

# AMSER Case of the Month

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A 31 y/o M presents with epistaxis, anosmia, and right-sided ocular proptosis

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# Patient Presentation

## HPI:

A 31-year-old male with a PMH of nasal polyposis who presented to the ENT clinic due to progressive nasal obstruction, epistaxis, loss of smell, and right-sided ocular proptosis. Denies any new onset headaches or vision loss.

## Objective:

- Vital signs WNL
- Physical Exam: Anterior rhinoscopy demonstrated diffuse, vascular nasal polyps extending to the nasal floor bilaterally with left-sided septal deviation. Maxillary sinus tenderness to palpation. Right-sided proptosis with lid lag on closure. Facial sensation intact.

What Imaging Should We Order?

# ACR Appropriateness Criteria

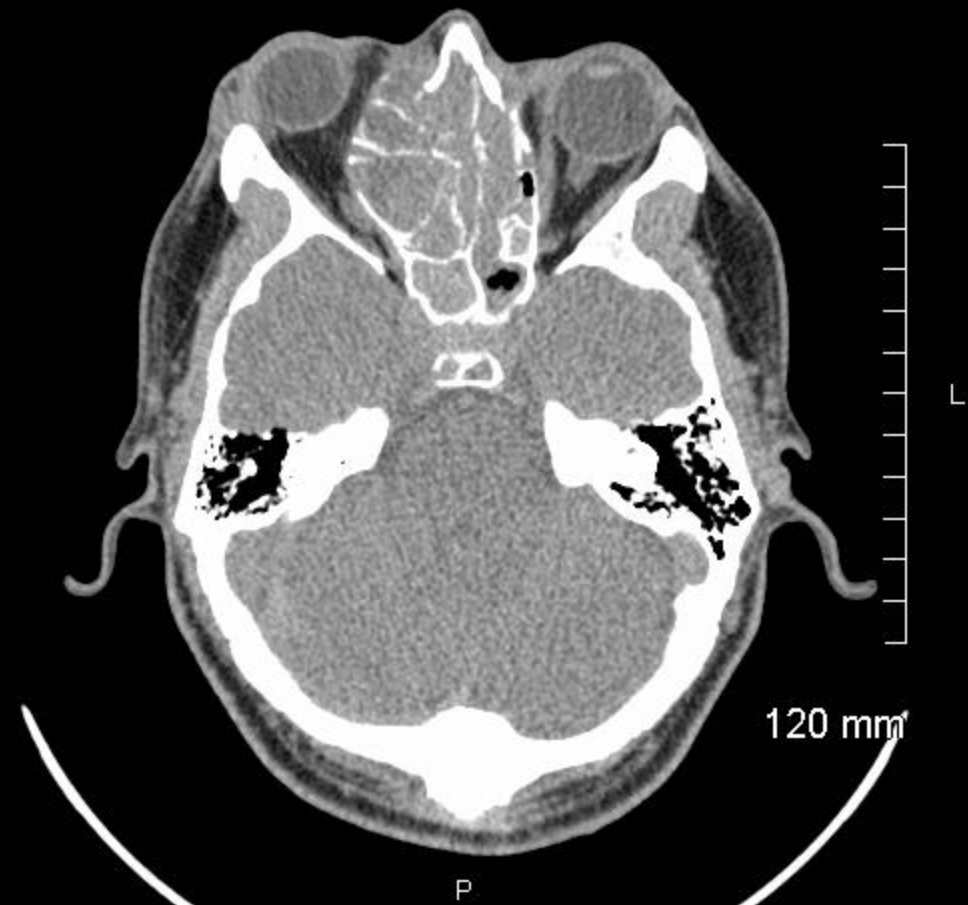
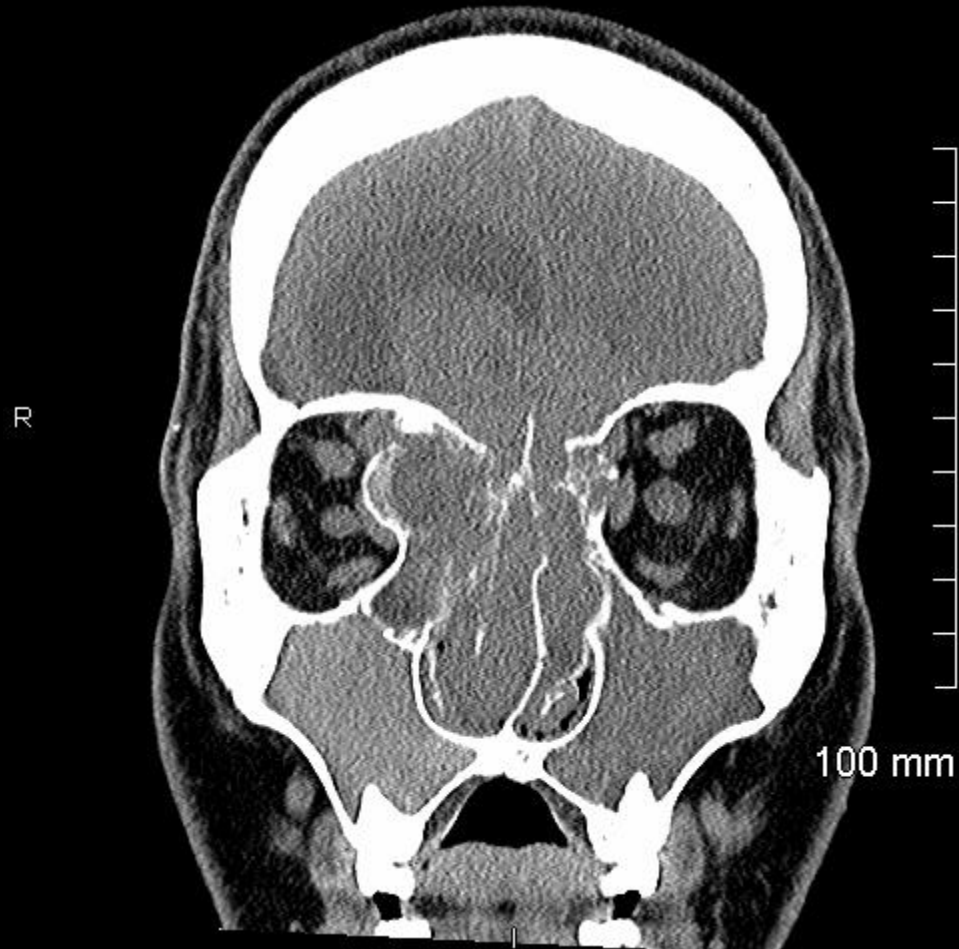
## Variant 5: Suspected sinonasal mass. Initial imaging.

Procedure	Appropriateness Category	Relative Radiation Level
MRI orbits face neck without and with IV contrast	Usually Appropriate	0
CT maxillofacial with IV contrast	Usually Appropriate	☼☼
CT maxillofacial without IV contrast	Usually Appropriate	☼☼
MRI head without and with IV contrast	May Be Appropriate	0
MRI head without IV contrast	May Be Appropriate	0
MRI orbits face neck without IV contrast	May Be Appropriate	0
CT head with IV contrast	May Be Appropriate	☼☼☼
Radiography paranasal sinuses	Usually Not Appropriate	☼
Arteriography craniofacial	Usually Not Appropriate	☼☼☼
MRA head with IV contrast	Usually Not Appropriate	0
MRA head without and with IV contrast	Usually Not Appropriate	0
MRA head without IV contrast	Usually Not Appropriate	0
MRI head with IV contrast	Usually Not Appropriate	0
MRI orbits face neck with IV contrast	Usually Not Appropriate	0
CT cone beam paranasal sinuses without IV contrast	Usually Not Appropriate	☼☼
CT head without and with IV contrast	Usually Not Appropriate	☼☼☼
CT head without IV contrast	Usually Not Appropriate	☼☼☼
CT maxillofacial without and with IV contrast	Usually Not Appropriate	☼☼☼
CTA head with IV contrast	Usually Not Appropriate	☼☼☼
SPECT or SPECT/CT paranasal sinuses	Usually Not Appropriate	☼☼☼
FDG-PET/CT skull base to mid-thigh	Usually Not Appropriate	☼☼☼☼

This imaging modality was ordered by the Otolaryngologist

# CT Findings unlabeled

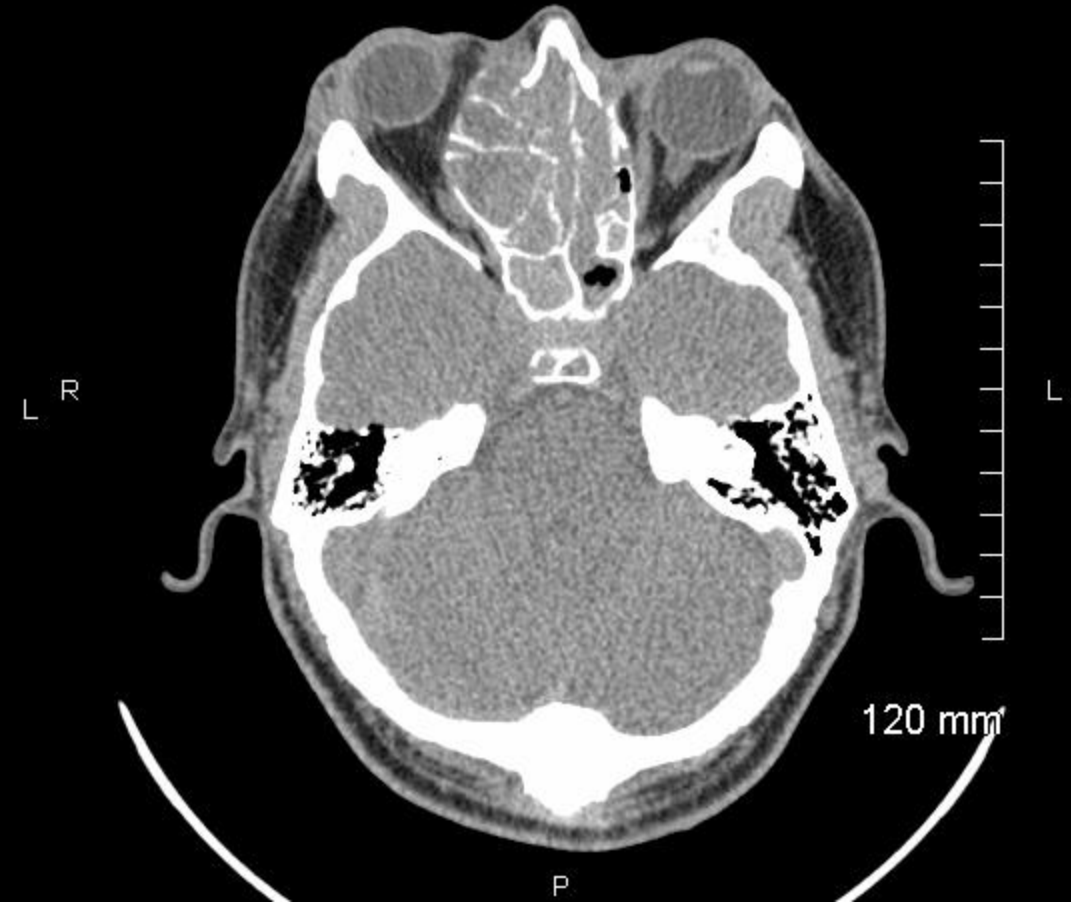
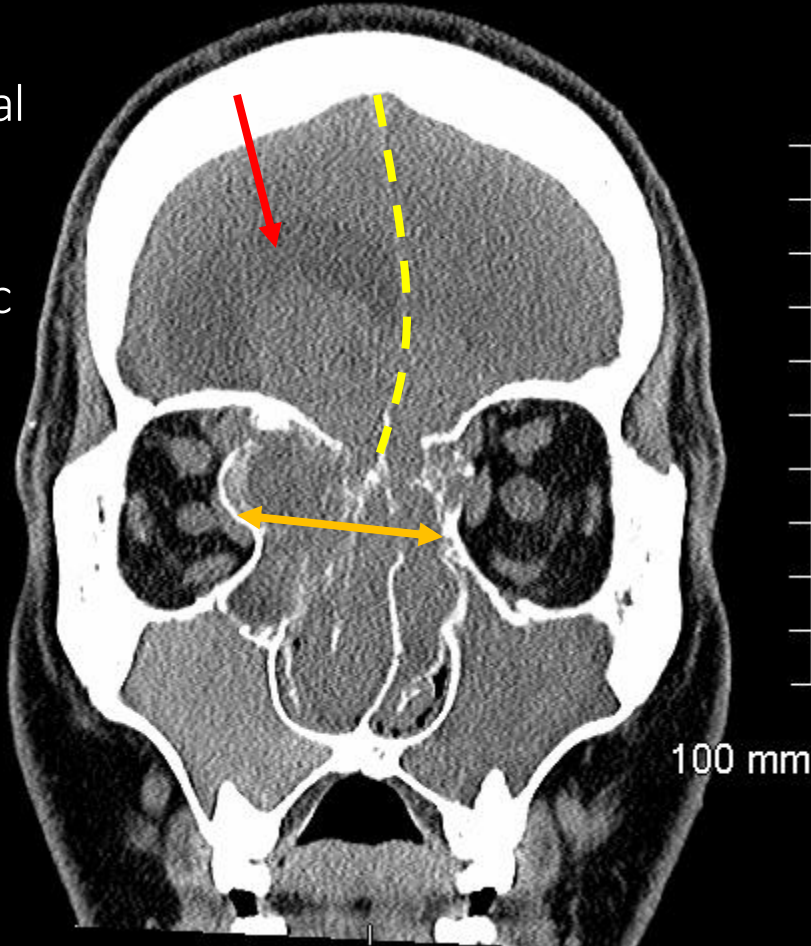
CT maxillofacial without IV contrast



# CT Findings labelled

Large lobulated mass that appears to originate at the olfactory recess with intracranial extension into the anterior cranial fossa and poorly characterized prominent involvement of the paranasal sinuses and nasal cavity (→)

Associated mass effect on the bilateral frontal lobes (- -) with moderate amount of vasogenic edema in the right frontal lobe (→)



What Follow-Up Imaging Should We Order?



# ACR Appropriateness Criteria

## Variant 4:

Adult. Suspected extraaxial brain tumor based on prior imaging. Pretreatment evaluation.

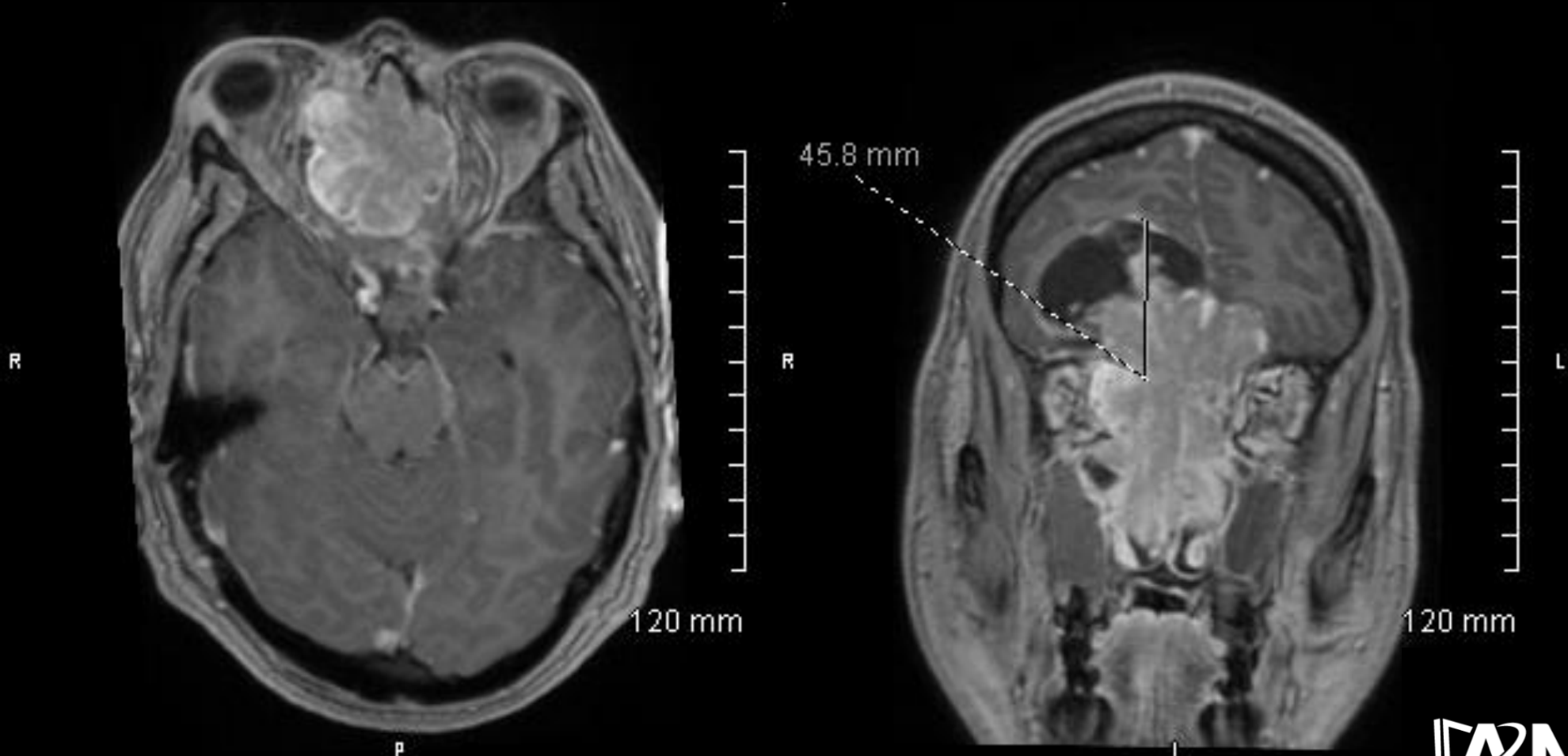
Procedure	Appropriateness Category	Relative Radiation Level
MRI head without and with IV contrast	Usually Appropriate	○
MRI complete spine without and with IV contrast	May Be Appropriate	○
MRI functional (fMRI) head without IV contrast	May Be Appropriate	○
MRI head without IV contrast	May Be Appropriate	○
DOTATATE PET/CT brain	May Be Appropriate	⊕⊕⊕
DOTATATE PET/MRI brain	May Be Appropriate	⊕⊕⊕
MR spectroscopy head without IV contrast	Usually Not Appropriate	○
MRI complete spine with IV contrast	Usually Not Appropriate	○
MRI complete spine without IV contrast	Usually Not Appropriate	○
MRI head perfusion with IV contrast	Usually Not Appropriate	○
MRI head perfusion without IV contrast	Usually Not Appropriate	○
MRI head with IV contrast	Usually Not Appropriate	○
MRI head without IV contrast with DTI	Usually Not Appropriate	○
CT head with IV contrast	Usually Not Appropriate	⊕⊕⊕
CT head without and with IV contrast	Usually Not Appropriate	⊕⊕⊕
CT head without IV contrast	Usually Not Appropriate	⊕⊕⊕
FDG-PET/CT brain	Usually Not Appropriate	⊕⊕⊕
FDG-PET/MRI brain	Usually Not Appropriate	⊕⊕⊕
Fluciclovine PET/MRI brain	Usually Not Appropriate	⊕⊕⊕
Fluciclovine PET/CT brain	Usually Not Appropriate	⊕⊕⊕

This imaging modality was ordered by the Otolaryngologist



# MRI Brain/Orbit with and without contrast

Post contrast T1 imaging demonstrates an extremely large invasive sinonasal mass infiltrating intracranially, invading the orbits and obstructing the nasopharyngeal airway.

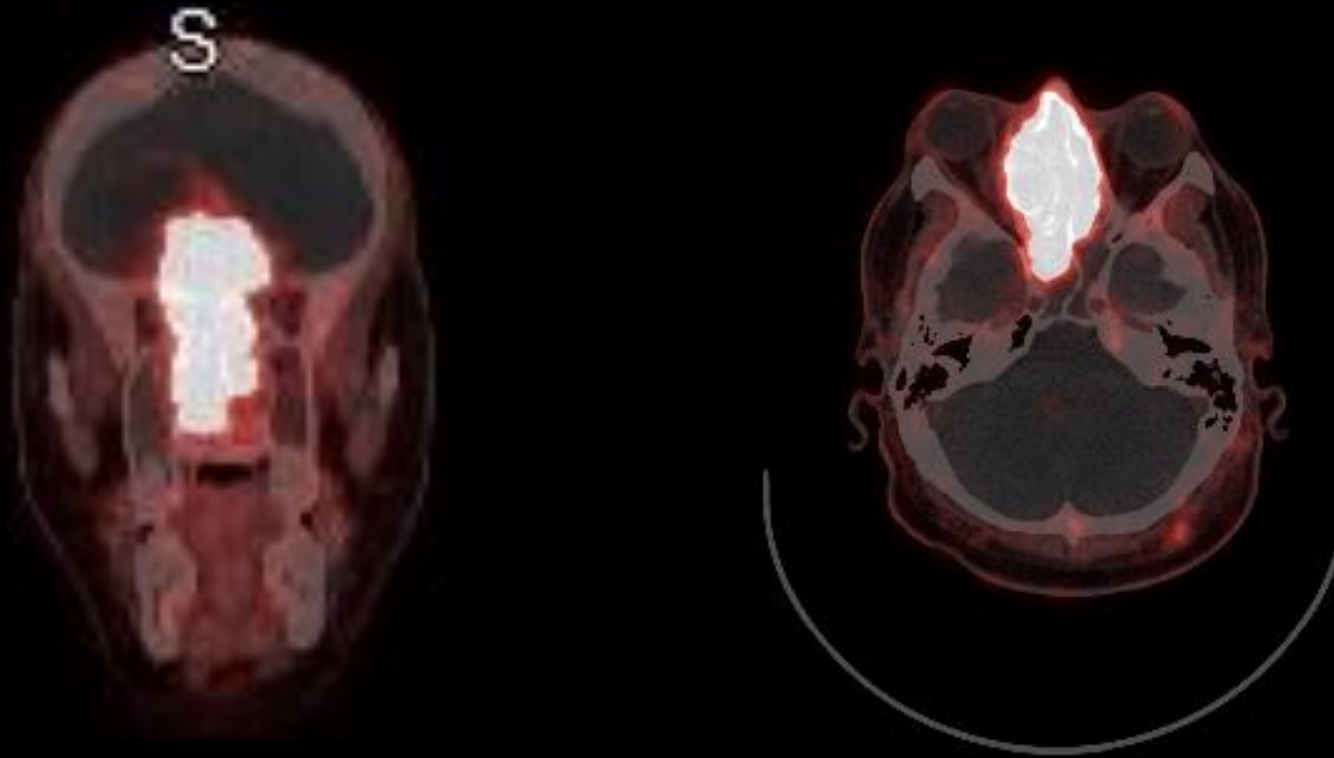


# DOTATATE PET/CT

- PET DOTATATE uses a somatostatin analog to identify neuroendocrine tumors, including olfactory neuroblastoma.<sup>2</sup>
- This imaging modality is used for initial staging, preoperative assessment, and detection of metastatic disease.
- In areas of high FDG PET/CT background activity, such as the brain and skull base, PET DOTATATE may reveal lesions not otherwise detected.<sup>2</sup>

# DOTATATE PET/CT

Prominent uptake of Dotatate within the large, invasive sinonasal mass with intracranial extension. No nodal disease or distant metastatic disease identified.



Final Dx:

Olfactory Neuroblastoma

# Case Discussion

- Olfactory neuroblastomas, also referred to as esthesioneuroblastomas, are tumors that originate from the olfactory epithelium of the superior nasal cavity.<sup>4</sup>
- The clinical presentation of olfactory neuroblastomas is often delayed with symptoms including rhinorrhea or epistaxis. Symptoms may persist for months and patients can develop anosmia when intracranial tumor extension is present (25-30% at diagnosis).<sup>3</sup>
- On imaging, masses typically emerge in the superior olfactory recess and involve the anterior and middle ethmoid air cells. Local bone destruction occurs as the tumor grows and may invade superiorly into the anterior cranial fossa and laterally into the orbits. Additionally, the tumor may invade across the midline into the contralateral nasal cavity or obstruct of the ostia of the paranasal sinuses.<sup>3</sup>
- Cervical and retropharyngeal nodal metastases are present in 10-44% of cases at diagnosis.<sup>5</sup>

# Case Discussion

- While CT cannot definitively distinguish olfactory neuroblastomas from other similar tumors, it helps assess bony destruction. Typically, it demonstrates a homogenous soft tissue mass with relatively uniform enhancement and may highlight focal calcifications.<sup>1</sup>
- MRI imaging demonstrates heterogenous intermediate signal on T1 and T2. As in this case, peritumoral cysts may be present if intracranial extension is present, suggestive of olfactory neuroblastoma.<sup>1</sup>
- Treatment often includes chemotherapy and/or radiotherapy with surgical excision. The presence of distant metastases significantly impacts prognosis. While the 5-year survival rate is 60% in the absence of metastases, the prognosis significantly worsens to 0% with distant metastases.<sup>5</sup>

# References:

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2. Dadgar, Habibollah PhD\*; Norouzbeigi, Nasim MD\*; Ahmadzadehfar, Hojjat MD, MSc†,‡; Assadi, Majid MD§. 68Ga-DOTATATE and 18F-FDG PET/CT for the Management of Esthesioneuroblastoma of the Sphenoclivar Region. Clinical Nuclear Medicine 45(8):p e363-e364, August 2020. | DOI: 10.1097/RLU.00000000000003133
3. Rosengren JE, Jing BS, Wallace S, Danziger J. Radiographic features of olfactory neuroblastoma. AJR Am J Roentgenol. 1979 Jun;132(6):945-8. doi: 10.2214/ajr.132.6.945. PMID: 108974.
4. Sasajima T, Kinouchi H, Tomura N, Watarai J, Mizoi K. High uptake of 123I-metaiodobenzylguanidine related to olfactory neuroblastoma revealed by single-photon emission CT. AJNR Am J Neuroradiol. 2000 Apr;21(4):717-20. PMID: 10782784; PMCID: PMC7976631.
5. Zollinger LV, Wiggins RH 3rd, Cornelius RS, Phillips CD. Retropharyngeal lymph node metastasis from esthesioneuroblastoma: a review of the therapeutic and prognostic implications. AJNR Am J Neuroradiol. 2008 Sep;29(8):1561-3. doi: 10.3174/ajnr.A1114. Epub 2008 May 22. PMID: 18499797; PMCID: PMC8119040.