IgG4-Related Disease in Head & Neck

- Recently recognized idiopathic systemic fibroinflammatory condition "Immunoglobulin G4"
- Tumor-like lesion with dense lymphoplasmacytic infiltrate rich in IgG4-positive cells
- Adult patients > 50 years of age
- Male predominance
- Indolent disease course; patients feel relatively well despite multiorgan involvement
- Etiology: Unknown
- Excellent response to glucocorticoid therapy

Granulomatosis with polyangiitis

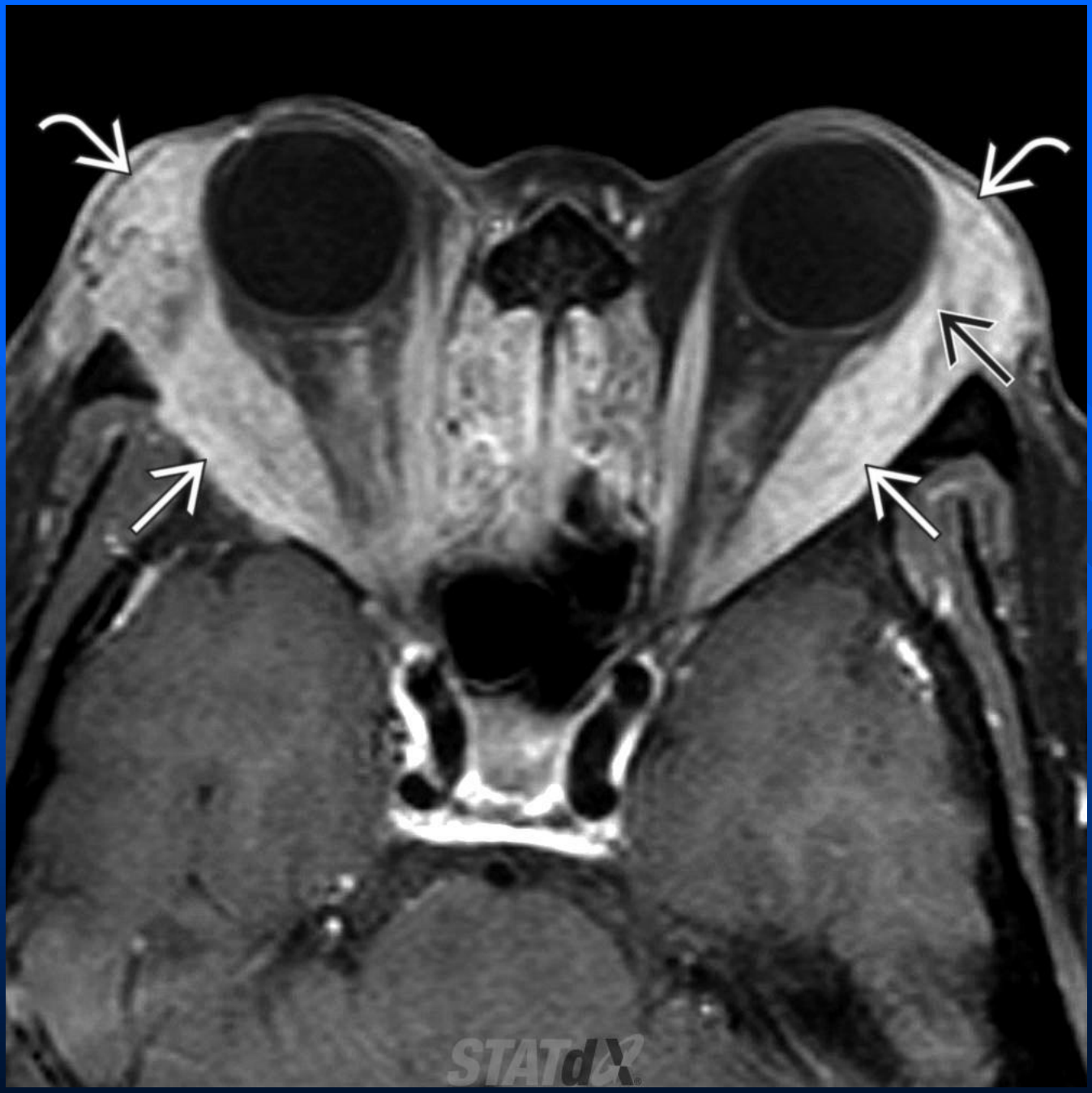
- Previously recognized head and neck conditions now acknowledged to fall within spectrum of IgG4-RD
  - Mikulicz syndrome
  - Kuttner tumor
  - Riedel thyroiditis
  - Inflammatory pseudotumor
  - Eosinophilic angiocentric fibrosis

Presentation

- **Most common signs/symptoms**
  - Head and neck region is commonly involved
    - » Ophthalmic disease > salivary glands > thyroid gland, lymph nodes, sinonasal cavities, laryngeal lesion
  - Typically presents as tumor-like lesion with indolent disease course (develops over months/years)
    - » Bilateral > unilateral proptosis ± visual disturbance due to optic nerve compression
    - » Bilateral firm submandibular > parotid swelling
    - » ± hypothyroidism &/or goiter
  - Constitutional symptoms subtle or absent
- **Other signs/symptoms**
  - Involvement outside head and neck
    - » Neurologic: Pituitary hypophysitis, hypertrophic pachymeningitis
    - » Chest: Lung involvement, mediastinitis, pleuritis
    - » Abdomen: Type 1 AIP, sclerosing cholangitis, cholecystitis, renal disease, retroperitoneal fibrosis
    - » Vascular: Aortitis/periaortitis, etc.
  - Longstanding allergic or atopic manifestations are common

Imaging

- Head and neck region commonly involved, especially orbits and salivary glands
- Bilateral chronic dacryoadenitis ± sialadenitis
  - Bilateral symmetrical > asymmetrical; affects lacrimal > submandibular > parotid glands
  - Involved glands enlarged; rounded contour with nodular or diffuse hypoechoic pattern and nondisplaced hypervascularity on USG
- Orbital involvement could be extraocular muscle myositis ± pseudotumor ± perineural disease
- Cervical lymph node and thyroid gland involvement less common
- Lesions typically T2 hypointense on MR due to high cellularity and fibrosis
- US more sensitive in detecting subclinical sialadenitis or dacryoadenitis
- MR comprehensively assesses head and neck involvement including retrobulbar and sinonasal spaces; T2W and T1W C+ sequences are helpful



Axial T1WI C+ FS MR shows IgG4-related orbital myositis of lateral recti (white solid arrow) and mildly enlarged lacrimal glands (white curved arrow). Homogeneous enhancement is typical. Orbital myositis involves tendon insertion (black solid arrow), as opposed to thyroid ophthalmopathy.