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Radiologic Findings of an Advanced Sinonasal Tumor

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Agenda

- Our patient presentation
- Differential diagnoses
- Menu of radiologic tests
- Relevant anatomy
- Radiologic findings
- Pathology findings
- Small cell sinonasal carcinoma



Our patient – initial presentation

- 58 y/o female
- CC: sinus pain, right facial pain for 2 weeks
- HPI:
 - Dull, pressure-like pain across right cheek and forehead
 - Initially responded to course of Augmentin (amoxicillin clavulanate)
 - Worsening symptoms in past week
 - Blurry vision in right eye
 - No fever, chills



Our patient – clinical history

- PMH:
 - DMII
 - HTN
 - Asthma
- Family Hx: non-contributory
- Physical Exam:
 - HEENT: oropharynx w/o eschar, R mild facial swelling
 - Neuro: cranial nerves intact



Differential diagnoses for sinus pain

- Vascular:
 - Cerebral venous thrombosis
- Infection:
 - Rhinosinusitis
 - Mucormycosis
- Neoplasm (benign):
 - Sinonasal polyposis
 - Mucocele
- Neoplasm (malignant):
 - Olfactory neuroblastoma
 - Sinonasal carcinoma
 - Sinonasal Ewing's sarcoma
 - Metastatic cancer (e.g. melanoma, NHL)



The vast majority of people with sinus pain just have rhinosinusitis (aka the common cold)...

How do you decide who needs imaging and further workup?



Indications for imaging sinonasal disease

- Imaging is indicated in 2 circumstances:
 - 1) chronic rhinosinusitis
 - 2) suspected mass lesion
- Our patient:
 - Worsening sinusitis despite Augmentin treatment
 - Focal neurologic symptoms (blurry vision), suspicion for mass lesion



Now that you've decided this patient needs imaging and further workup...

Which radiological studies should you order?



Menu of radiologic tests

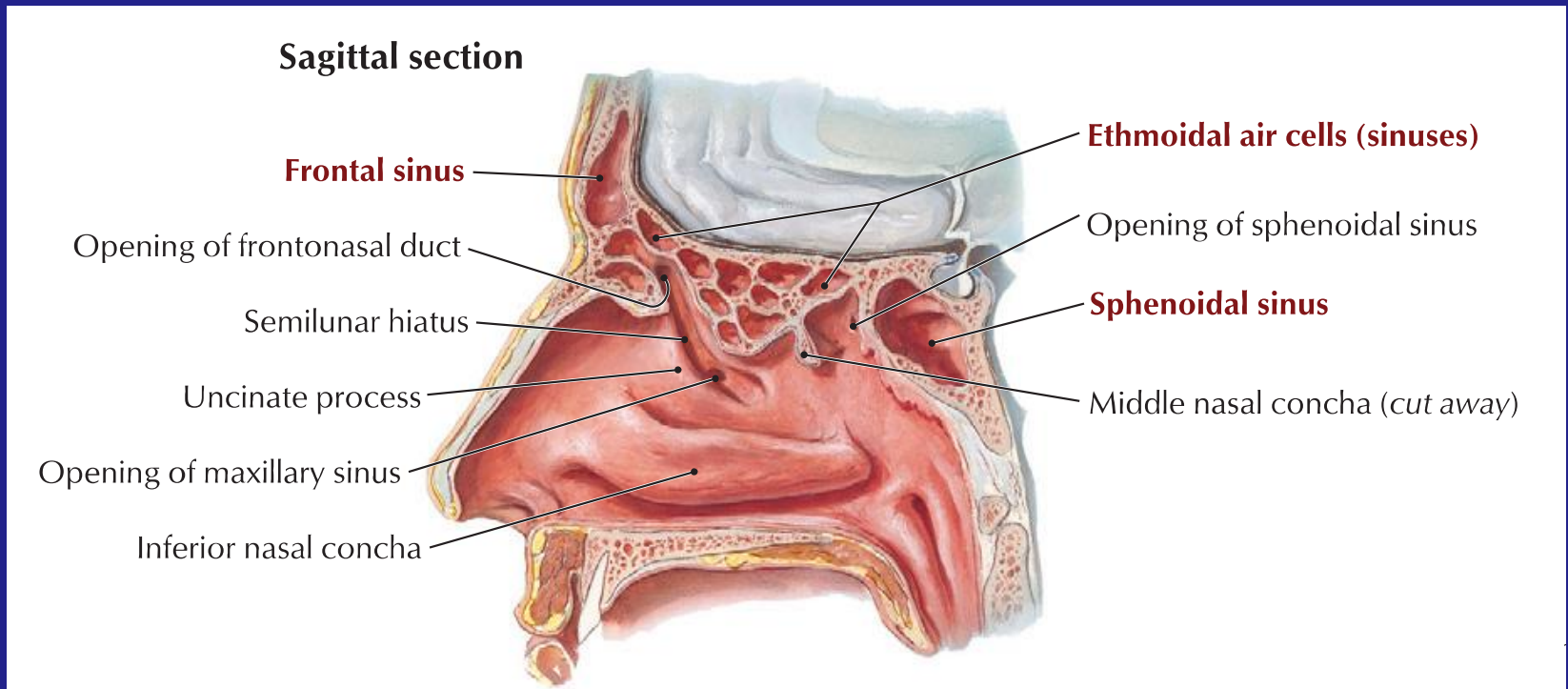
- CT scan
 - Best for anatomical detail of paranasal sinuses
 - Useful for staging of disease, especially bony erosions
- MRI
 - Complementary to CT study
 - Visualizes sinonasal, intraorbital and intracerebral invasion of mass
- SPECT (single positron emission computed tomography)
 - $^{123}\text{MIBG}$ radiotracer localizes to neuroblastomas
- Digital subtraction angiography
 - Visualizes vasculature
 - Relatively more invasive than CTA



Before looking at the images, let's
review some sinonasal anatomy

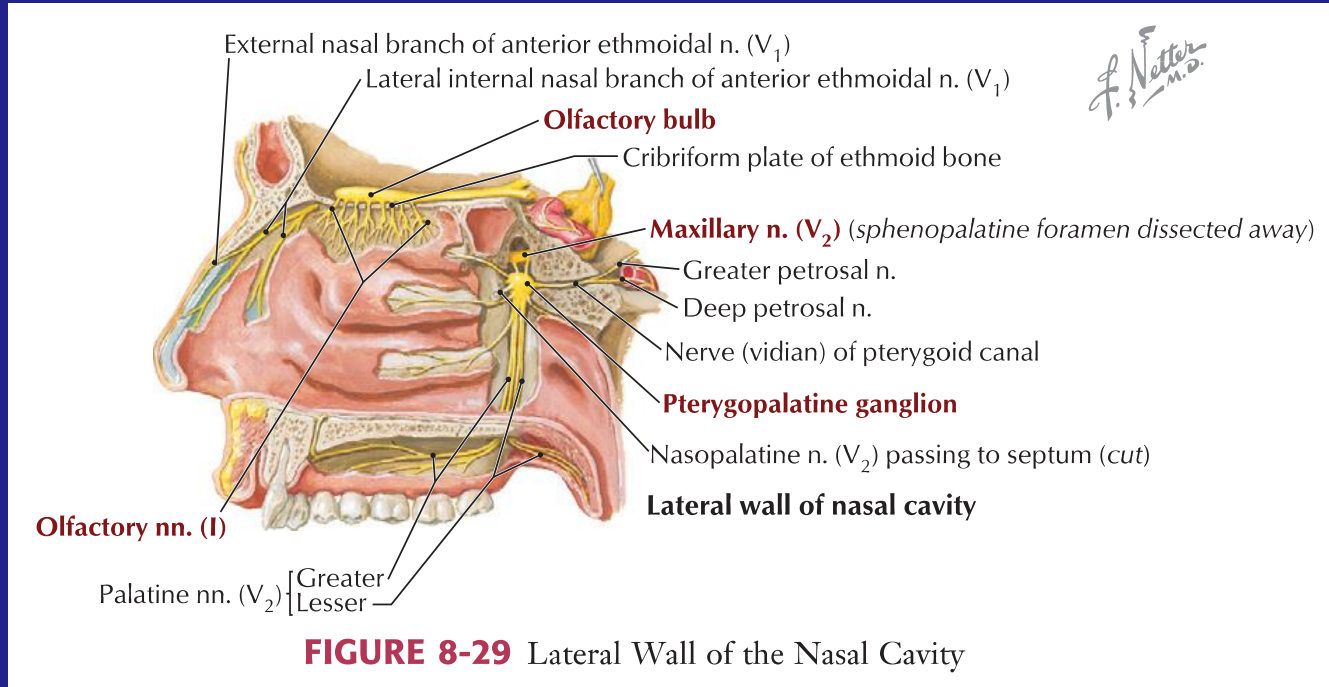


Paranasal sinus anatomy



- There are 4 sinuses: frontal, ethmoid, sphenoid, maxillary (not pictured)
- Sinuses secrete mucous to moisturize nose, protect from microorganisms
- Sinuses drain mucous via openings called ostium, a tumor obstructing an ostium could cause unilateral swelling and sinus pain

Olfactory nerve anatomy



J. Netter M.D.

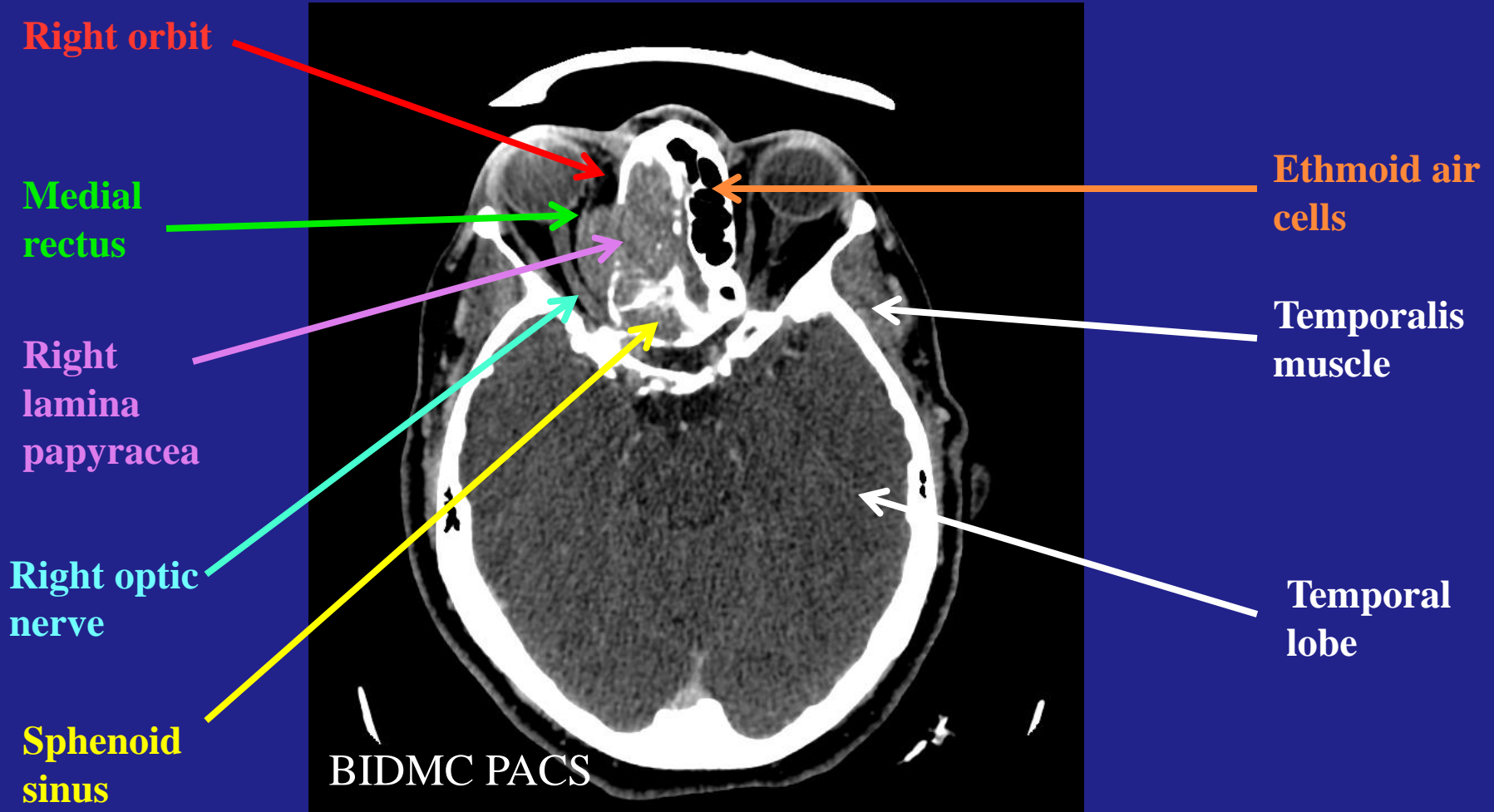
- The olfactory nerve arises from mucosa in the upper part of the nasal cavity, then extends through the cribriform plate within the ethmoid bone to synapse on the olfactory bulb in the brain
- The cribriform plate is a passageway through which sinus infections (especially *Naegleria fowleri*) and cancer can invade the brain



The initial imaging study is almost always CT, because:

- 1) it provides anatomical bony detail*
- 2) it's fast (on the order of seconds)*
- 3) it's relatively inexpensive*

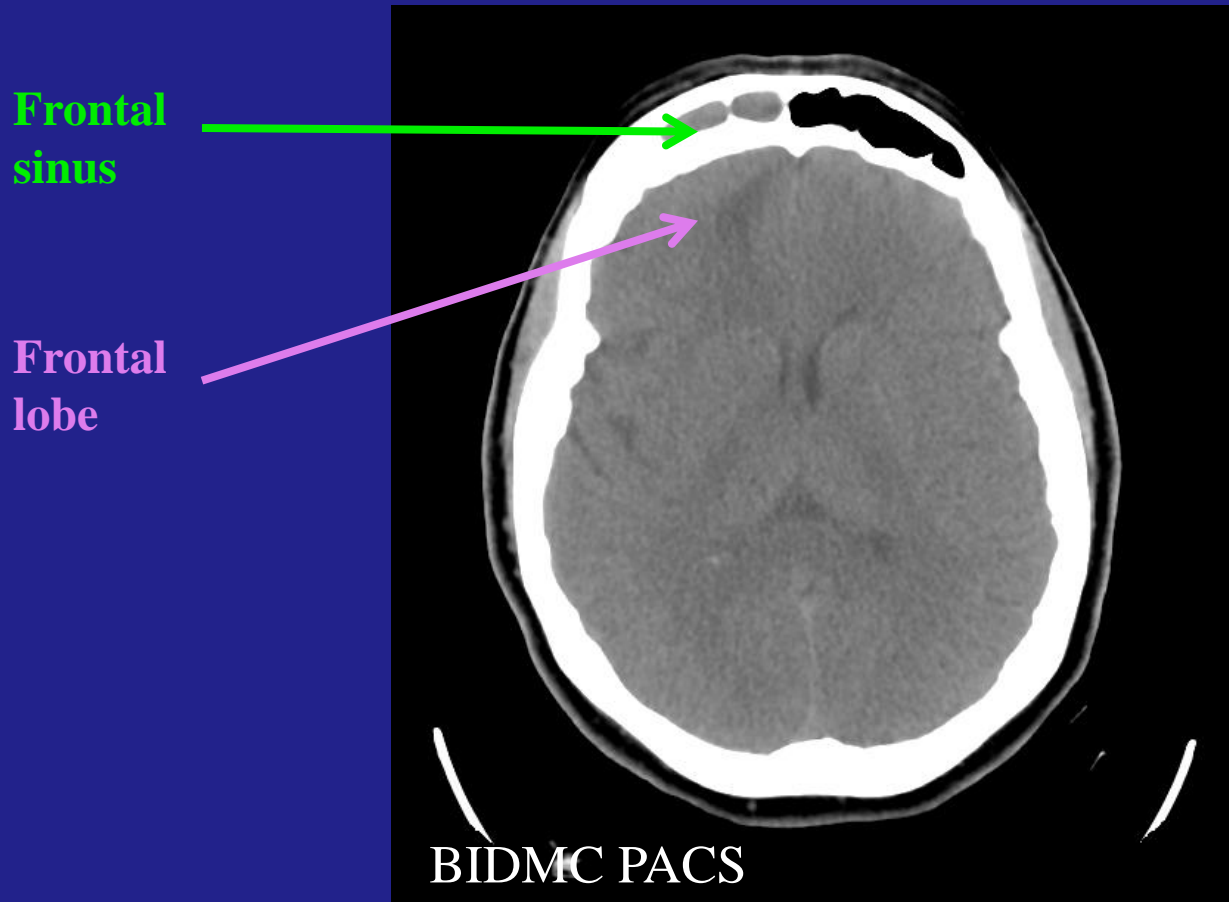
Our patient – C+ Axial view CT showing mass



- There is a soft tissue mass occupying the sphenoid & ethmoid sinuses
- Mass invades into right orbit, causing dehiscence of papyracea bone and impinging on the medial rectus



Our patient – C+ Axial view CT showing frontal sinus & lobe involvement



- Enhancement of right frontal sinus suggests tumor invasion
- Inhomogeneity of right medial frontal lobe may be edema surrounding invasive tumor

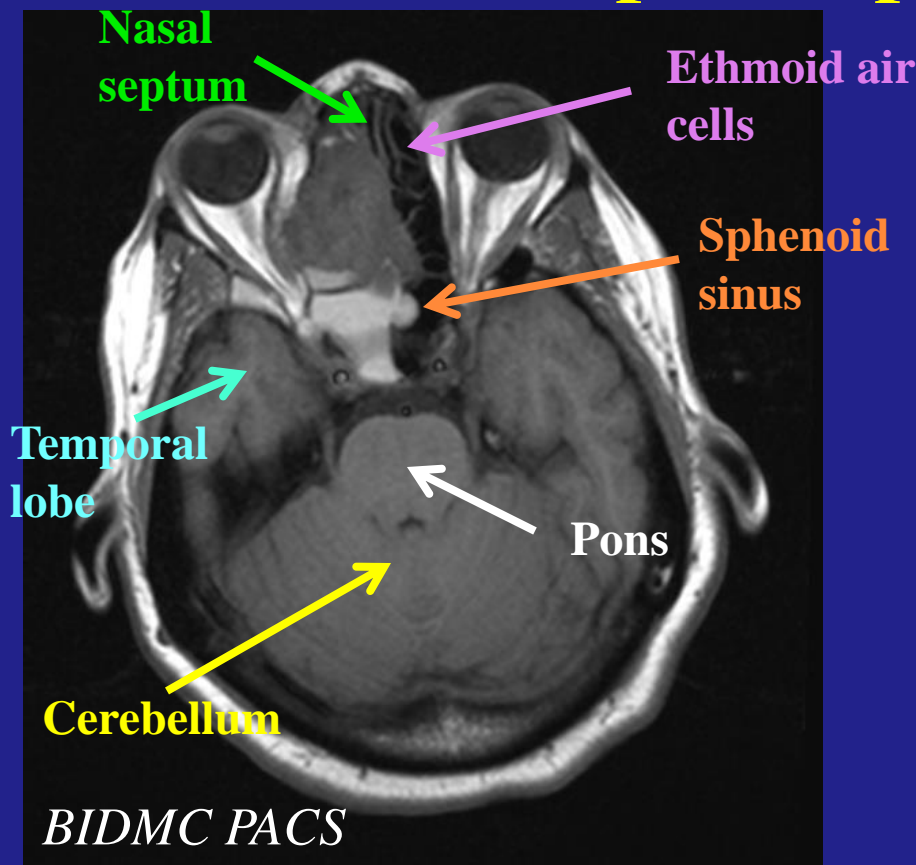


Positive findings on CT should be followed up with an MRI study...

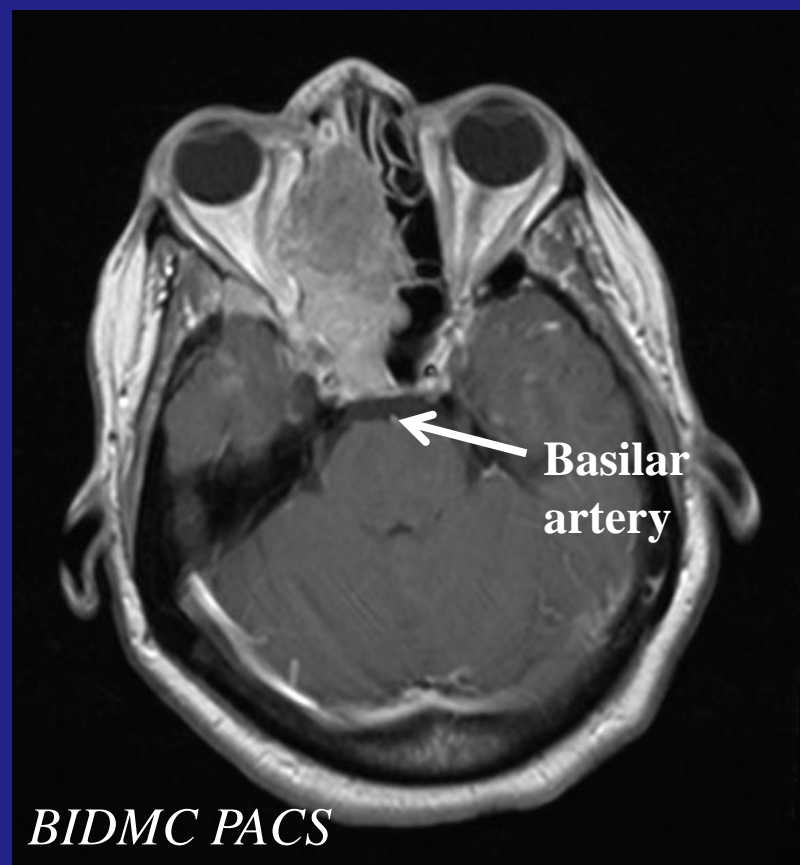
MRI can provide more soft tissue detail (and tumors are comprised of soft tissue!)



Our patient: Contrast-enhancing mass on T1 MRI pre & post-Gadolinium



T1, Pre-Gd



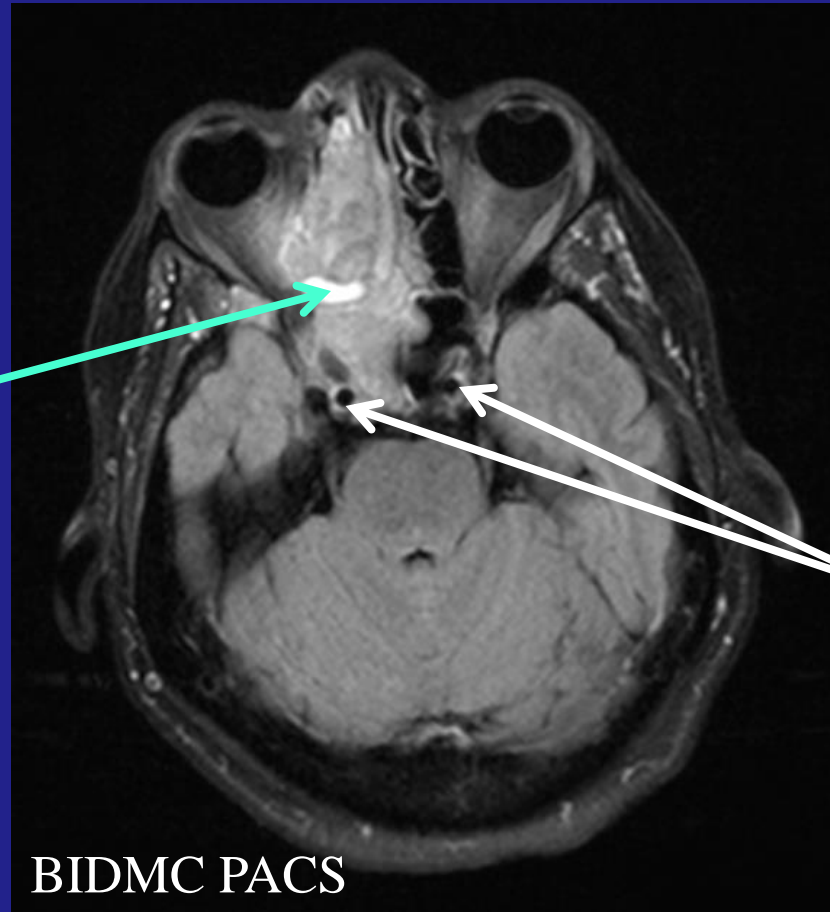
T1, Post-Gd

- MRI visualizes the mass with greater soft tissue detail than CT
- Mass enhances with Gd, suggesting tumor has infiltrated blood-brain-barrier



Our patient – MRI T2 FLAIR axial view showing edema surrounding mass

Edema

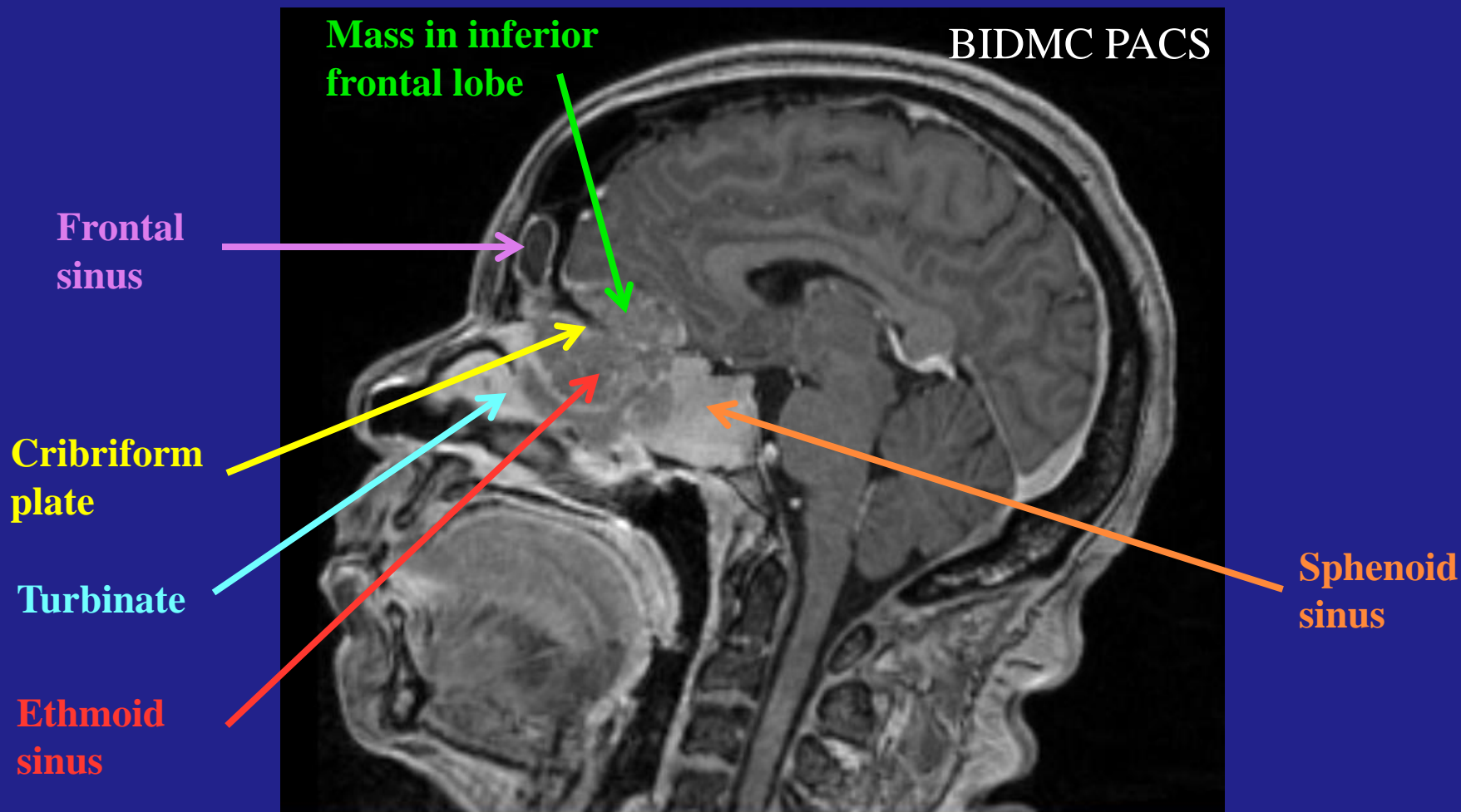


Internal carotid
arteries

- FLAIR sequences highlight edema
- Hyperintensity surrounding mass indicates reactive edema and inflammation



Our patient – MRI T1 sagittal view showing mass extending through cribriform plate



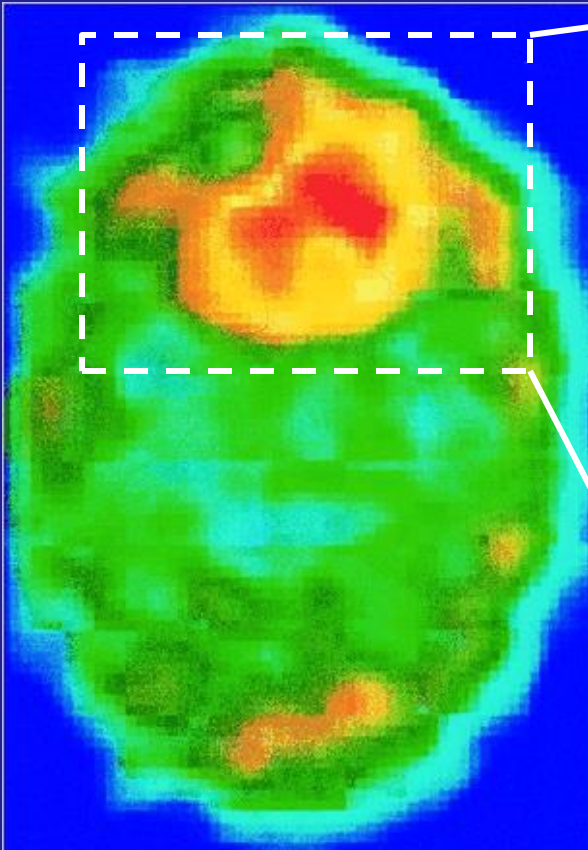
- On sagittal view, mass can be seen extending from ethmoid sinus through cribriform plate into medial inferior frontal lobe



CT and MRI are standard of care...

*Following are less-commonly used
SPECT and subtraction angiogram
studies on comparison patients*

Comparison Patient #1: ^{123}I -MIBG SPECT



- Warm colors indicate areas of relative radiotracer accumulation
- ^{123}I -MIBG preferentially localizes to neuroblastomas, thus SPECT can be used as a confirmatory diagnostic test
- This radiological finding is consistent with bilateral frontal lobe olfactory neuroblastomas

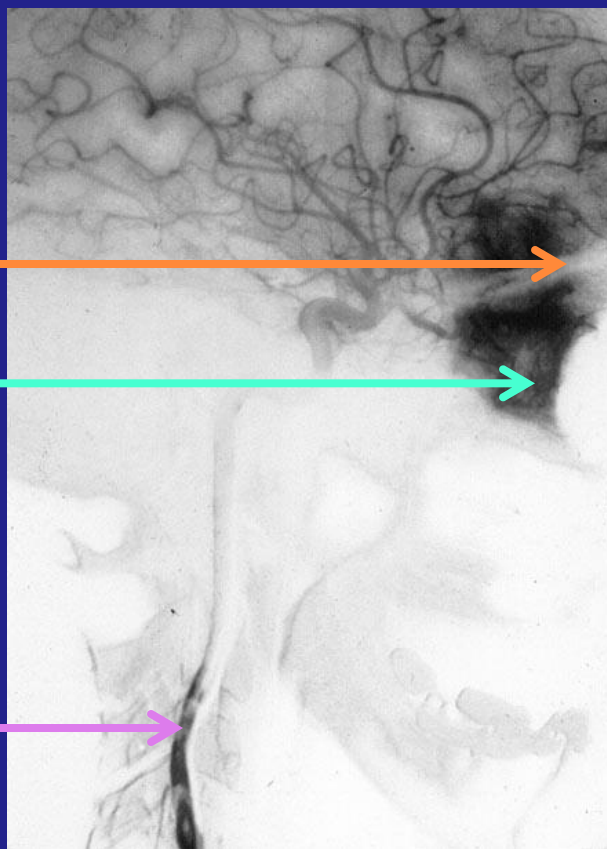


Comparison Patient #2: Subtraction angiogram

**Cribriform
plate**

**Tumor in
ethmoid
sinus**

**Common
carotid
artery**



- Subtraction angiogram visualizes vascular structures
- Fast-growing tumors induce angiogenesis, increasing the density of blood vessels in the region of the tumor
- This will appear hyperintense on an angiogram, as seen here with a tumor crossing the cribriform plate
- This test is non-specific and relatively more invasive than a CTA, so it's rarely used



When radiological studies are only
able to narrow the differential...

*We then proceed to biopsy and
pathology studies for diagnostic
clarification*



Our patient – Pathology findings

- Cytology:
 - Monotonous sheets of medium-sized cells with coarse chromatin and scant cytoplasm
 - High mitotic activity
- Staining:
 - Cytokeratin: diffusely positive
 - Chromogranin: patchy positive
 - Synaptophysin: patchy positive
 - S-100: focal staining at periphery of tumor lobules
- Conclusion:
 - Likely olfactory neuroblastoma
 - Cytokeratin staining is atypical, consider also sinonasal small cell neuroendocrine carcinoma and sinonasal Ewing sarcoma



Our patient: Clinical course

- Presumptive diagnosis: small cell sinonasal carcinoma
 - Olfactory neuroblastoma deemed less likely because of diffuse cytokeratin staining
- Soon after, patient developed monocular vision loss in right eye and was admitted to begin treatment
- CT scans showed interval enlargement of mass with infiltration into optic nerve
- Patient received high-dose steroids and cisplatin/etoposide induction chemotherapy (now on cycle 3)
- Chemotherapy succeeded in shrinking tumor and improving extraocular eye motility, though right eye still has no light perception



Small cell sinonasal carcinoma

- Epidemiology:
 - Extrapulmonary manifestations of small cell carcinoma (SCC) account for only 4% of SCC cases
 - Sinonasal SCC is even more rare, only 76 cases have been reported
- Presentation:
 - Symptoms of nasal obstruction, discharge and recurrent epistaxis are often mistaken for benign etiologies
 - This delays diagnosis, so patients often present in advanced stages
- Histology:
 - Indistinguishable from pulmonary SCC
 - Sheets of small blue cells arranged in sheets or nests, cells have scant cytoplasm with hyperchromatic nuclei
- Management & Prognosis:
 - No management guidelines exist because tumor is so rare
 - Chemotherapy using cisplatin/etoposide followed by radiotherapy has been effective in some patients
 - Surgery is only reserved for chemotherapy non-responders
 - One and five-year survivals are approximately 57% and 10%



Summary – clinical findings

- Differential diagnosis for sinus pain is broad, ranging from benign causes like rhinosinusitis (the common cold) to malignant neoplasms like high-grade sinonasal carcinoma
- Diagnosis requires both imaging and pathology studies
- Sinonasal carcinomas often present as advanced stage disease, even with chemotherapy and adjunct radiation, thus prognosis is poor



Summary – radiological findings

- There are 2 indications for imaging:
 - 1) worsening sinusitis symptoms despite treatment
 - 2) suspected mass lesion (e.g. focal neurological signs)
- Menu of radiologic tests:
 - CT: visualizes bony anatomy, useful for staging disease
 - MRI: complementary to CT study, better for soft tissue
 - SPECT: ^{123}I -MIBG radiotracer localizes to neuroblastomas, is diagnostic for olfactory neuroblastomas
 - Digital subtraction angiography: visualizes highly vascular areas, such as tumors
- Our patient's radiological findings:
 - Large soft tissue mass occupying right frontal, ethmoid and sphenoid sinuses causing osseous dehiscence and invading into right inferior frontal lobe and right orbit
 - Mass enhances with contrast and has surrounding edema



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