Idiopathic orbital inflammation masquerade

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Nonmalignant non specific inflammatory lesion involves orbital tissue and may simulates a neoplasm without a known local or systemic cause

Diagnosis of exclusion, based on patient history, clinical picture, response to steroids, and occasionally by biopsy

Etiology unknown immune mediated

Third most common cause of orbital inflammation

Represents around 10% of orbital masses

The peak incidence fourth and fifth decade but it can also occur in children

no sex predilection

Unilateral presentation is more typical but bilateral presentations are not uncommon

There is no universally accepted classification for IOI

Based on the onset

- Acute
- Subacute
- Chronic

Acute

Abrupt onset of pain, proptosis and other inflammatory signs such as swelling and erythema

Chronic

IOI accounts for approximately 8%–10% of all orbital mass lesions *"orbital pseudotumor"*

Depending on the target tissues involved

Focal

Diffuse

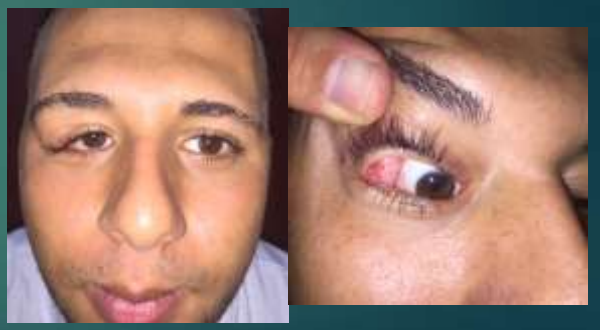
Anterior orbit

Posterior orbit

Focal

Myositis, dacryoadenitis, optic perineuritis, periscleritis and sclerotenonitis

Dacryoadenitis is the most commonly encountered subtype of IOI, accounting for approximately 50% of all IOIs



In 20% of patients, both lacrimal glands are affected, either simultaneously or sequentially

Chronic bilateral lacrimal mass lesion

Diffuse

Anterior orbit

Posterior orbit

Orbital apex syndrome

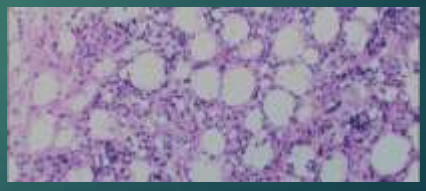
Intracranial extension "cavernous sinus"

Histopathological classification

Classical or Cellular

Granulomatous
Eosinophilic
Vasculitic
Desmoplastic /Fibrous

polymorphous lymphoid infiltrate with varying degrees of fibrosis.



Diagnosis

Clinical	History & Exclusion & steroids response
Radiological	CT MRI
Laboratory	Thyroid,Collagenic,IgG4
Biopsy	Histopathology

Clinical

Five main locations in order of frequency:
Lacrimal gland (darcryoadenitis)
Extraocular muscles (myositis)
Anterior orbit
Orbital apex
Diffuse

Imaging

Computed tomography (CT)

Magnetic resonance imaging (MRI)

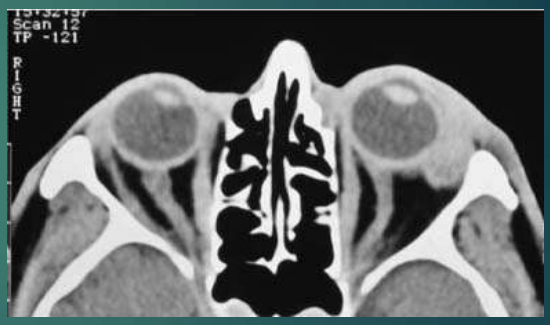
CT

Dacryoadenitis

Diffuse enlargement with shape preservation

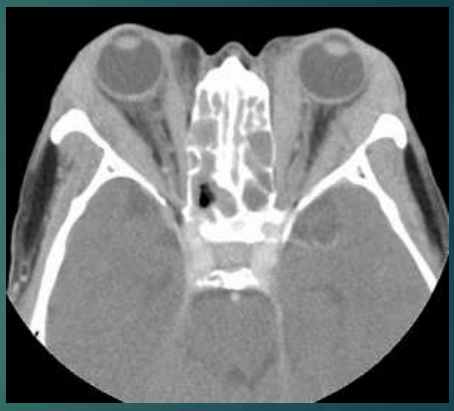
No bone erosion or remodeling

Expansion along the lateral orbital wall



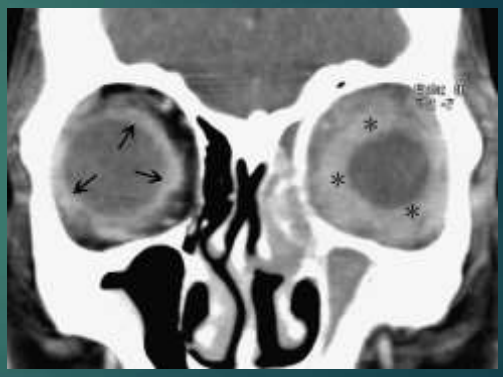
Myositis

Tubular involvement without tendon sparing



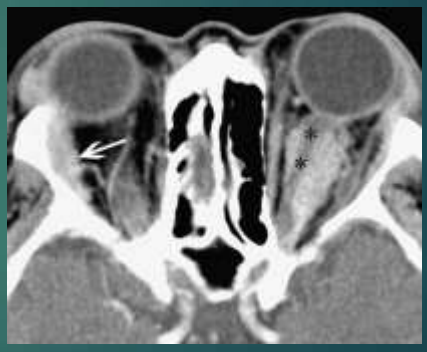
Courtesy of Z.X. Ding

Sclera, episclera, Tenon's capsule, and uvea



Courtesy of Z.X. Ding

CT image of optic nerve involvement with sheath enhancement ("tramline" sign) (asterisks), white arrow showing right lacrimal gland enlargement



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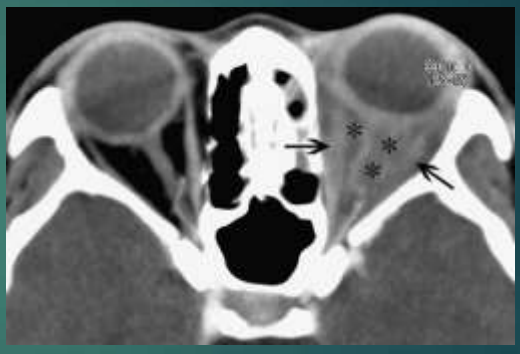
Diffuse type

Orbital fat
Diffuse infiltration and inflammation



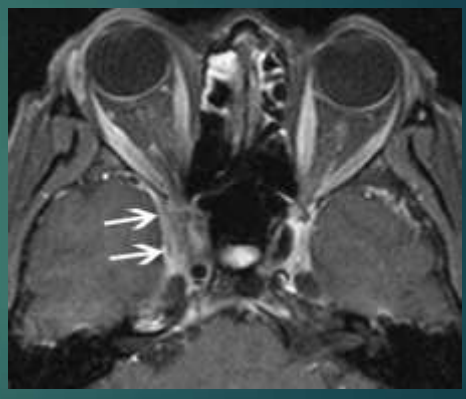
Courtesy of Z.X. Ding

MRI

For any manifestion of orbital apex affection

Extra orbital extension

The cavernous sinus and middle cranial fossa are the two most common locations for intracranial extension



MRI, fat-saturated, T1-weighted image with white arrows showing extension into the cavernous sinus.

Prognostic value

Lesions that appear hyper intense compared with cerebral cortex respond well to corticosteroid therapy

Lesions that are hypointense or isointense compared with extraocular muscle respond poorly.

BIOPSY

Fine needle

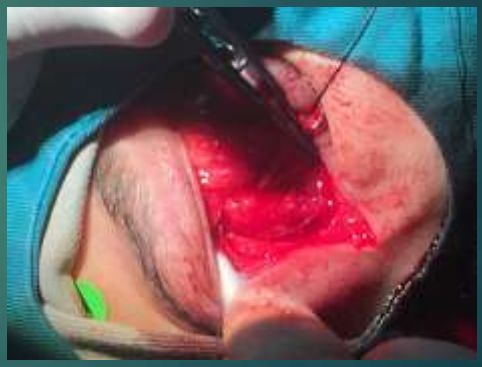
Incisional

Excisional

47 years

Recurrent attacks of dacryoadenitis

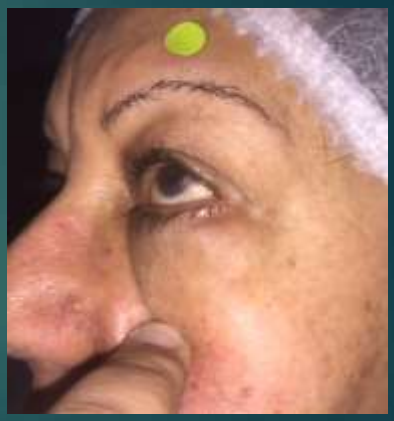
Moderate response to steroids

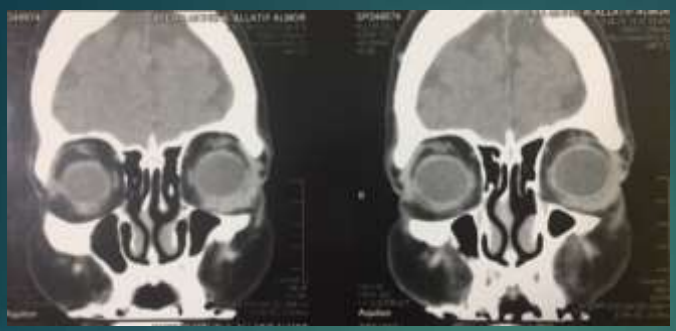


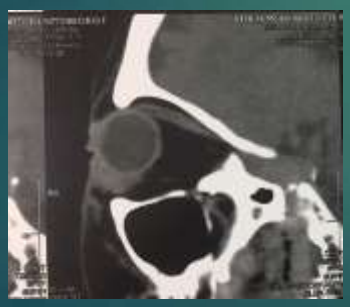
64 years

Chronic anterior orbital mass

Attacks of low grade inflammation



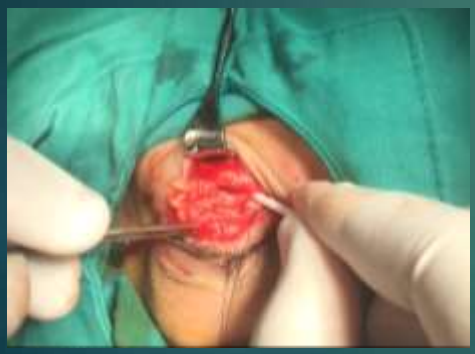




Referred by: Prof. Dr. Ibrahim Allam
Examination: Orbit and Brain
Techniques: Post Contrast Axial and Coronal Cuts

REPORT

- *Ill defined tumefactive soft tissue thickening is seen along the inferior and aspects of the left eye globe, measuring about 2.5x0.9cm, abutting the insertions of the inferior and lateral recti muscles, which show otherwise normal girth…?? Inflammatory pseudo tumor for contrast enhanced MRI assessment*



Differential diagnosis

Dacryoadenitis

Immunoglobulin G4-related ophthalmic disease (IgG4-ROD)

Thyroid eye disease

Lymphoma

IgG4-RD

IgG4-RD was first presented in 2001 in a Japanese study of patients with autoimmune pancreatitis

The orbit was the first extra-pancreatic site of IgG4-RD reported in the literature.

IgG4-related disease (IgG4-RD) is a systemic condition in which fibroinflammatory lesions rich in IgG4+ plasma cells can be present in single or multiple organs of the body. When the disease manifests in the eye or orbit, it is referred to as IgG4-related ophthalmic disease (IgG4-ROD), which most commonly causes painless swelling of the lacrimal gland(s)

Immunoglobulin G4-related ophthalmic disease (IgG4-ROD) most frequently involves lacrimal gland but can also affect the orbital soft tissue, optic nerve, *trigeminal nerve branches, especially infraorbital nerve*, sclera, choroid, and orbital adnexa.

What is the difference??

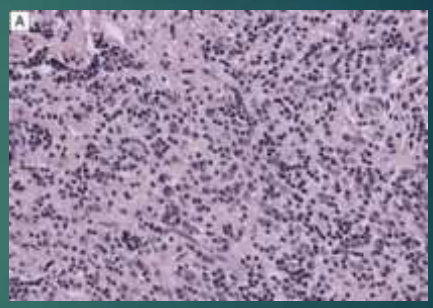
Clinical	less inflammation More bilateral Association with sialoadenitis
CT	Infraorbital nerve enlargement

Histopathology

There are 3 major pathologic features of IgG4-RD:

Dense lymphoplasmacytic infiltrate
Focally storiform fibrosis
Obliterative phlebitis

Blood test shows elevated serum IgG4 (≥135 mg/dl).



IgG4-ROD diagnostic criteria *Goto et al 2015*

1.Imaging studies show enlargement of the lacrimal gland, trigeminal nerve, or extraocular muscle as well as masses, enlargement, or hypertrophic lesions in various ophthalmic tissues.

2. Histopathologic examination shows marked lymphocyte and plasmacyte infiltration, and sometimes fibrosis. A germinal center is frequently observed. IgG4+ plasmacytes are found and satisfy the following criteria: ratio of IgG4+ cells to IgG+ cells of 40% or above, or more than 50 IgG4+ cells/HPF (x400).

3. Blood test shows elevated serum IgG4 (≥135 mg/dl).

Definite IgG4-ROD is defined as 1, 2 and 3. Probably IgG4-ROD is defined as 1 and 2. Possible IgG4-ROD is defined as 1 and 3.

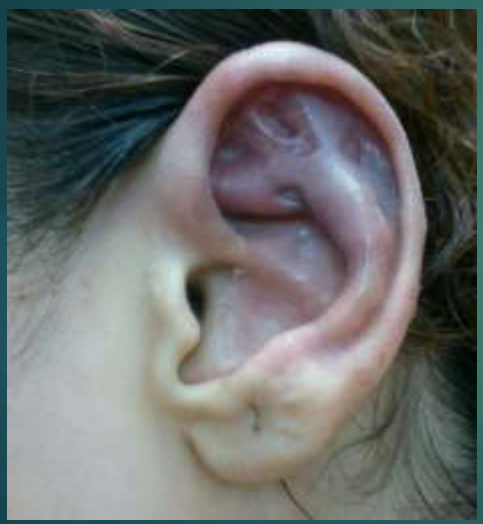
Collagenic disorders

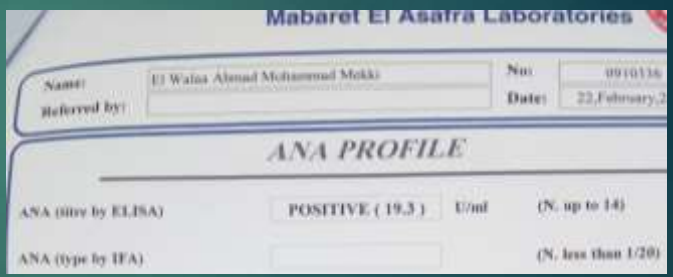
Crohn's disease
Systemic lupus erythematous
Rheumatoid arthritis
Ankylosing spondylitis

42 years

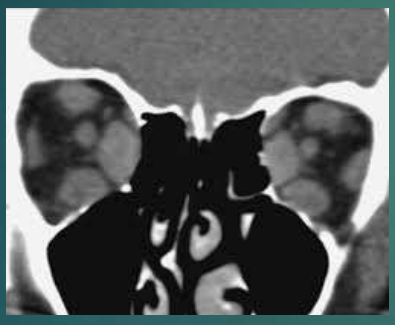
Six months history of bilateral dacryoadenitis

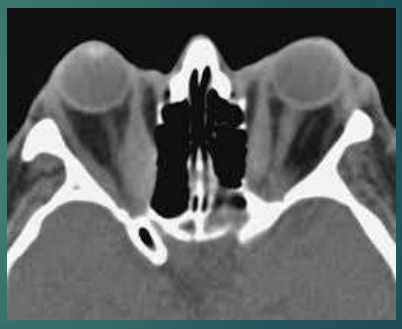
Moderate steroid responder

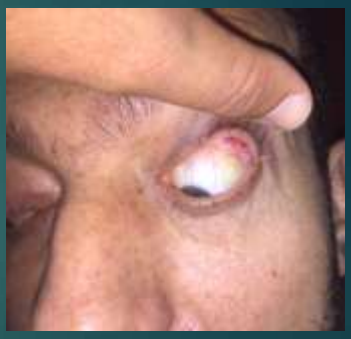




Thyroid eye disease







Lymphoma

Age

Progression

Steroids response

Biopsy

Treatment

Steroids *systemic* *local*

Immunosuppressants

Cytokine/protein specific biologic agents

Radiotherapy

Oral steroids

The mainstay of therapy
diagnostic sensitivity of 78 %

Recurrence rate of 50-60 %.

High dose oral steroid for 2-3 weeks followed by slow tapering over 3 months

Intraorbital injection of triamcinolone acetonide 20-40 mg

Immunosuppressants

Cyclophosphamide
200mg/day is used to treat patients with recurrence on steroid therapy

Cyclosporine 2-5mg/kg

Methotrexate
7.5-12.5mg/kg

Cytokine/protein specific biologic agents

Adalimumab

Infliximab

Rituximab

Tocilizumab

Radiotherapy

Radiation – Radiotherapy is used to treat patients intolerant or resistant to steroids.

1500 – 2500 cGy over 10-15 days is appropriate in steroid resistant cases

Average time taken for response to radiotherapy is 3-8 months.

Thank you