AMSER Case of the Month September 2024

Exophthalmos and temporal fullness

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Patient Presentation

- HPI: 53-year-old female with no significant PMH presenting with a three-year history of progressive left eye and temple “puffiness,” and retroorbital pressure. She reported no pain or visual changes.
- Vitals: BP 133/75, Temp 98.7
- ROS: No vision changes, no hearing difficulties, no dental abnormalities.
- Physical exam: Mild proptosis of the left eye. Extraocular movements intact. Pupils equal, round and reactive to light and accommodation. Left temple fullness compared to the right and left eyelid swelling.
- Pertinent Labs: None.

What Imaging Should We Order?

Select the applicable ACR Appropriateness Criteria

Variant 2: **Nontraumatic orbital asymmetry, exophthalmos, or enophthalmos. Initial imaging.**

Procedure	Appropriateness Category	RRL
MRI orbits without and with IV contrast	Usually Appropriate	O
CT orbits with IV contrast	Usually Appropriate	☢ ☢ ☢
CT orbits without IV contrast	May Be Appropriate	☢ ☢ ☢
CTA head and neck with IV contrast	May Be Appropriate	☢ ☢ ☢
MRA head and neck without and with IV contrast	May Be Appropriate	O
MRI head without and with IV contrast	May Be Appropriate	O
MRI orbits without IV contrast	May Be Appropriate	O
MRA head and neck without IV contrast	May Be Appropriate (Disagreement)	O
MRI head without IV contrast	May Be Appropriate	O
Arteriography cervicocerebral	May Be Appropriate	☢ ☢ ☢
CT head with IV contrast	May Be Appropriate	☢ ☢ ☢
CT head without IV contrast	May Be Appropriate	☢ ☢ ☢
CT head without and with IV contrast	Usually Not Appropriate	☢ ☢ ☢
CT orbits without and with IV contrast	Usually Not Appropriate	☢ ☢ ☢
X-ray orbit	Usually Not Appropriate	☢

These imaging modalities were ordered by the physician

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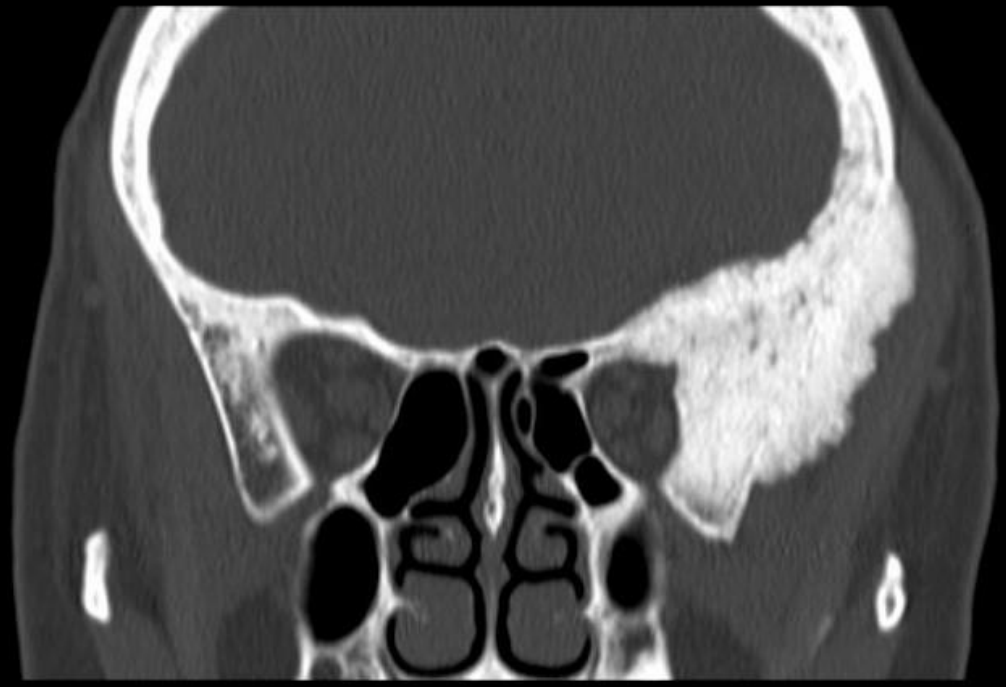
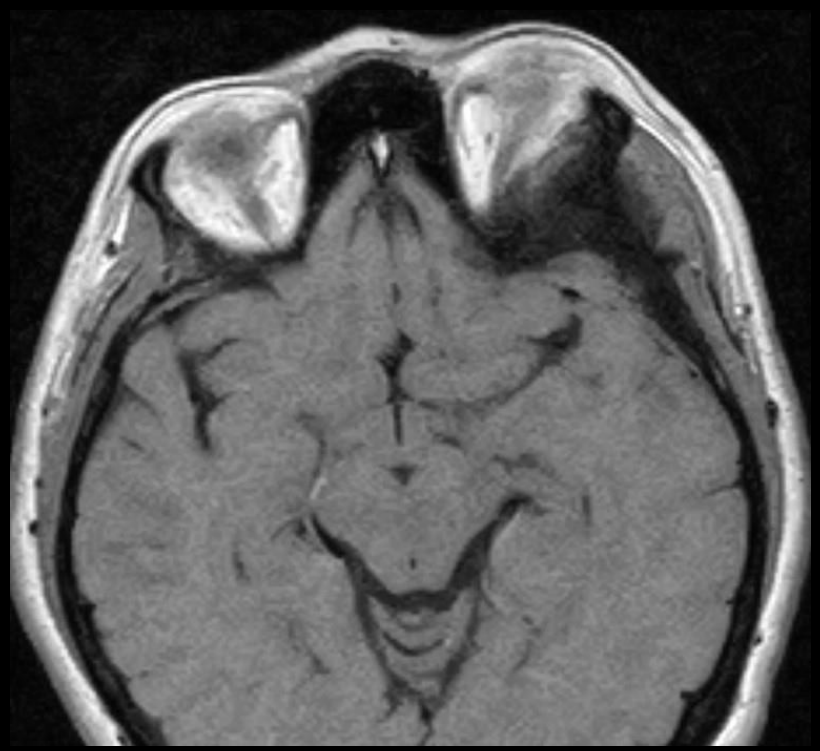
Findings (unlabeled)



CT Coronal

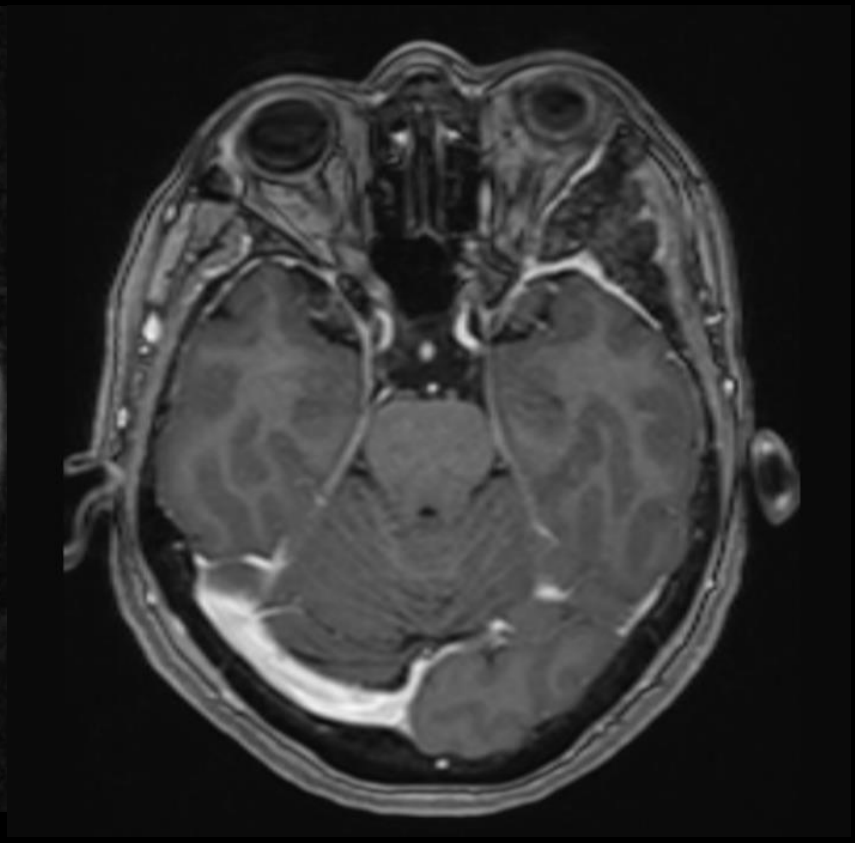
CT Axial

Findings (unlabeled)



T1 Axial

T2 Axial



T1 + Contrast

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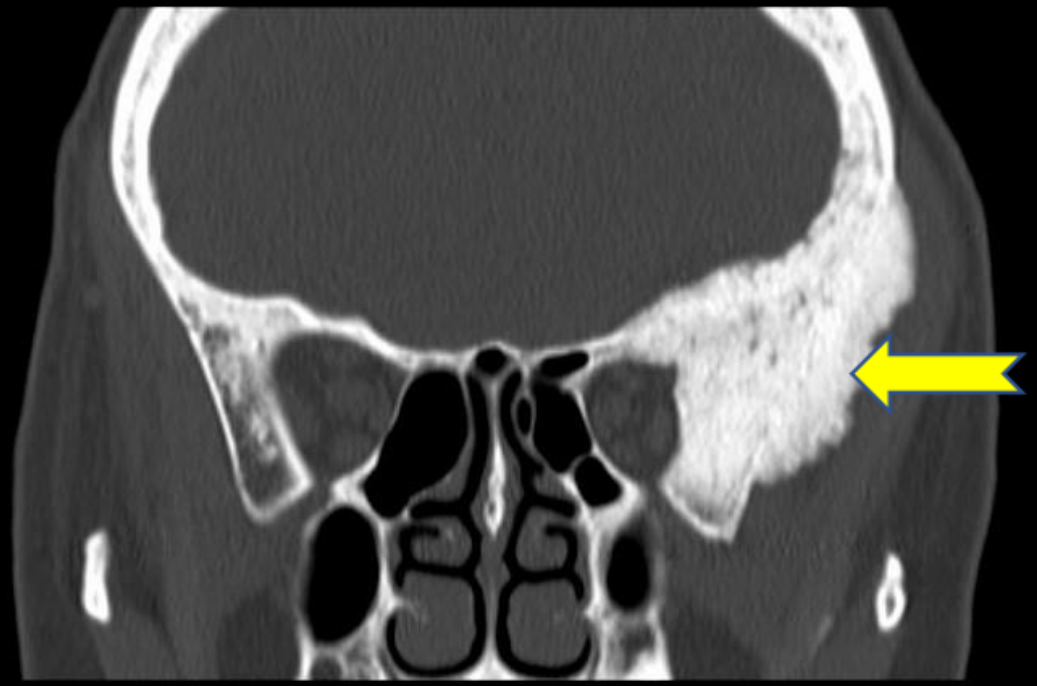
Findings: (labeled)

-Expansile sclerotic lesion with extensive hyperostosis of the left frontal, sphenoid, and squamosal temporal bones as well as the lateral wall of the left orbit.
-Associated left globe proptosis.



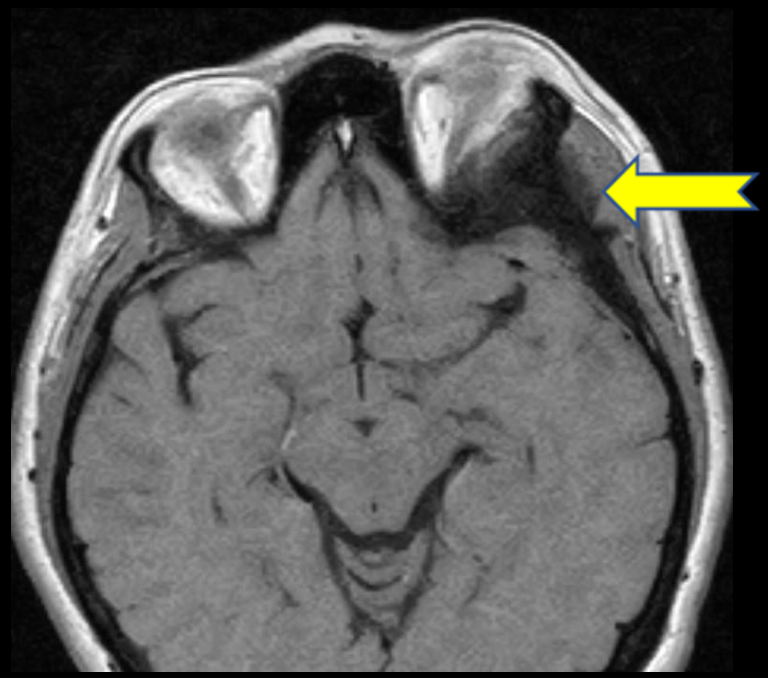
CT Coronal

CT Axial

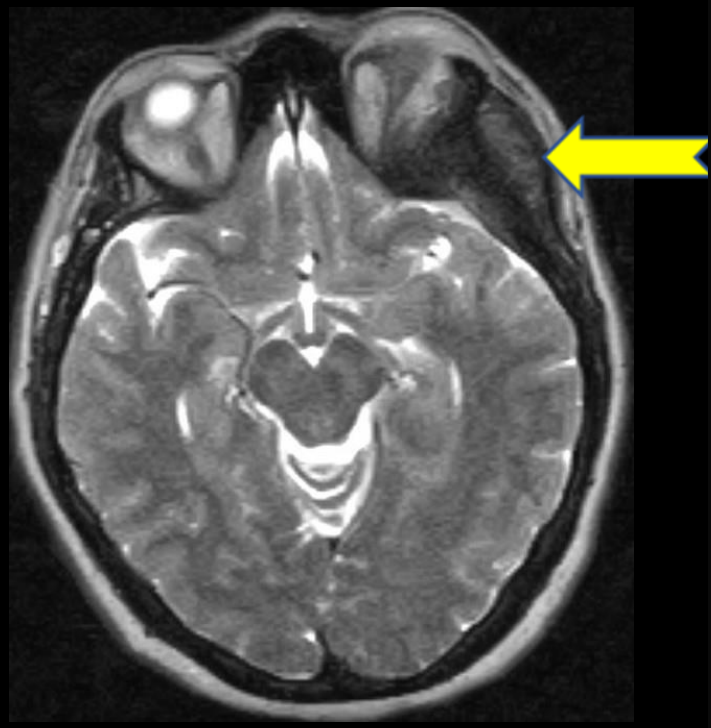
Findings: (labeled)

-T1/T2 hypointense expansile lesion involving the greater wing of the left sphenoid bone and lateral wall of the left orbit.

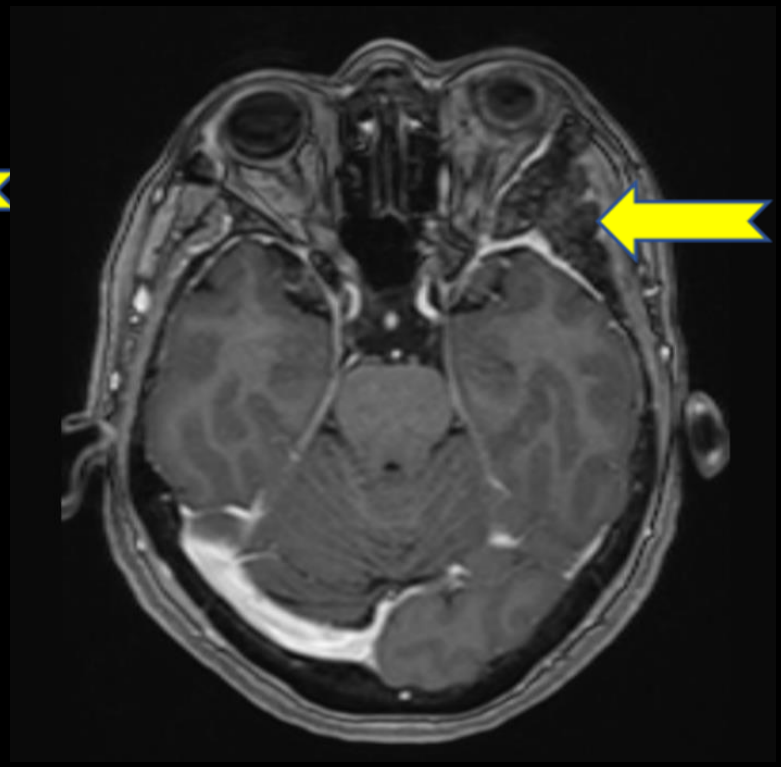
-Associated faint internal lesions enhancement with overlying dural enhancement of the middle cranial fossa and enhancement along the left lateral orbital wall.



MRI T1 Axial



MRI T2 Axial



MRI T1 + Contrast

Final Dx:

Primary Intraosseous Meningioma (PIM)

Primary Intraosseous Meningioma (PIM)

- Definition: A rare subtype of extradural meningiomas that originates within bone.
- Epidemiology:
  - Extradural compartment meningiomas are estimated to represent less than 2% of all meningioma cases.
  - Similarly to dural based meningiomas, PIMs occur more frequently in females.
- Etiology: Pathogenesis is unknown but is theorized to result from entrapment of arachnoid cap cells within the cranial sutures during birth.

Primary Intraosseous Meningioma (PIM)

- Clinical features:
  - Clinical presentations range from asymptomatic to significant neurological symptoms, including visual loss, exophthalmos, and headaches.
  - Cranial nerve/spinal cord compression and intracranial mass effect may cause variable symptoms.
- Differential Diagnosis:
  - Fibrodysplasia
  - Osteoma
  - Sclerotic metastasis
  - Paget disease of bone

Primary Intraosseous Meningioma (PIM)

- Radiographic features:
  - Computed tomography:
    - Iso- to hyperdense.
    - Approximately half of lesions will demonstrate sclerotic features, with the remainder variably displaying mixed or lytic changes.
  - MR Imaging:
    - Homogenous iso- to mildly hypointense on T1-weighted images.
    - T2-weighted images are typically hypo- to isointense.
    - Enhancement after contrast administration.

Primary Intraosseous Meningioma (PIM)

- Tumor Grading:
  - PIM grading follows the same WHO grade system as intracranial meningiomas.
  - Several studies suggest a higher proportion of PIMs are malignant when compared to intracranial meningiomas.
- Treatment:
  - If symptomatic, total tumor removal with wide surgical resection followed by cranial reconstruction is the treatment of choice.
  - In cases of unresectable or malignant PIM—radiotherapy, chemotherapy, and bisphosphonate therapy will be considered.

Primary Intraosseous Meningioma (PIM)

- Our Patient
  - The patient opted for definitive proton radiation therapy as the primary treatment. Surgery was discussed but given her lack of significant bothersome symptoms, the risks of surgery and necessity of extensive reconstruction were felt to outweigh the benefits. Furthermore, even with surgery, there might be a need for postoperative radiation therapy if all margins could not be adequately addressed.
  - Following treatment, she will receive long term imaging surveillance.

References:

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