56-year-old female with left-sided eye "bulge"

Joseph Ryan, MD, PhD Gary Gong, MD, PhD

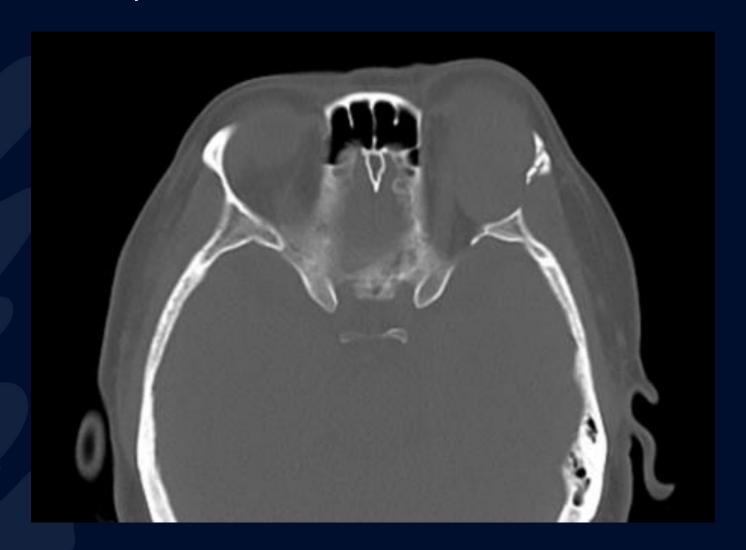












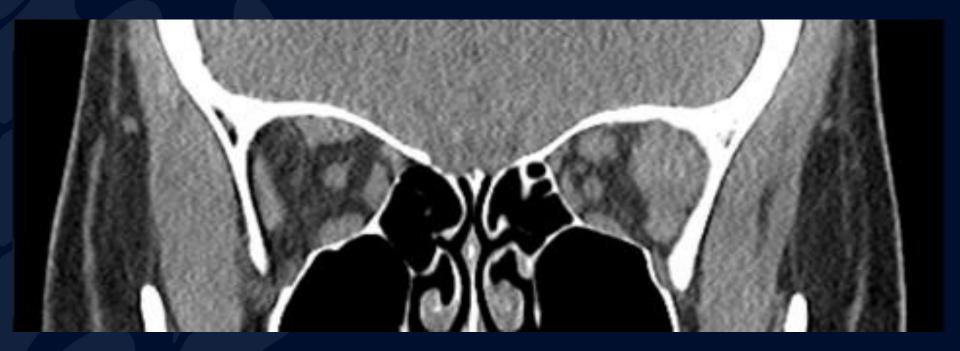




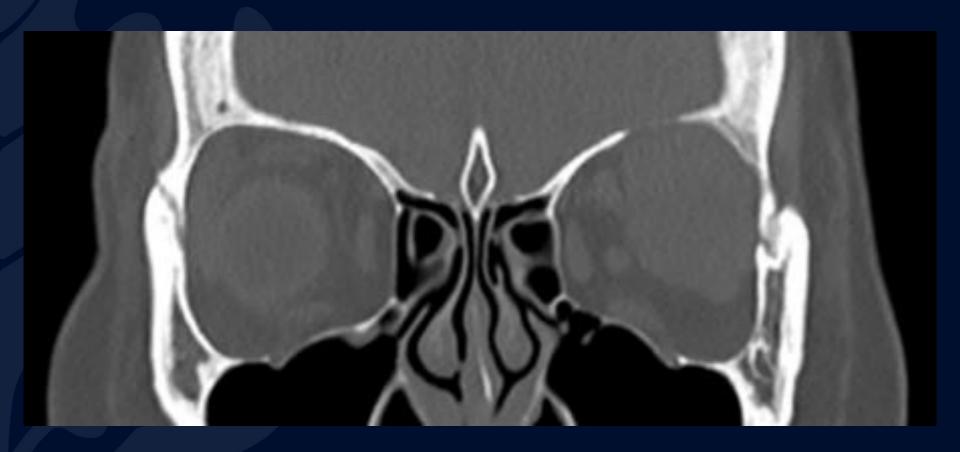


























Orbital Lymphoma



Orbital Masses

- Broad spectrum of benign and malignant entities
- Categorized based on location and histology
- Cross-sectional imaging supplements clinical fundoscopic evaluation; imaging features reflect tissue composition
 - MRI gold standard; provides essential anatomic information about ocular structures involved, perineural spread, intracranial extension



Orbital Masses

- Vasculogenic lesions (17%), lymphoid-derived, lacrimal gland, optic nerve/meningeal, metastatic, peripheral nerve, primary melanoma (1%), and other rare ones: fibrocytic, myogenic, lipogenic, myxoid. (Compiled from Shields, et. al., case study)
- Anatomic compartment → possible tissue(s) of origin
- Evaluation of the bony orbit is crucial when orbital soft tissue mass is present
- Osseous remodeling (slow) vs. erosion (fast/aggressive)
- Bony foramina carry nerves → check for perineural spread, intracranial extension (optic canal, superior orbital fissure, infraorbital canal)



Orbit Anatomy

The orbit is divided into the intraconal space, extraconal space, muscle cone, ocular compartment

- Muscle cone → 5 of the 6 extraocular muscles (not the inferior oblique)
 converge at orbital apex to form tendinous ring (annulus of Zinn); divides
 the intra- and extra-conal spaces
- Ocular compartment/globe → continuous with CNS, comprises 3 layers (not typically visualized unless choroidal or retinal detachment
 - Sclera collagenous tissue layer continuous anteriorly with cornea, posteriorly with dura
 - Uvea vascular pigmented layer, consists of iris, ciliary body, and choroid; choroid very vascular, most frequent location for mets
 - Retina light sensory, continues posteriorly as optic nerve



Lymphoproliferative Lesions / Lymphoma

- Most common primary orbital tumor in adults >60 y/o. Subtypes include the following:
 - Lymphoid hyperplasia
 - Atypical lymphoid hyperplasia
 - Ocular adnexal (malignant) lymphoma
 - most common (67-90%), 24% of all space occupying orbital tumors in patients >60 years old
- Lesions may be 2/2 systemic lymphoma or primary
 - 30% with orbital lymphoma have systemic lymphoma within 10 years
 - MALT-subtype Non-Hodgkin most common



Orbital Lymphoma

- Typically present with painless proptosis, palpable mass, mildly restricted ocular motility.
 - Most (76%) unilateral, however if bilateral lesions in elderly, think lymphoma
 - 50% diffuse ill-defined, 50% smooth circumscribed
- T1-iso to muscle, T2-hyperintense to orbital fat; Uniform enhancement
- Tendency to mold to orbital structures, such as the globe, optic nerve, and orbital wall; tumor can cause bone remodeling, occasionally erosion
- Histology will show a range from benign reactive lymphoid hyperplasia, to highly mitotic atypical lymphocytes (typically B cell origin)



Orbital Lymphoma Mimics

Myositis Pseudotumor

- Idiopathic inflammation of rectus muscle
 - Usually presents with acute pain, periorbital edema
 - Responds rapidly to steroids
- Lymphoma usually pushes rectus, but not directly involved

Other mimics

Granulomatosis with polyangiitis (Wegener's), IgG4-related disease (autoimmune), Erdheim-Chester disease (non-Langerhans histocytosis)

RADIOLOGY

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