

Grand Rounds



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Subjective

- CC: Double vision for 2 weeks
- HPI: 66 year old WF referred to the adult strabismus clinic with a 2 week history of vertical diplopia secondary to a partial 3rd nerve palsy of the left eye. Pt feels diplopia has continued to worsen over the past week
- Past Ocular History: Dry eyes (Restasis, AT's)
- PMH: Depression
- Medications: Depakote, Dilantin

Subjective

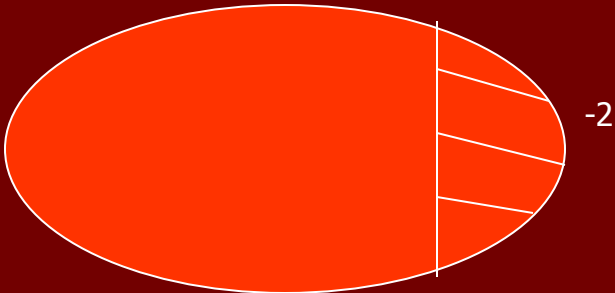
- ROS:
 - 3-4 mo headaches and dizziness.
 - Denies jaw claudication, fatigue, weight loss or additional neurological deficits.
 - No recent trauma or illness.
- Social history: lives with husband, denies smoking or alcohol use

Objective

	OD	OS
<u>Va(Cc):</u>	20/25	20/25
<u>External:</u>	Left Ptosis Inferior orbital fullness OD>OS	
<u>Pupils:</u>	Equally round and reactive- No RAPD	
<u>IOP:</u>	16	18
<u>Ant Seg:</u>	Nasal conjunctival injection OU	
<u>DFE:</u>	WNL OU	

Extra-ocular Movements

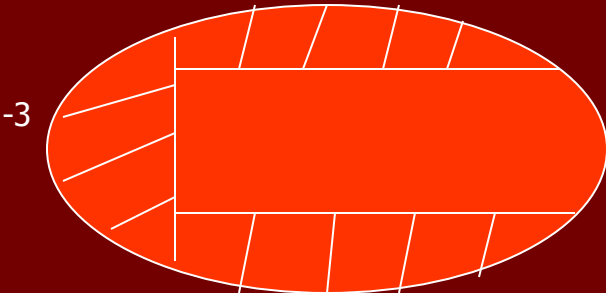
OD



10 \triangle L HT

16 \triangle XT

OS



Clinical Photos



Assessment

- 66yo WF with acute onset partial 3rd nerve palsy OS
- Unexplained adduction deficit OD
- Orbital fullness OD>OS

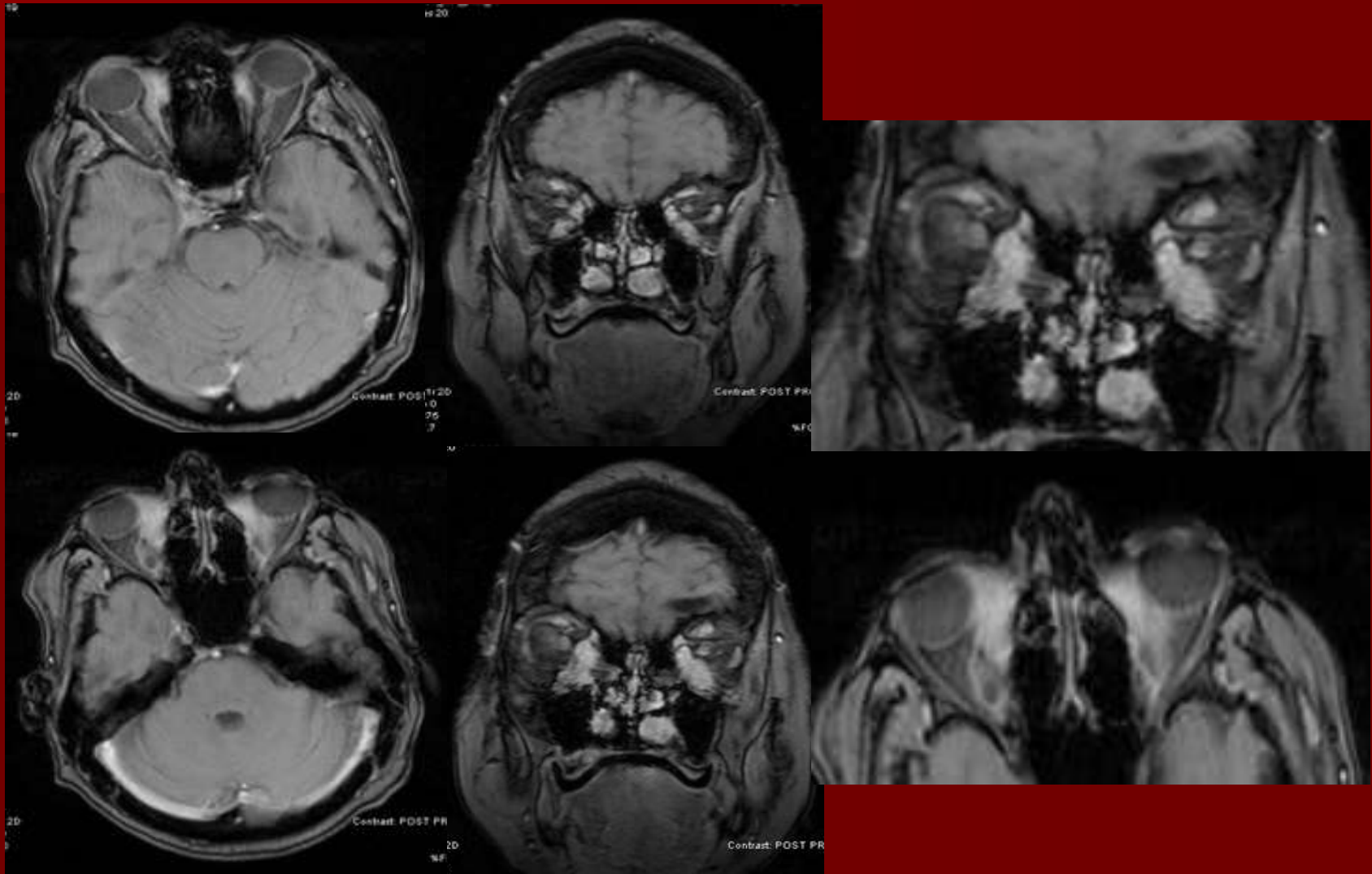
Plan

- Refer to PCP for evaluation
- MRI/MRA head and orbits

Labs/Medical Evaluation

- Glucose: 96 mg/dL
- BUN: 19 mg/dL
- Creatinine: 0.8 mg/dL
- Sodium: 137 mmol/L
- Potassium: 4.5 mmol/L
- Chloride 96 mmol/L
- ESR/CRP: wnl
- Blood Pressure: Within normal range when measured twice daily for one week

Imaging



- **Bilateral anterior medial orbital masses with associated enhancement of the medial and inferior rectus muscle groups bilaterally**

Assessment

- 66yo WF with partial 3rd nerve palsy OS
- Infiltrative process involving the medial and inferior rectus muscles bilaterally

Differential Diagnosis

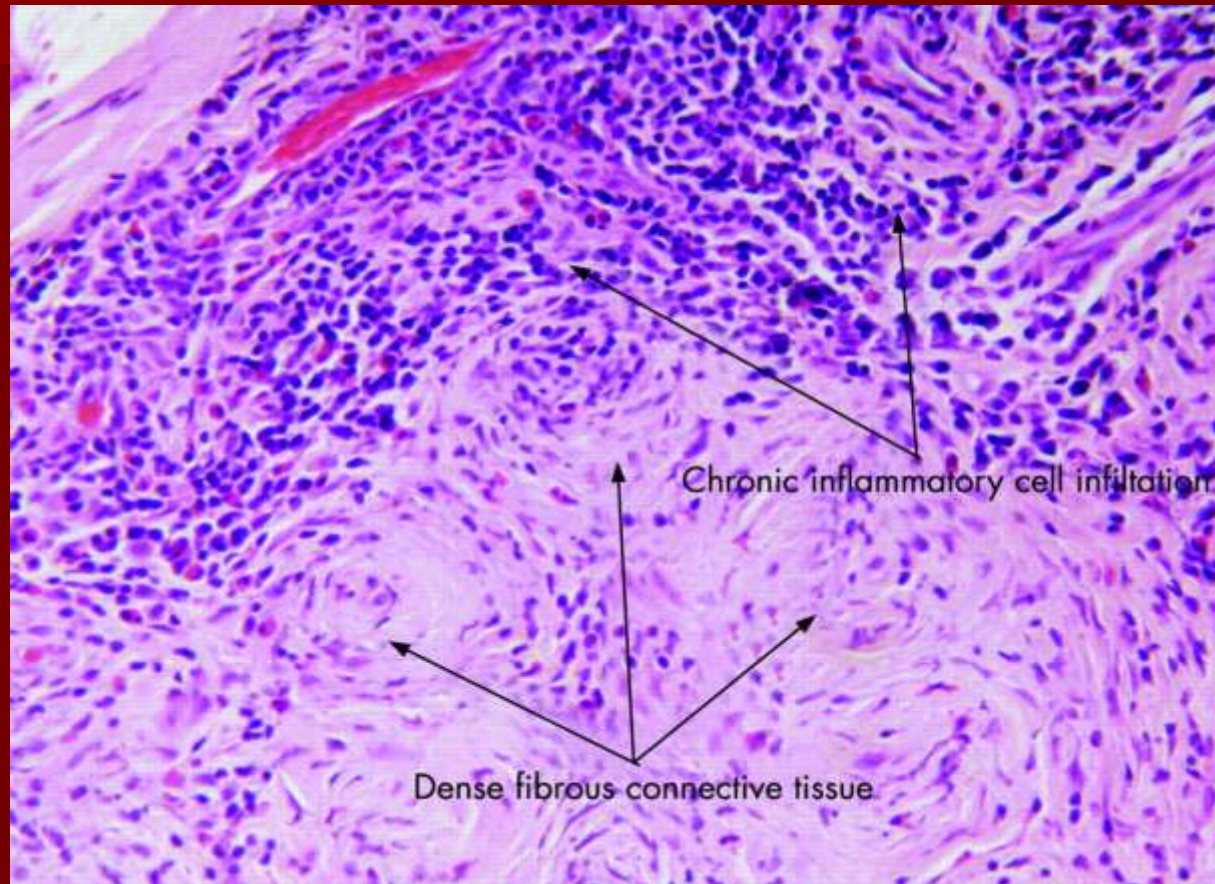
- Metastatic Breast Carcinoma
- Lympho-proliferative disorder
- Idiopathic orbital inflammatory syndrome
- Amyloidosis
- Myeloma
- Histiocytosis
- Infectious

Plan

- Refer to Oculoplastics service for evaluation and biopsy

- Breast exam in the OR did not reveal any masses or suspicious changes
- Biopsy results revealed:
 - Extensive fibrosis with prominent chronic active inflammation
 - Non caseating granulomas
 - Adjacent focal chronic myositis
 - No apparent neoplastic features recognized
 - The Congo red stain is negative for amyloid
 - AFB, GMS and PAS are negative for microorganisms.
 - Immunophenotyping failed to reveal a monoclonal B cell population or an abnormal T-cell immunophenotype

Pathology



Sclerosing orbital inflammatory disease

- Patient started on 80mg oral Prednisone
- Symptoms improving

Discussion: Idiopathic orbital inflammatory syndrome (IOIS)

- Heterogeneous group of disorders characterized by orbital inflammation without any identifiable local or systemic causes.
- It is a rare clinical entity and a diagnosis of exclusion
- Accounts for approximately 5-10% of all orbital “tumors”

Discussion

- Can affect any structure in the orbit
- Presentation can range from abrupt to insidious onset

Discussion

Table 1 Signs and symptoms at presentation

Sign or symptom	Percentage of patients with signs or symptoms
Pain	58.3 (14/24)
Swelling/mass	79.2 (19/24)
Diplopia	37.5 (9/24)
Proptosis	62.5 (15/24)
EOM restriction	54.2 (13/24)
Decreased vision/RAPD	20.8 (5/24)
Ptosis	16.7 (4/24)

EOM, extraocular movement; RAPD, reactive afferent papillary defect.
[Br J Ophthalmol. 2007; 91\(12\): 1667–1670.](#)

Discussion

Table 2 Orbital site involved

Orbital site of pseudotumour	Percentage of patients
Lacrimal gland	54.2 (13/24)
Extraocular muscle	50.0 (12/24)
Orbital fat	75.0 (18/24)
Sclera	4.2 (1/24)
Optic nerve	20.8 (5/24)
Other*	8.3 (2/27)

*One patient had eyelid mass, and one patient had a medial canthal mass.
[Br J Ophthalmol. 2007; 91\(12\): 1667–1670.](#)

Scleritis



Orbital Mass



Myositis



Dacryoadenitis



Discussion

- Diagnosis is often made on the basis of the clinical response to systemic corticosteroids.
- Thyroid eye disease, malignancy and fungal infections may partially respond to corticosteroids
- Histological diagnosis is important

Histological Subtypes

- Classical orbital pseudotumour:
 - inflammatory cells with variable amounts of tissue edema and fibrosis.
- Idiopathic sclerosing orbital inflammation:
 - connective tissue sclerosis and hyalinisation predominated with a paucity of inflammatory cells
- Granulomatous orbital pseudotumour:
 - histiocytic infiltration and multinucleated giant cells
- Vasculitic orbital pseudotumour:
 - vasculitis of the small blood vessels the defining feature.

Discussion

Table 3 Histopathological diagnosis

Histopathology	Percentage of patients
Classical orbital pseudotumour	41.2 (9/24)
Sclerosing orbital pseudotumour*	50.0 (13/24)
Granulomatous orbital pseudotumour	4.1 (1/24)
Vasculitic orbital pseudotumour	4.1 (1/24)
Eosinophilic orbital pseudotumour	0 (0/24)

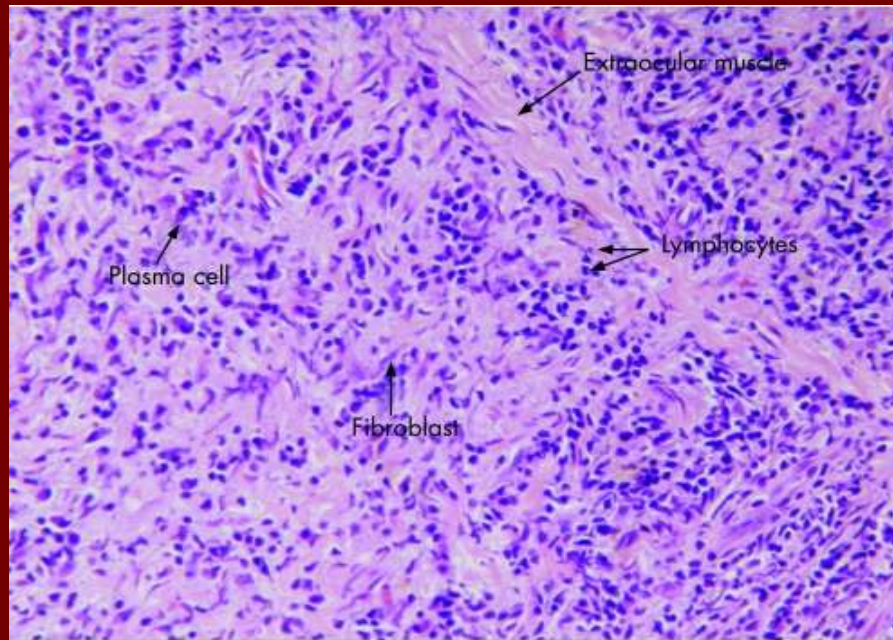
*Three patients developed classical orbital pseudotumour in the first biopsy and subsequently diagnosed as having the sclerosing form in a repeat biopsy.

From:

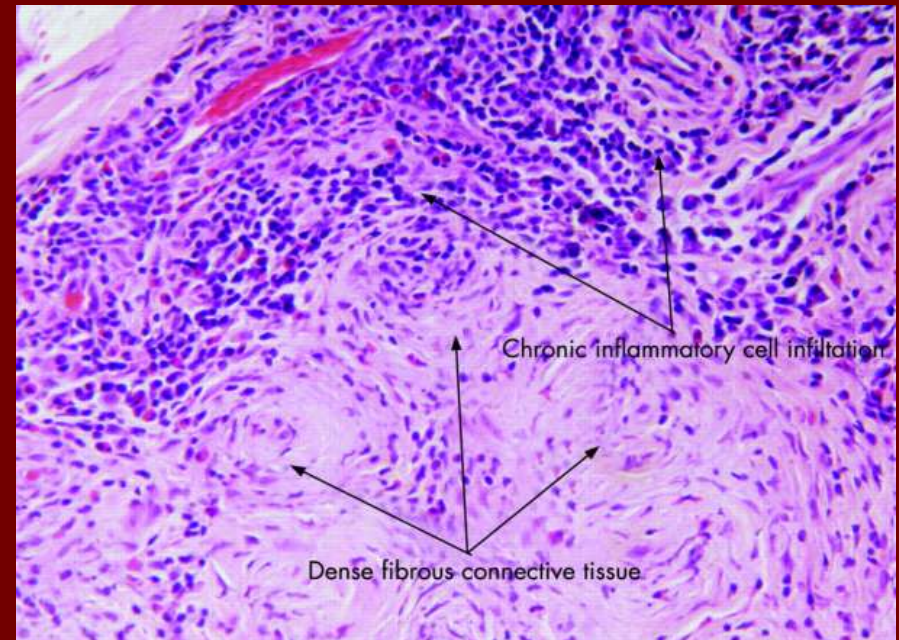
[Br J Ophthalmol. 2007; 91\(12\): 1667–1670.](#)

Histopathology

Classical idiopathic orbital inflammatory disease



Sclerosing orbital inflammatory disease



Treatment

- Steroids are the mainstay of treatment
- High relapse rate
- Radiotherapy
- Steroid sparing/chemotherapeutic agents
 - Methotrexate, azathioprine, cyclophosphamide, cyclosporine

Discussion

Table 4 Treatment received for the management of IOIS

Treatment received	Percentage of patients
Observation alone	20.8 (5/24)
Antibiotics	41.2 (10/24)
Steroid oral	79.2 (19/24)
Steroid intravenous	4.2 (1/24)
Other chemotherapeutic agent*	29.2 (7/24)

*Methotrexate, azathioprine, mycophenolate or cyclosporin.
[Br J Ophthalmol. 2007; 91\(12\): 1667–1670.](#)

Discussion

Table 5 Number of drugs used to maintain remission

No. of drugs	Percentage of patients
0	20.8 (5/24)
1	50.0 (12/24)
2	25.0 (6/24)
3	4.2 (1/24)

Br J Ophthalmol. 2007; 91(12): 1667–1670

Thank You