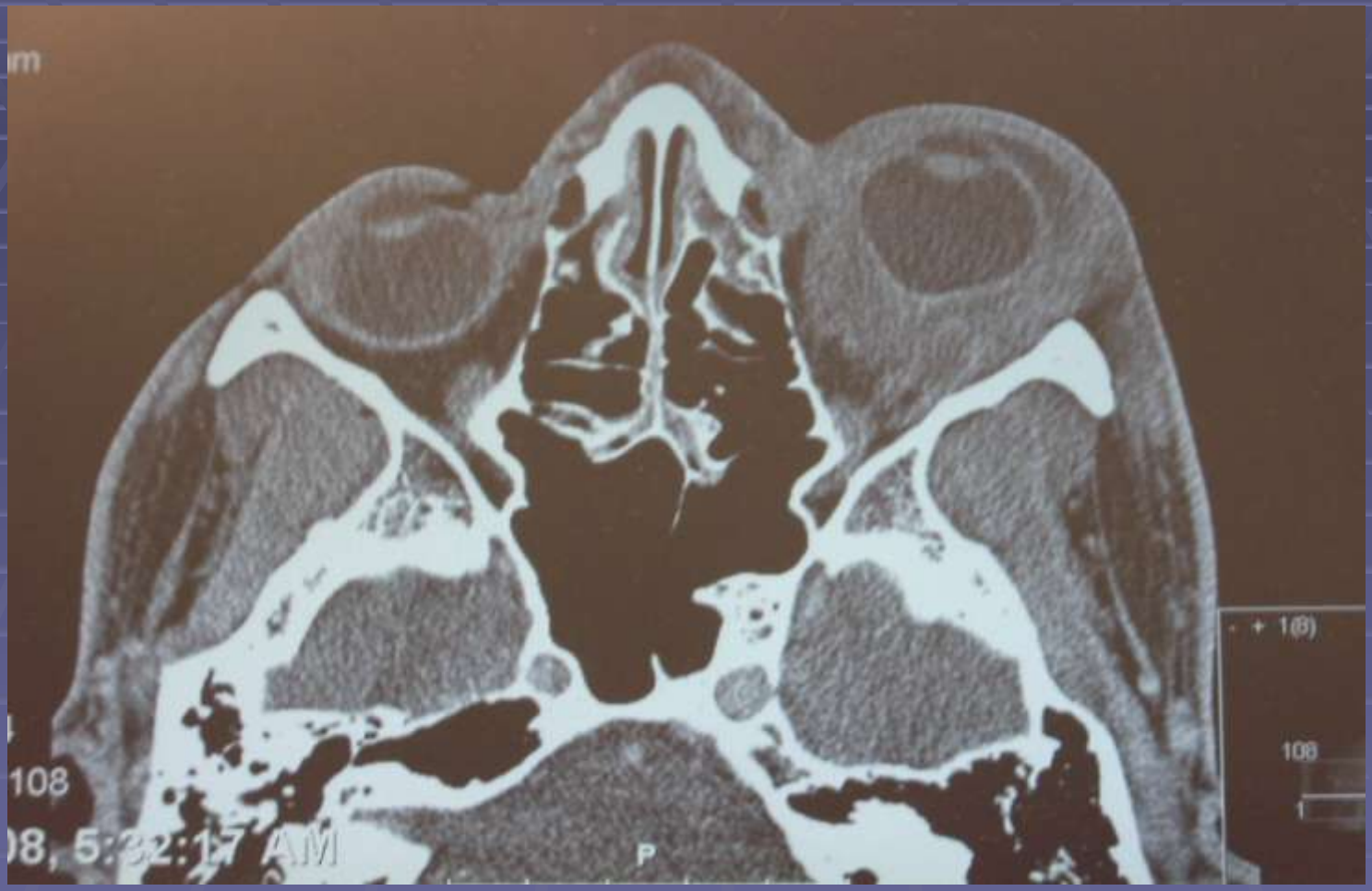

Differential Diagnosis

- Orbital Cellulitis
- Idiopathic Orbital Inflammation
- Sarcoidosis
- Neoplasm
- Thyroid Orbitopathy

Workup

- CT Orbits w and w/o contrast
- Admission for IV Antibiotics
- Blood Cultures
- CBC
- ESR
- CRP
- ANA
- TSH, T3, T4
- ACE
- Frequent daily follow up by ophthalmology

Imaging



CT Reading

- Infilitration of intraconal fat plane encasing extraocular muscles and optic nerve, resulting proptosis. Thickening/enhancement of posterior sclera. No evidence of retrobulbar organized fluid collection. Bony margins are intact.

Assessment/Plan

Orbital Cellulitis Left Eye versus Idiopathic Orbital Inflammatory Pseudotumor

-Continue broad spectrum antibiotics (Zosyn started in ER)

- Addition of IV Vancomycin. Infectious Disease evaluation recommended

-CXR

-Follow-up three times a day

-Reevaluation for consideration of starting steroids….

- Patient was followed three times a day, minimal improvement on IV Antibiotics
- CXR showed no acute disease process
- Labs: WBC: 9.5, ESR 84, CRP 70.2, ACED ANA: neg, ANCA: neg, TFTS: WNL
- Solumedrol 80 Mg IV twice a day was approximately 36 later
- Continued close followup…..

Idiopathic Orbital Inflammation

- Nonspecific Orbital Inflammation
- Usually idiopathic, though less frequently a specific local systemic disease might be identified as cause.
- First described in 1903 by Gleason and Busse/Hochheim.
- Classified specifically in1905 Birsch-Hirschfeld: orbital mass simulating a neoplasm
- Third Most Common Orbital Disease
- Accounts for 4.7-6.3% or orbital disorders

- Can afflict children and adults
- Idiopathic tumor-like inflammation consisting of pleomorphic cellular response and a fibrovascular response.
- Usually confined to the orbit
- Usually self limited course

Subclassification

- Based on anatomical target
- Dacryoadenitis
- Myositis
- Preseptum, Sclera, Episclera, Posterior Tenons, Uvea
- Optic Nerve Sheath (Inflammatory Optic Neuritis)
- Superior Orbital Fissure, Cavernous Sinus (Tolosa-Hunt Syndrome)
- Diffuse involvement of orbital fatty tissues
- Orbital Mass: heterogeneous composition invading extraorbital structures extending along optic nerve sheath from globe to optic canal

Symptoms/Presentation

- Will depend on tissues involved
- Acute Onset
- Deep, rooted, boring pain
- Extraocular muscle restriction
- Proptosis
- Conjunctival injection and chemosis
- Eyelid erythema
- Soft Tissue Swelling
- Impaired Visual Acuity
- Typically Unilateral

- Clinical presentation is often diagnostic and imaging may confirm diagnosis
- Prompt response to systemic steroids helps confirms diagnosis (metastases, ruptured dermoid cysts, infections may also improve with steroids)
- Thorough systemic evaluation should be done if any uncertainty
- Cases with atypical pain, limited inflammatory signs or fibrotic presentation known as sclerosing NSOI may occur
- Biopsy may be required in those cases and for those that fail to respond rapidly or recurrence after treatment is discontinued.

Laboratory and Imaging

- Peripheral Blood Eosinophilia
- Elevated ESR
- ANA levels
- CSF pleocystosis
- CT: diffuse enlargement of lacrimal gland, thickening of extraocular muscles, thickening of tendon insertion, inflammatory infiltrate of retrobulbar fat pad, contrast enhancement of sclera (ring sign)

Histopathology

- Cellular infiltrate consisting of lymphocytes, plasma cells, eosinophils with variable degrees of reactive fibrosis.
- Fibrosis becomes more marked as process becomes more chronic

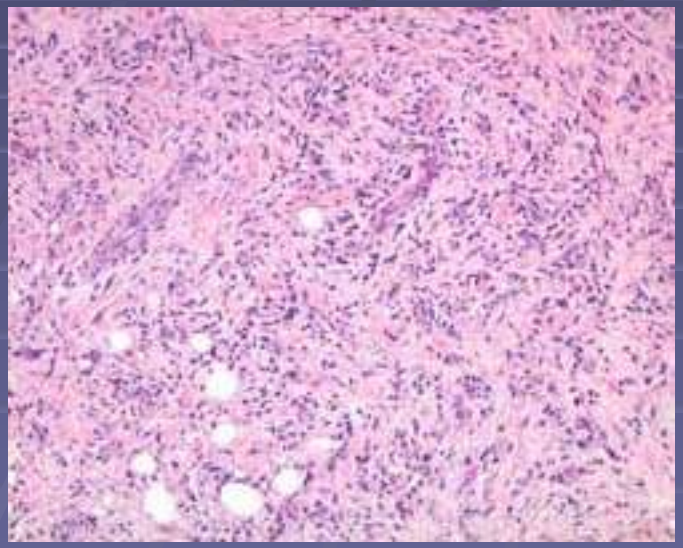


Photo courtesy of Kumar: Robbins and Cotran: Pathologic Basis of Disease, 7th ed.

Treatment

- Systemic corticosteroids
- Adults doses typically 1mg/kg prednisone
- Acute cases generally response rapidly with abrupt resolution of pain
- Steroid use can be tapered as soon as clinical response is complete
- Slow! Below 40mg/da and very slowly below 20mg/day based on response
- Rapid reduction may cause recurrence of signs/symptoms

Treatment continued..

- Topical steroids may help superficial inflammatory reaction and AC reaction
- Some believe pulse dosed IV dexamethasone followed by prednisone results in clinical improvement when oral prednisone fails to control.
- Incomplete response/recurrence suggest need for biopsy after which when diagnosis confirmed orbital radiation, antimetabolites or alkylating agents with continued steroids may be useful
- Rarely orbital decompression is necessary in case with progressive optic neuropathy.

Intraorbital Injection of Triamcinoline Acetonide in Patients with Idiopathic Orbital Inflammation

- Prospective, noncomparitive, interventional case series
- Twenty to 40mg/ml triamcinoline acetonide was injected intraorbitally (intralesional or perilesional)
- Injection repeated at 4 weeks intervals if complete resolution not achieved
- Patients assessed for local and systemic complications, Va, fundoscopy, IOP, Blood pressure, serum glucose measured at each visit.
- Ten patients: 4 orbital masses, 6 lacrimal gland involvement

Continued……

- Substantial improvement (1 patient) and complete resolution (8) was noted over follow up of 9.8 months
- The one patient with no response to first injection refused further treatment
- Complication: Nausea/vomiting in 1 patient isolated episode after 1st injection
- No increase in IOP or systemic complications.
- Conclusion: Injection of corticosteroid is effective any may be considered 1st line treatment in select patients.

Retrospective Review: Distribution, Clinical Features and Treatment Outcome

- Ten year review of idiopathic orbital inflammation at one institution
- Ninety eyes in 65 patients studied
- Diagnoses: isolated dacyroadenitis (21), isolated myositis (19), concurrent dacryoadenitis and myositis (5), orbital apex syndrome (6), inflammation involving preseptum, supraorbital region, sclera, Tenon capsule, orbital fat or nerve (14)
- Mean age 45 years (at presentation)
- Pain and periorbital swelling most common clinical features: 69% and 75% of patient respectively
- Seventeen patients with bilateral involvement
- Biopsy performed in 19 with atypical presentation or failure to respond

Continued

- Patients treated with steroids alone : 45
- Steroids and subsequent radiation: 8
- Steroids and NSAIDS: 6
- NSAIDS only :2
- Radiation with steroids: 1
- Surgical debulking: 1
- Forty-one patients (63%) were treatment successes
- Twenty-four (37%) failures: 1 patient underwent exenteration
- Their conclusion: Systemic steroids with slow taper is established 1st line treatment but refractory cases seen in this study reflect need for a more systematic approach to study of disorder and for therapeutic alternatives to steroids.

Our Patient

- After initiation of systemic steroids patient had subjective improvement with decreased pain, and improvement in Va.
- Patient continued to receive course of antibiotics.
- Next day patient was beginning to have improvement in extraocular movements. Edema of upper lid decreased, chemosis resolved, and injection began to improve.
- Never developed APD or had deficits in color vision by plate testing.

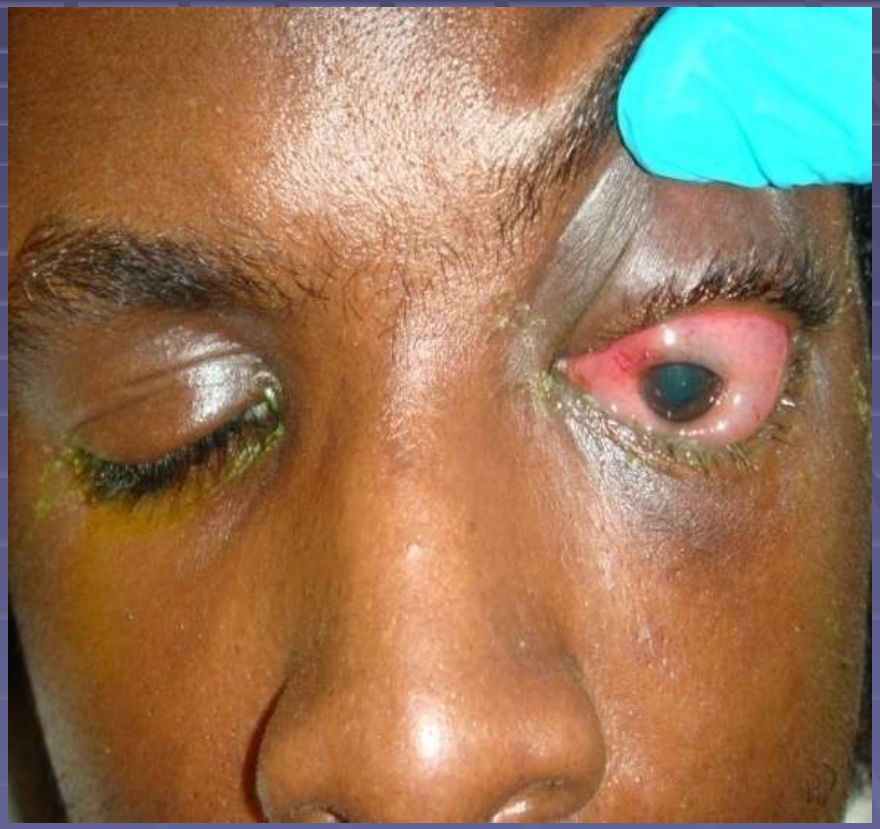
Continued…..

- Patient was discharged home 5 days later on oral antibiotics and 100mg Prednisone
- Patient seen in clinic 2 days after discharge for follow up
- EOMs at that visit: Medial 95%, Lateral 75%, Inf 80%, and 50% Superior
- Undfe: slight haziness of disc os (improved)
- Prednisone slow taper initiated
- Patient was to follow up in 1 week…..

Continued……

- Patient was lost to follow up on 3 occasions
- Would return after running out of prednisone (never completing taper)
- Symptoms recurred in days after stopping prednisone, increased swelling, diplopia, proptosis.
- Recurrent IOI due to medication non-compliance
- Restarted on Prednisone with instructions for slow taper
- Importance of compliance stressed

- Patient began to follow up as scheduled was doing well had slight limitation of upgaze and left gaze with horizontal diplopia.
- Diplopia resolved.
- Patient last seen May 1 2009
- No proptosis. EOMs: Inf: 100%, Add 100%, Sup 90%, Abd 95%
- Undfe: unchanged
- Continued on very slow prednisone taper
- Due to follow up May 22……



References

- Basic and Clinical Science Course: Orbit, Eyelids, and Lacrimal Systems pgs. 54-57
- Yanoff & Duker: *Ophthalmology*, 3rd ed. Chapter 12.12 Orbital Diseases
- Igal Leibovitch ,Venkatesh Prabhakaran, Garry Davis, Dinesh Selva. **Intraorbital Injection of Triamcinolone Acetonide in Patients with Idiopathic Orbital Inflammation**. *Archives of Ophthalmology* 2007; 125 (12) 1647-1651.
- Sonia J Ahn Yuen, Peter Rubin. **Idiopathic Orbital Inflammation Distribution, Clinical Features, and Treatment Outcome**. *Archives of Ophthalmology*. 2003; 121: 491-499

Thanks

- Dr. Purewal
- Dr. Rand
- Dr. Greenberg
- Dr. Frisbee
- Dr. Kartvelishvili
- Dr. Liu
- Dr. Yeager
- J.C.