

AMSER Case of the Month

September 2020

46 yo M with progressive dysphagia and decreased sensation of the right face, chest, and arms.

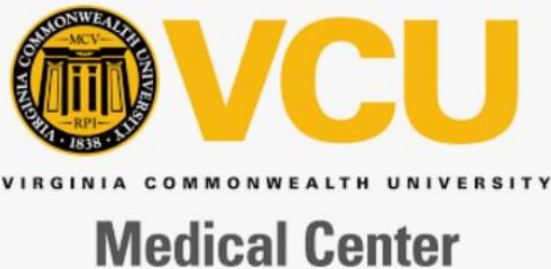
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Patient Presentation

- **HPI:** 46 yo M presents with 6 wks of decreasing PO intake and unintentional weight loss. Initial symptoms started in 2017, pt was evaluated by GI and underwent dilatation for stricture. Pt has also had ongoing voice hoarseness for which he underwent a procedure to treat a lateralized vocal cord in September 2019. Pt notes a significant change in symptoms since November 2019 when he developed a progressive intolerance to liquids and reflux of food. Over the same time, pt endorses decreased sensation to cold over his right face, chest, and arms.
- **ROS:** Denies odynophagia, fevers, chills, abdominal pain, headache, numbness, tingling, vomiting, diarrhea, and chest pain.
- **PMHx:** Acid reflux, chronic venous insufficiency, hoarseness of voice, thyroglossal cyst, hypertension, obesity, OSA
- **PSHx:** Non contributory
- **Family Hx:** Mother and father with heart disease.
- **Social Hx:** Denies tobacco or ILD use. Drinks alcohol 2-3x/ wk

Pertinent Labs

- BMP within normal limits
- CBC within normal limits

What Imaging Should We Order?

ACR Appropriateness Criteria

Variant 6: Delayed (greater than 1 month) postoperative development of dysphagia. Oropharyngeal or retrosternal. Initial imaging.

Procedure	Appropriateness Category	Relative Radiation Level
CT neck and chest with IV contrast	Usually Appropriate	⊕⊕⊕⊕
Fluoroscopy single contrast esophagram	Usually Appropriate	⊕⊕⊕
Fluoroscopy barium swallow modified	May Be Appropriate	⊕⊕⊕
Fluoroscopy biphasic esophagram	May Be Appropriate	⊕⊕⊕
Esophageal transit nuclear medicine scan	May Be Appropriate	⊕⊕⊕
CT neck and chest without and with IV contrast	Usually Not Appropriate	⊕⊕⊕⊕
CT neck and chest without IV contrast	Usually Not Appropriate	⊕⊕⊕⊕
Fluoroscopy pharynx dynamic and static imaging	Usually Not Appropriate	⊕⊕⊕

This imaging modality was ordered by the ER physician



ACR Appropriateness Criteria

Variant 4: New focal neurologic defect, fixed or worsening. Longer than 6 hours. Suspected stroke.

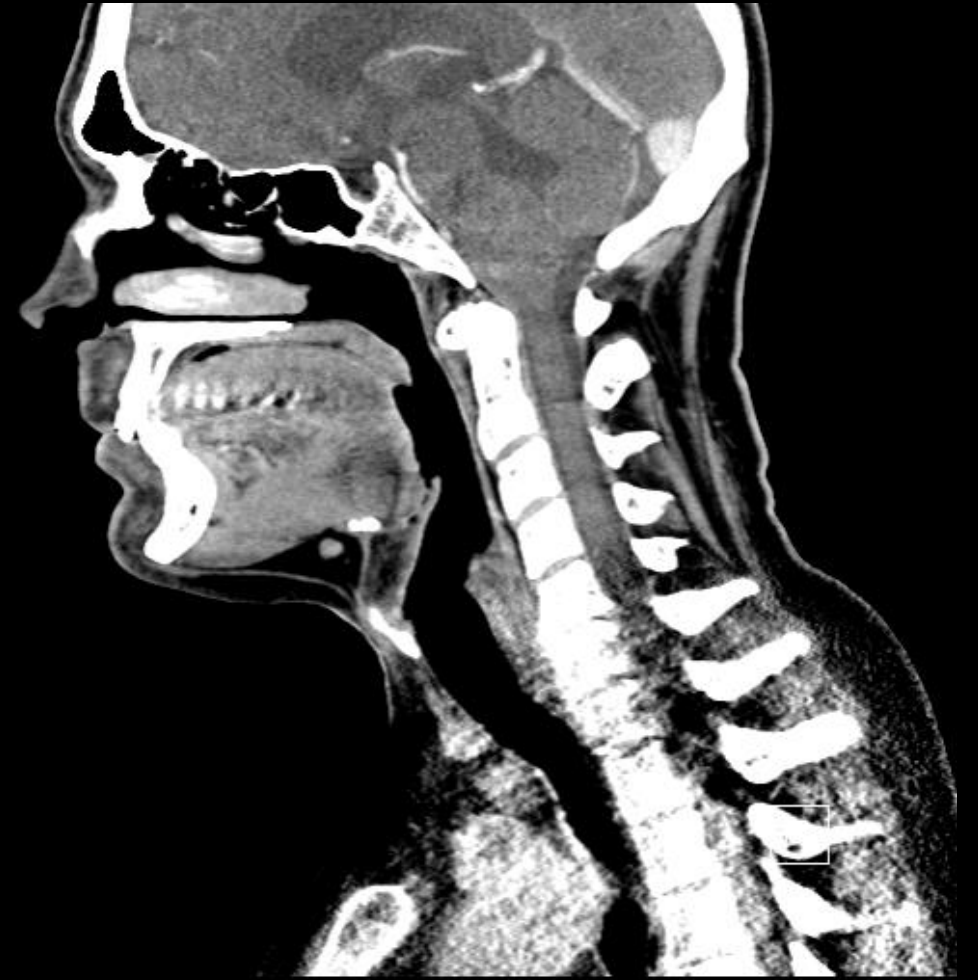
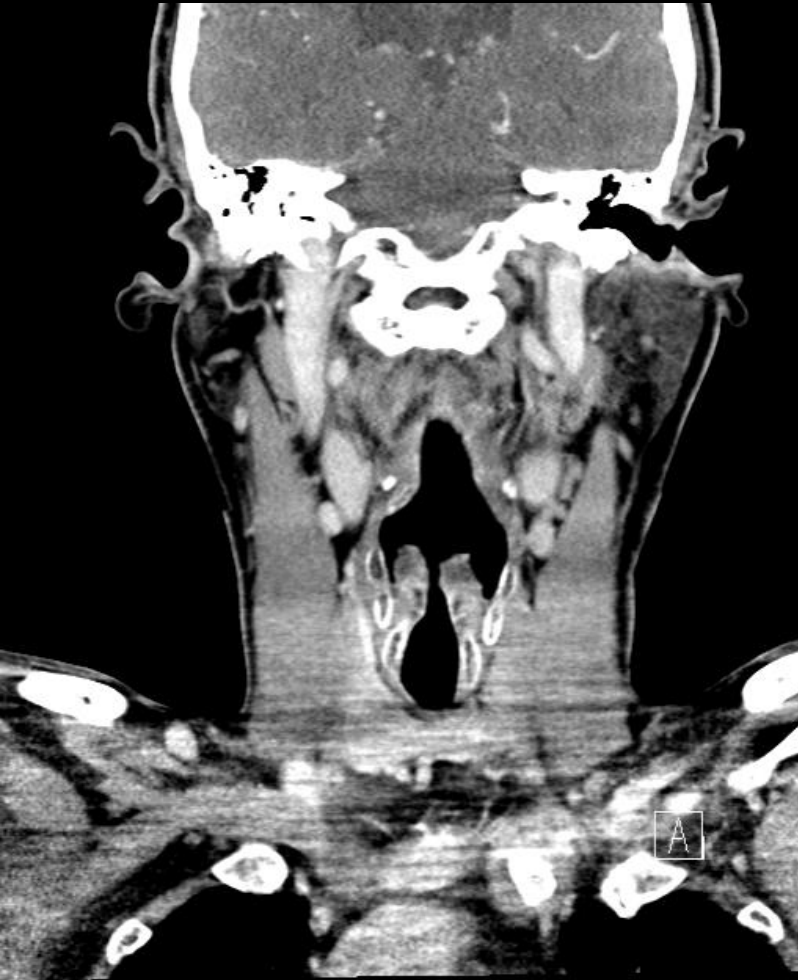
Radiologic Procedure	Rating	Comments	RRL.*
MRI head without IV contrast	8	Parenchymal brain imaging and CT or MR vascular imaging of the head and neck should be considered. Noncontrast head CT is often obtained first to assess for hemorrhage or large infarct. Can be useful if there is a contraindication to contrast. MRI is more sensitive than CT for acute infarct.	0
MRI head without and with IV contrast	8	Parenchymal brain imaging and CT or MR vascular imaging of the head and neck should be considered. Noncontrast head CT is often obtained first to assess for hemorrhage or large infarct. MRI is more sensitive than CT for acute infarct.	0
MRA head and neck without IV contrast	8	Can be obtained in conjunction with MRI head. Preferred MR vascular imaging of the head and neck includes noncontrast head MRA and contrast-enhanced neck MRA. May be useful in patients with renal failure or contrast allergies.	0
MRA head and neck without and with IV contrast	8	Can be obtained in conjunction with MRI head. Preferred MR vascular imaging of the head and neck includes noncontrast head MRA and contrast-enhanced neck MRA.	0
CT head without IV contrast	8	Noncontrast head CT is often obtained first to assess for hemorrhage or large infarct. MRI is more sensitive than CT for acute infarct.	☼☼☼
CTA head and neck with IV contrast	8	CTA can be obtained after NCCT.	☼☼☼
Arteriography cervicocerebral	6		☼☼☼
CT head perfusion with IV contrast	5		☼☼☼
MRI head perfusion with IV contrast	5		0
CT head with IV contrast	3		☼☼☼
CT head without and with IV contrast	3		☼☼☼
US duplex Doppler carotid	2		0

Rating Scale: 1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate

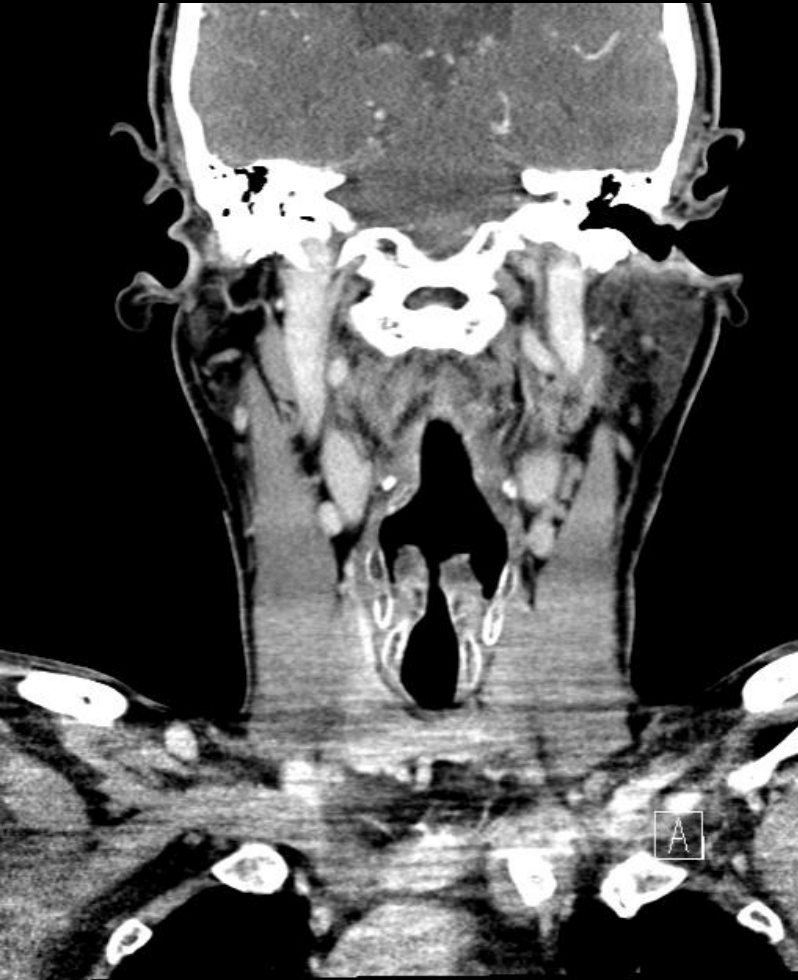
*Relative Radiation Level

This imaging modality was ordered by the attending physician upon admission

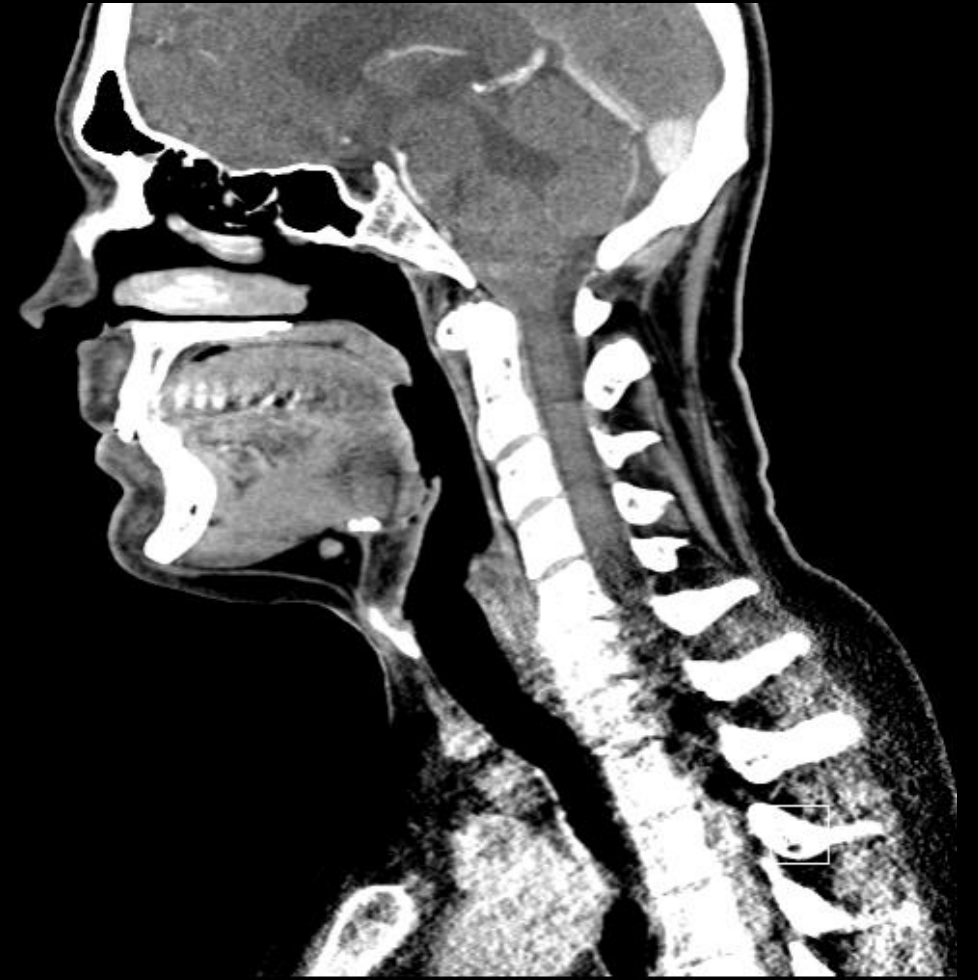
Findings: CT Neck (unlabeled)



Findings: CT Neck (labeled)



Unremarkable neck CT. No masses identified.

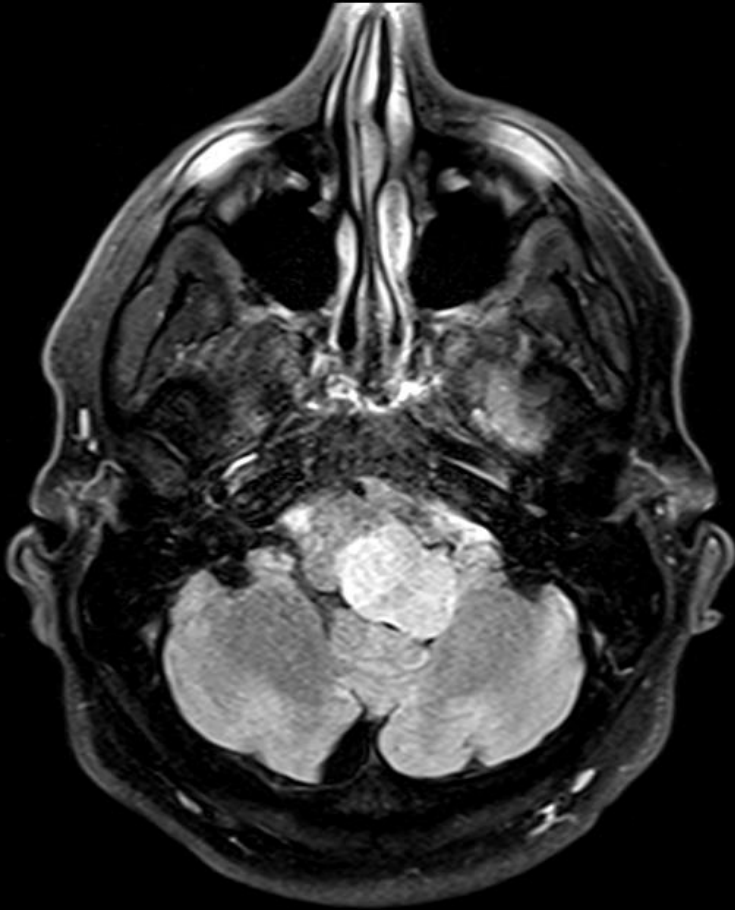


Findings: MRI Head (unlabeled)

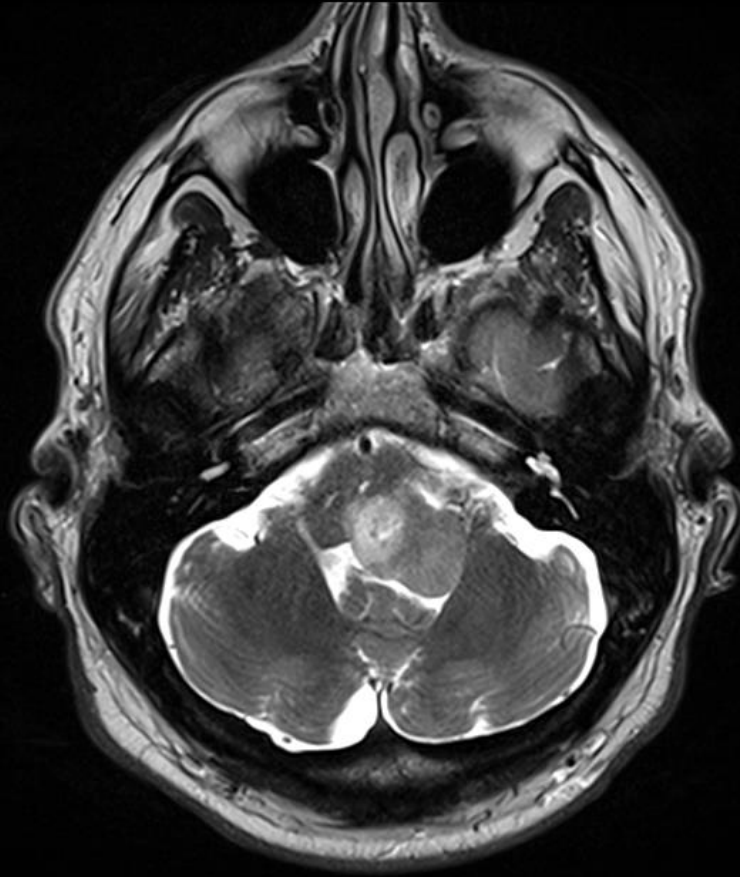


T1 Post-Contrast

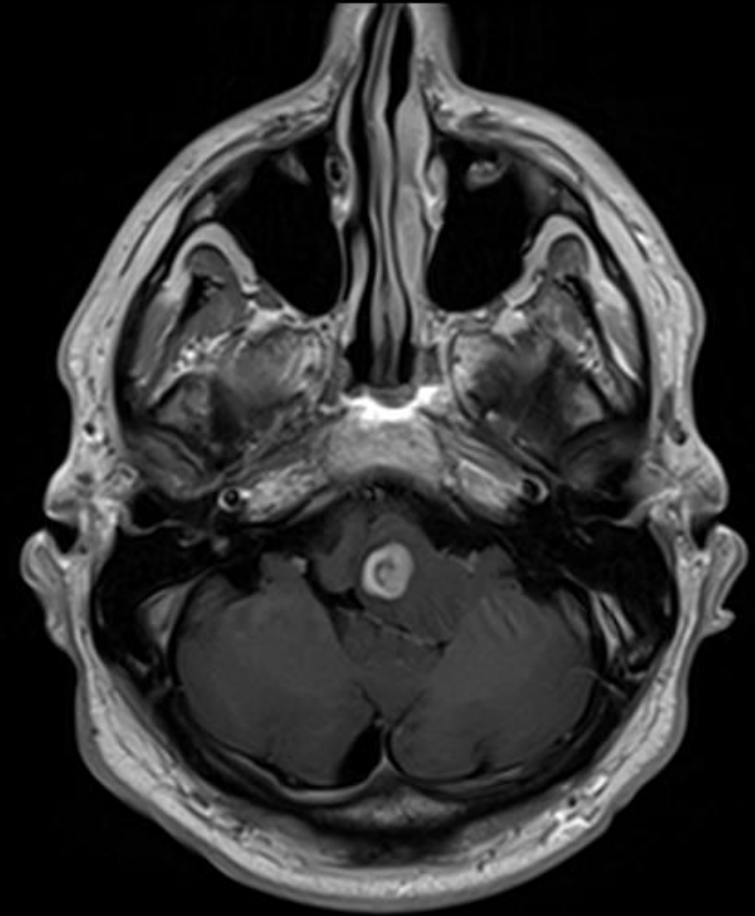
Findings: MRI Head (unlabeled)



T2 FLAIR

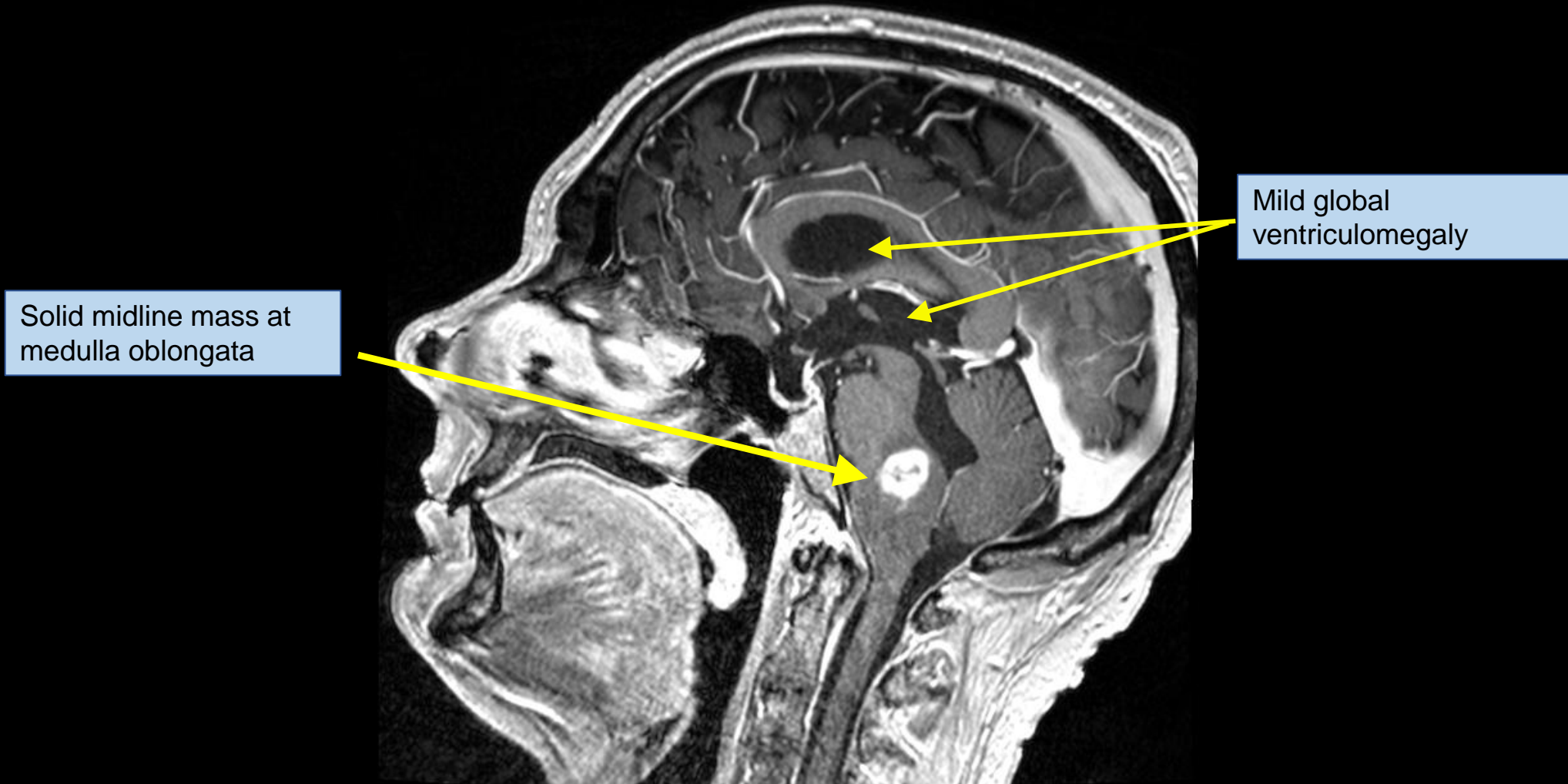


T2



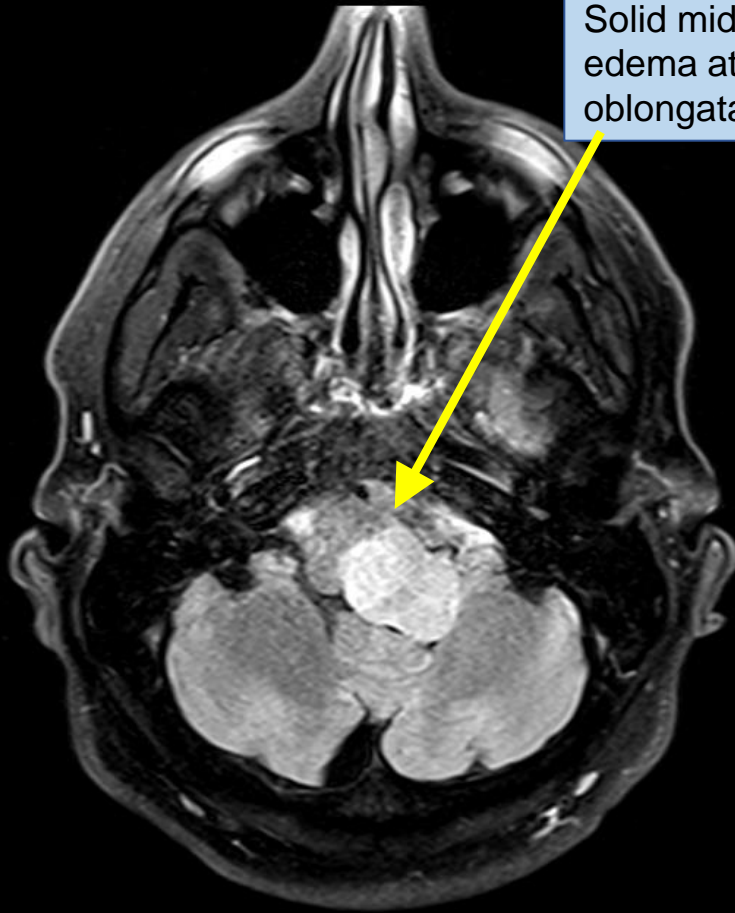
T1 Post-Contrast

Findings: MRI Head (labeled)



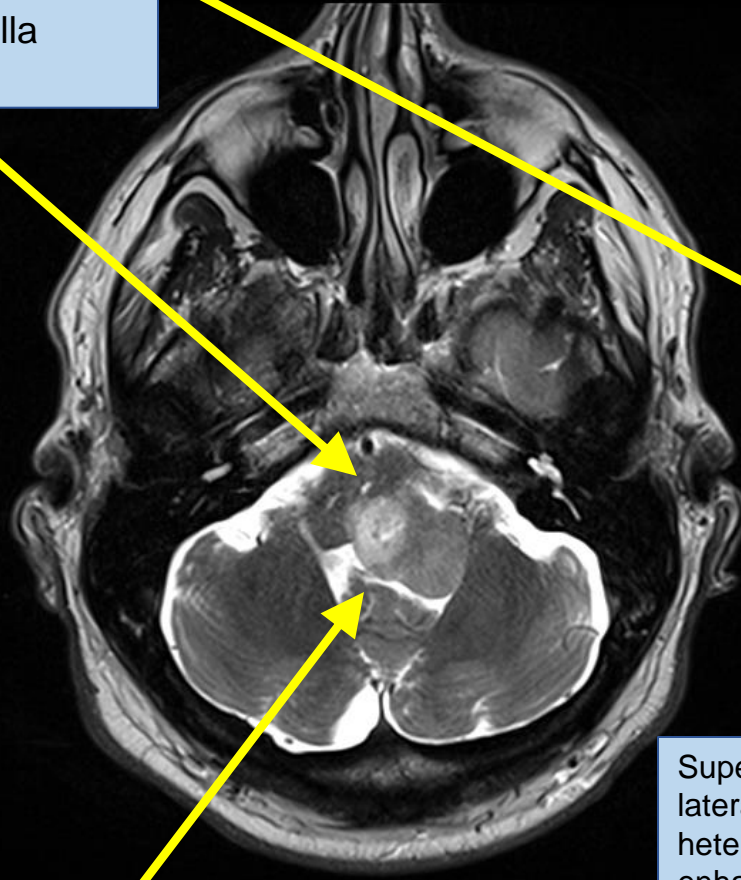
T1 Post-Contrast

Findings: MRI Head (labeled)



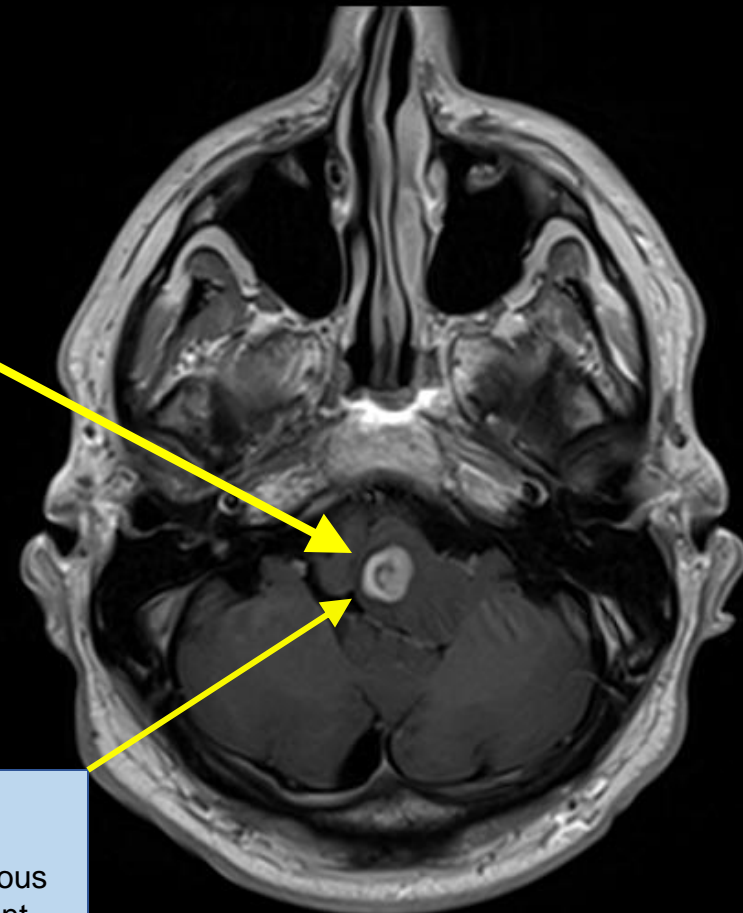
T2 FLAIR

Solid midline mass +
edema at medulla
oblongata



Effacement of
4th ventricle

T2 Post-Contrast



Superior R
lateral
heterogeneous
enhancement

T1 Post-Contrast

Final Dx:

Medullary Diffuse Midline Glioma
WHO Grade IV

Case Discussion – About Gliomas

- The term glioma describes a group of tumors that share features of glial cells (neuron supporting cells) in the brain.
- Gliomas are the most common primary intracranial neoplasm, accounting for 33% of all brain tumors.
- They are most commonly seen in adults, with an increase in incidence with increasing age.
- Only proven risk factor: ionizing radiation
- Gliomas are characterized based on **histological** and **genetic** features.

Case Discussion – Glioma Neuroimaging

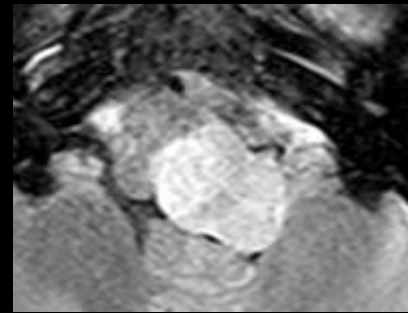
- MRI features

- High Grade Glioma

- T1: Hypointense
 - T1 Post-Contrast: Heterogeneous enhancement
 - T2 and FLAIR: Increased signal intensity
 - MRS: Increased choline, decreased N-acetylaspartate
 - Infiltrative features observed with anaplastic gliomas and glioblastomas: T2 hyperintensity of cortex and white matter.

- Low Grade Glioma

- **Nonenhancing**
 - Best appreciated on T2 or FLAIR



- PET: Increased uptake

- Glioblastoma: Ring enhancing with central necrosis


Case Discussion: Classification of Gliomas

- **Main Histologic Subtypes (WHO 2016):**

- **Diffuse astrocytic and oligodendroglial tumors:**

- **Astrocytoma:** Shares features with astrocytes.

- Anaplastic: Higher cellularity, nuclear atypia.



- **Diffuse midline glioma:** Requires all four criteria- H3 K27M mutant, diffuse, in the midline, a glioma.

- Glioblastoma: 10% of all gliomas, younger adults. Fast growing astrocytoma.

- **Oligodendroglioma:** Shares features with oligodendrocytes.

- **Oligoastrocytoma:** Shares features with oligodendrocytes and astrocytes.

- **Ependymal tumors:** Most common in children. Arises from cells lining cavities of brain and spinal canal.

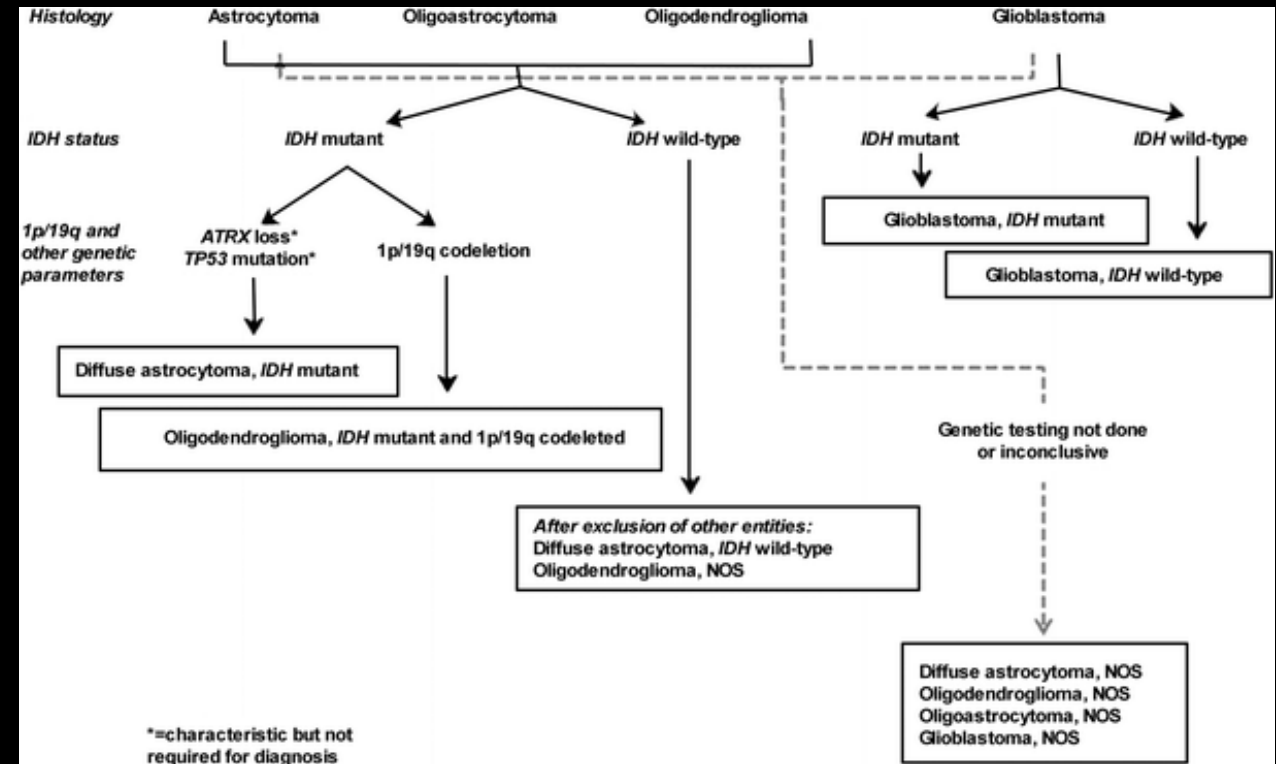
- **Neuronal and Mixed neuronal-glial:** Less common, wide variety of tumors displaying either neuronal or mixed neuronal-glial features.

- **Other gliomas**

- **Other astrocytic tumors**

Case Discussion: Classification of Gliomas

- Gliomas can be further characterized by their genetic/molecular features
- Key molecular diagnostic tests:
 - IDH1/IDH2 mutations
 - 1p/19q codeletion
 - ATRX mutation
 - TP53 mutation
 - BRAF alterations
- Our patient has a **diffuse midline glioma** which is characterized by a **H3-K27M mutation**



Louis et al. (International Agency for Research on Cancer, France, 2016)

- Gliomas can be further classified into WHO grade I – IV, based on the degree of anaplasia. The higher the grade, the more aggressive the tumor.
 - Grades I – II: Low-grade
 - Grades III – IV: High grade
 - Our patient is **WHO grade IV**.

References:

- Chen R, Smith-Cohn M, Cohen AL, Colman H. Glioma Subclassifications and Their Clinical Significance. *Neurotherapeutics*. 2017. doi:10.1007/s13311-017-0519-x
- Louis DN, Ohgaki H, Wiestler OD, Cavenee WK. World Health Organization Histological Classification of Tumours of the Central Nervous System. International Agency for Research on Cancer, France. 2016.
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