56-year-old female with left-sided eye “bulge”

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

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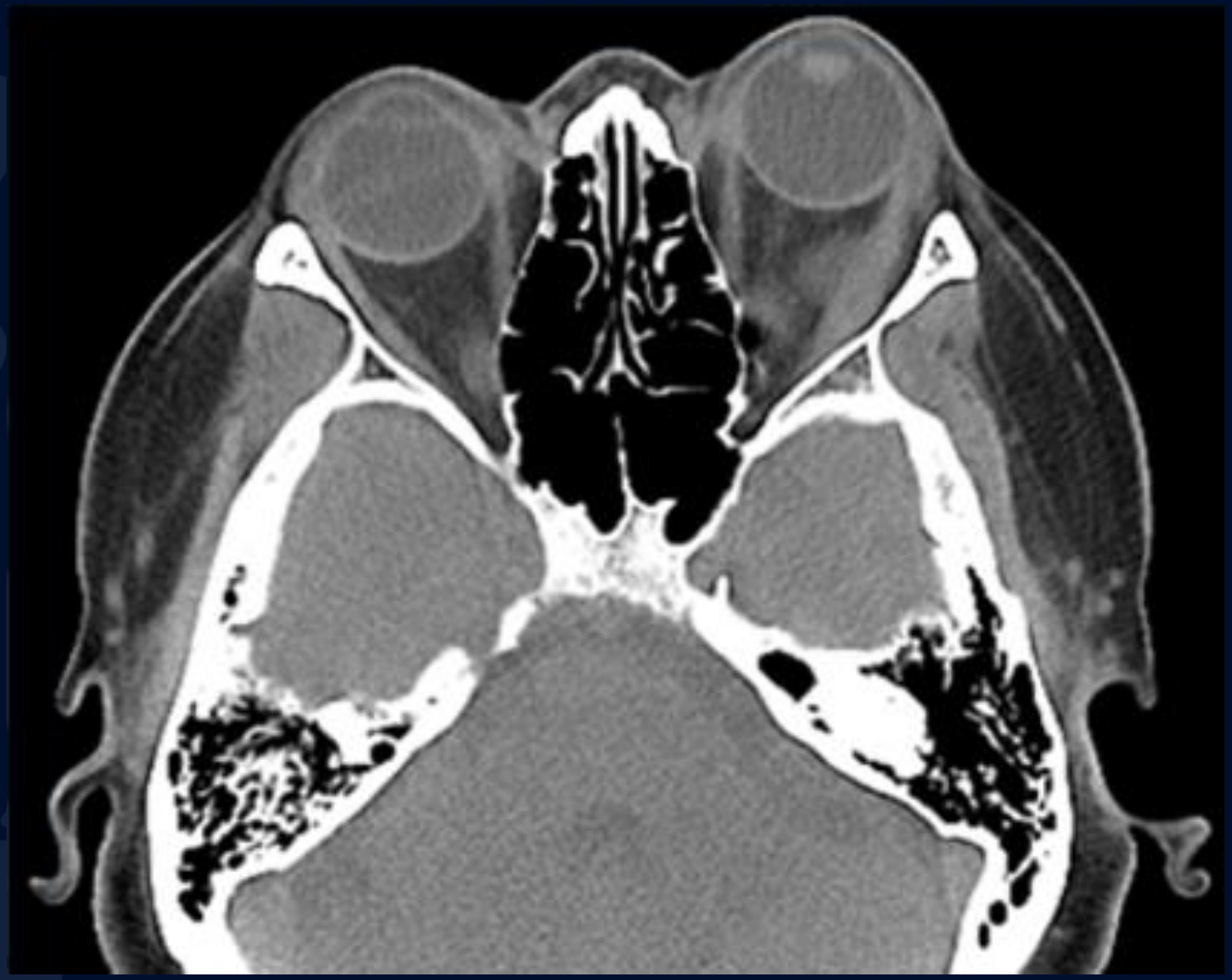
CT paranasal sinuses without IV contrast



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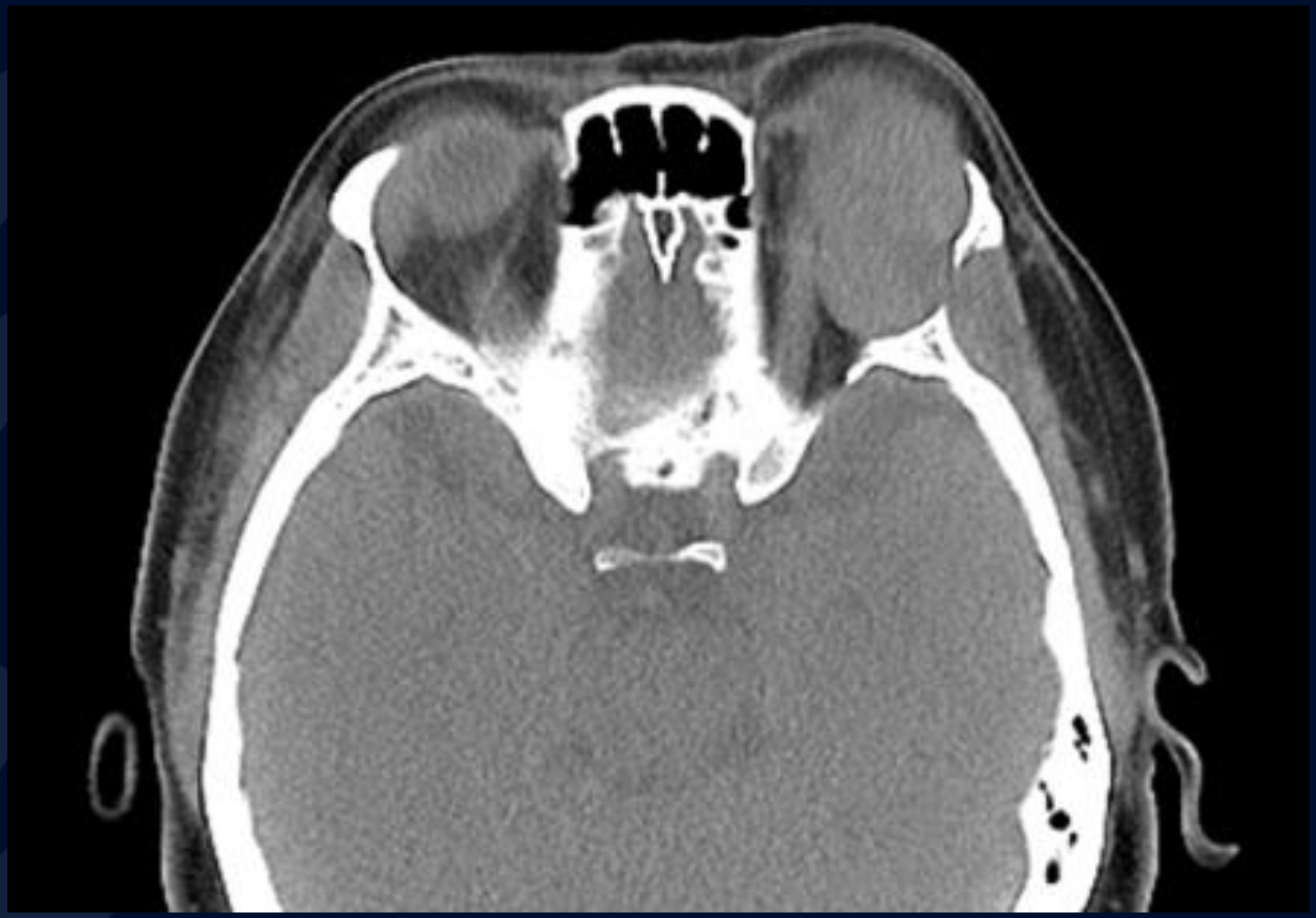

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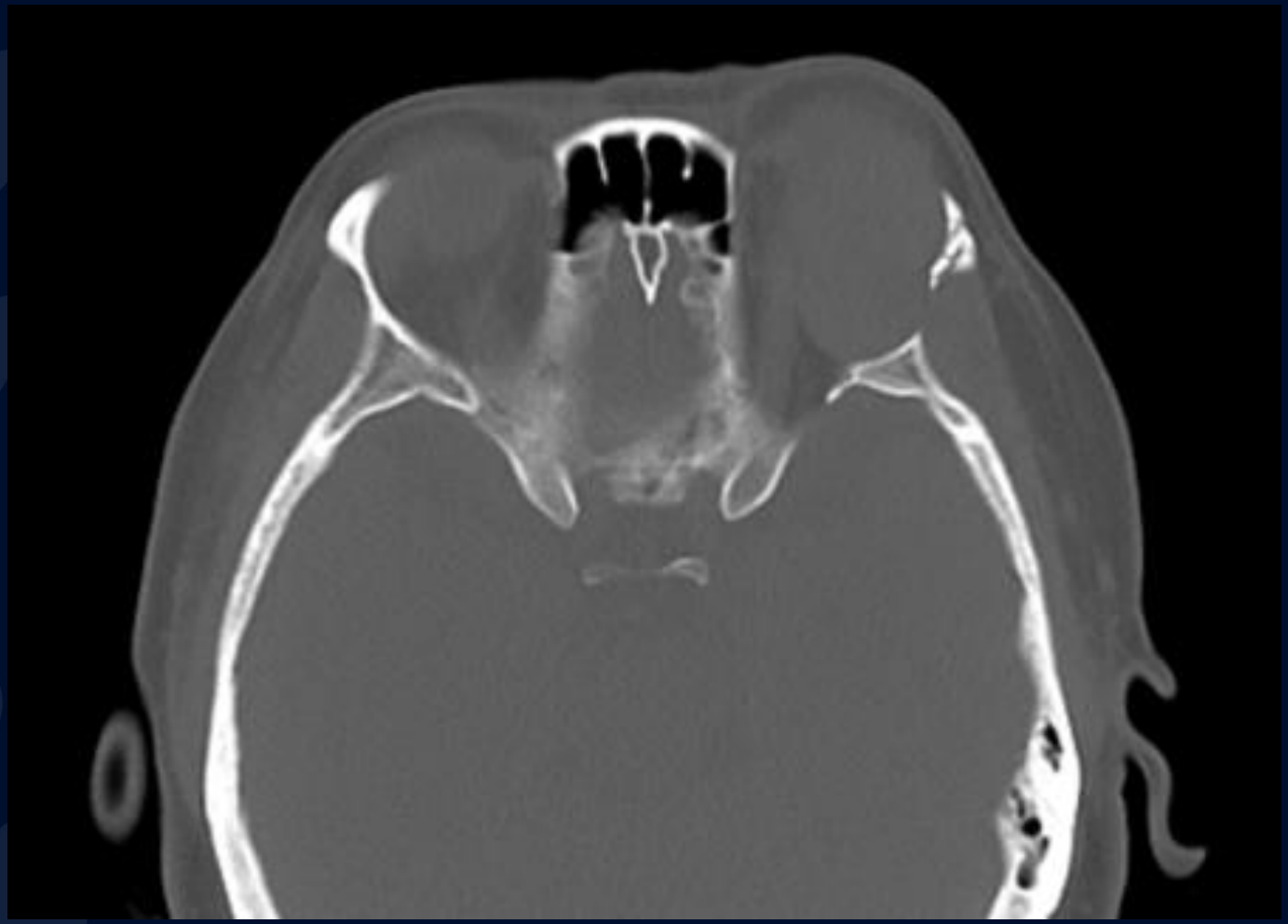

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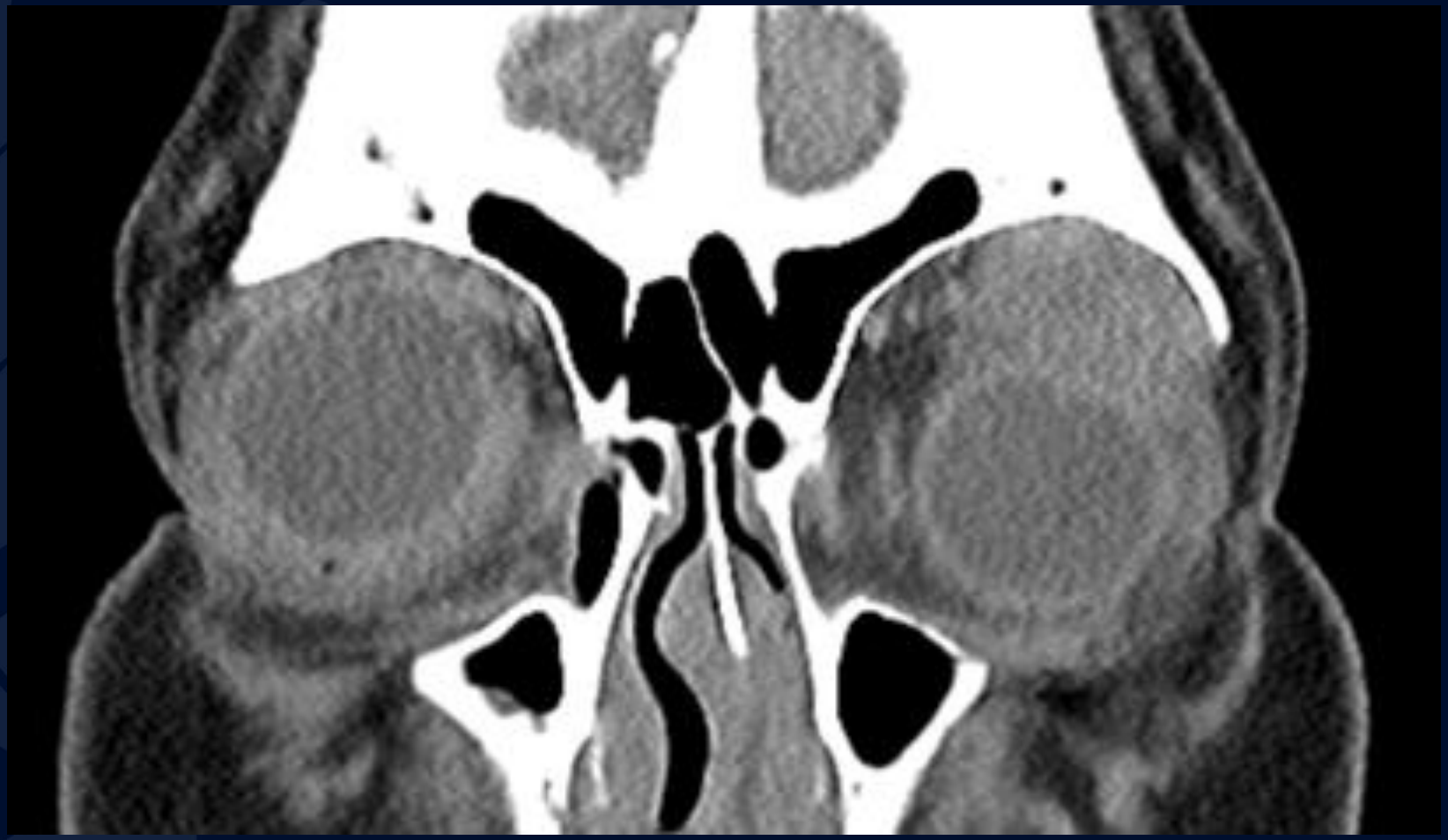

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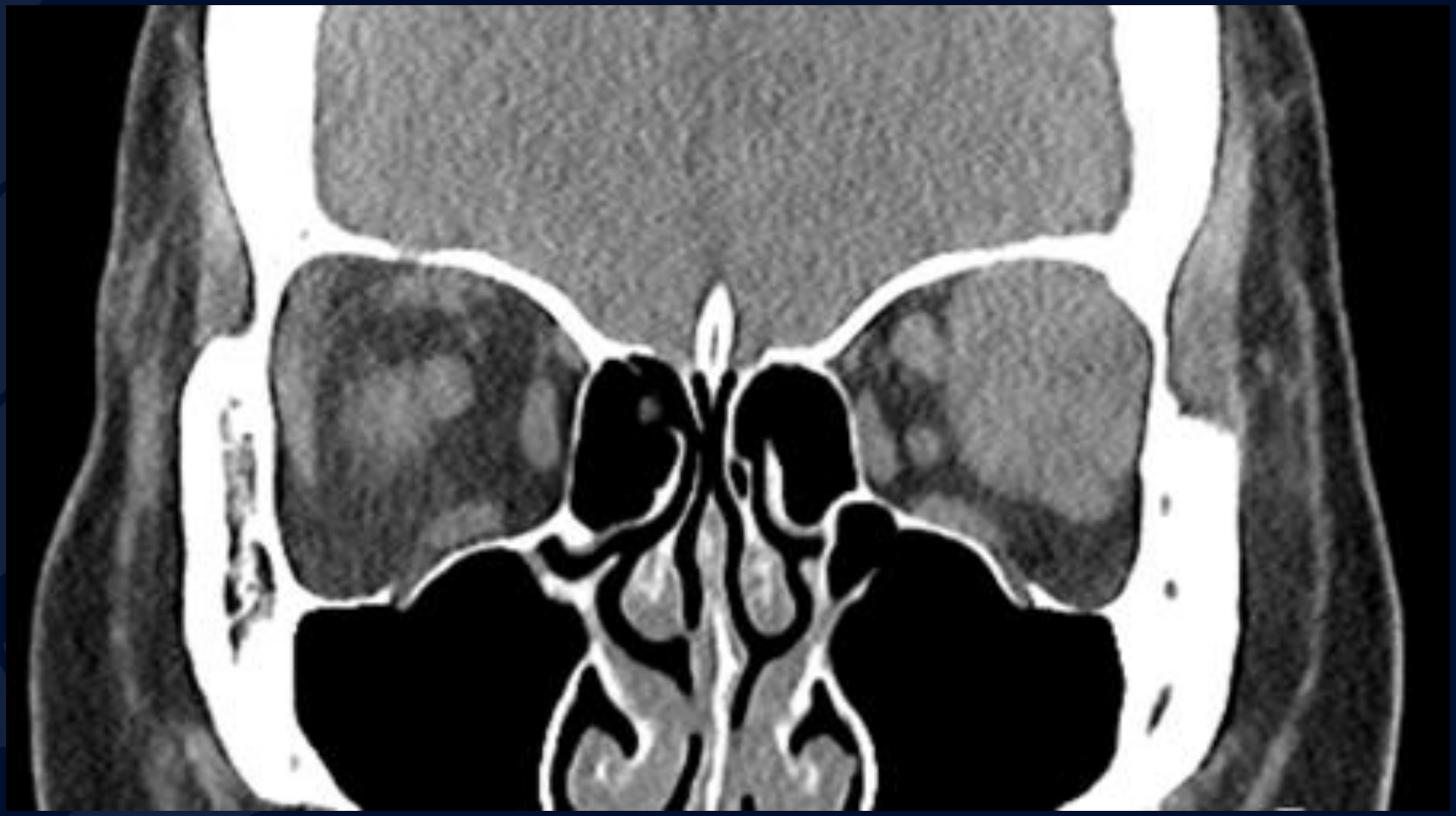

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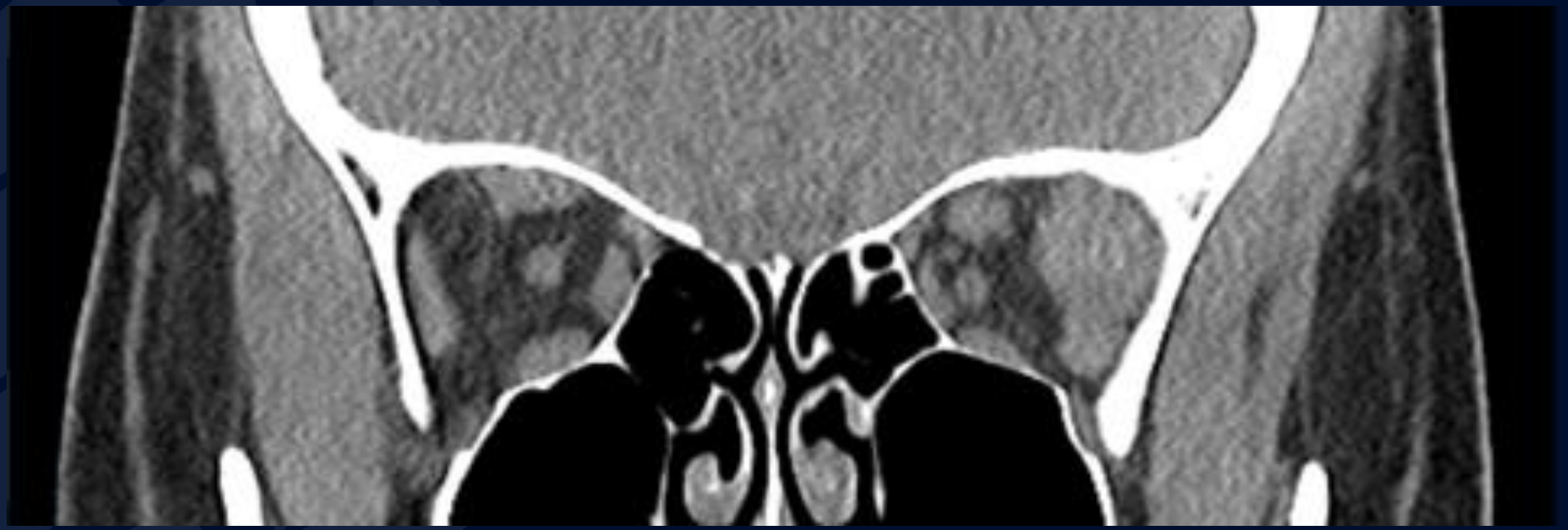

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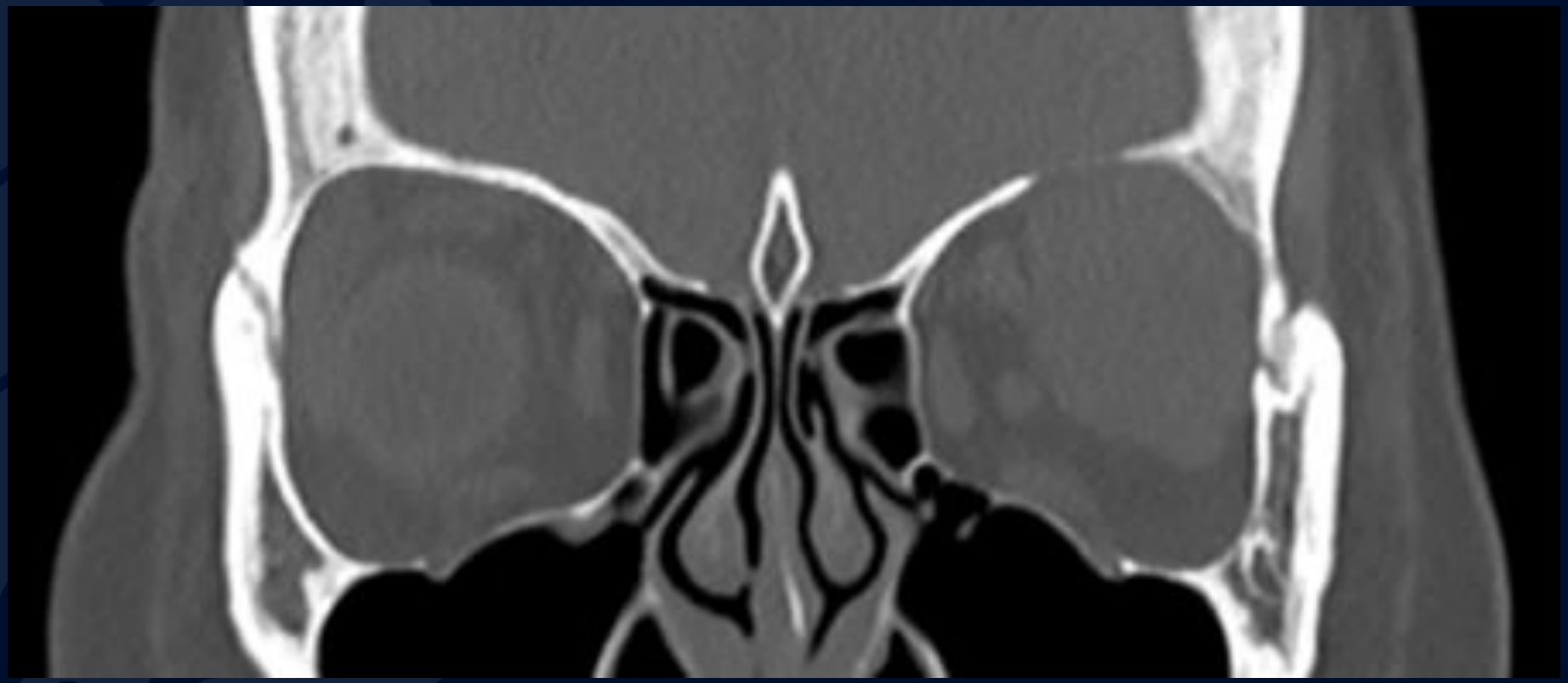

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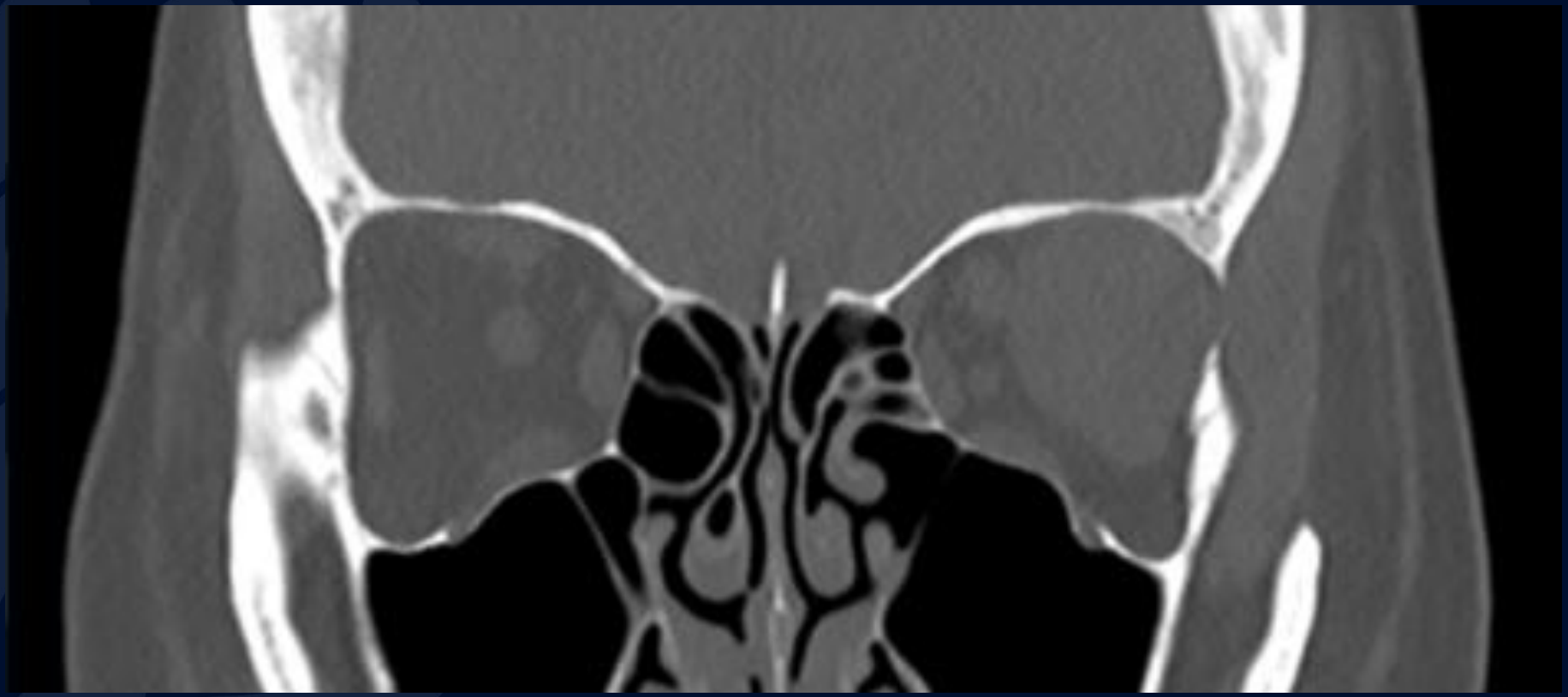

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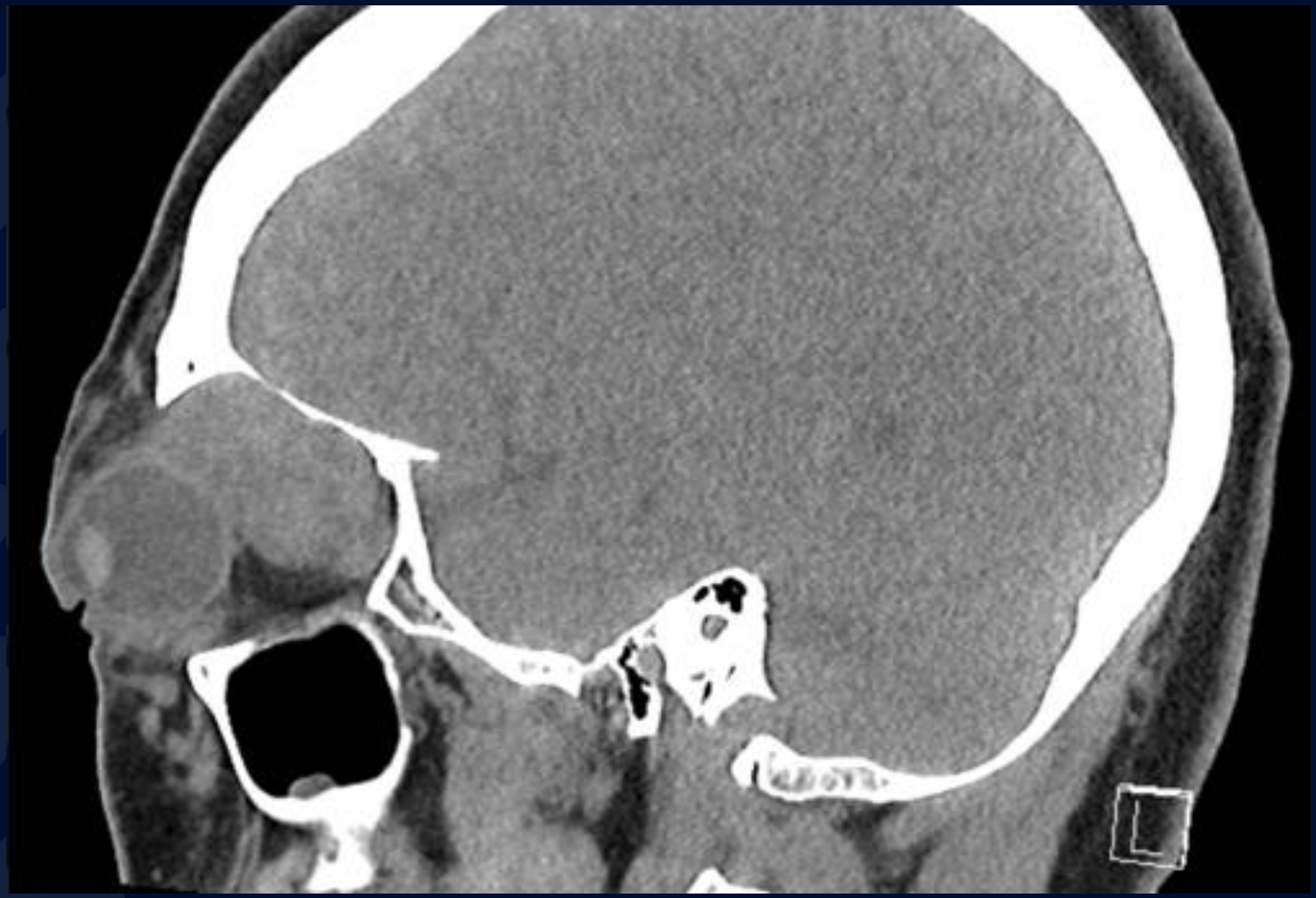

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Orbital Lymphoma

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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

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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)

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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

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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

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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)

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