

Case Presentation: 32 Year Old Woman Status Post Fall

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History of Present Illness

32 y.o. female presents to the ED 2 days after a fall at home involving a head injury. She states she hit her head on the edge of a cabinet as she fell, without losing consciousness.

Patient endorses recent '**discomfort**' on the **left side of her head and face** (left ear and cheek), which is **worse with loud noises**. She also noted associated **nausea**, though she denied vomiting. She tried Ibuprofen and Tylenol without a relief of the discomfort.

She denies history of visual changes, numbness, tingling, extremity weakness, dysarthria, aphasia, facial droop or substance use.

Additional History

Medical Hx: PCOS, Infertility

Surgical Hx: none

Family Hx: recent TIA in mother associated with vascular malformation

Social Hx: Patient immigrated from India several years ago. She is never smoker, denies alcohol or drug usage.

Examination and Initial Work-Up

147/85 83 97.7F 100%

Physical exam is benign with pertinent negatives below:

Patient is NAD, normocephalic and atraumatic. A&O x 3. No signs of contusion. Normal ear exams bilaterally. PERRL, no visual field deficit, intact EOM, no nystagmus.

Supple neck, full ROM in all extremities. CN II-XII intact, sensation intact, 5/5 strength in all extremities, no signs of focal neurological deficit, negative Romberg, normal finger-to-nose, normal rapid alternating movements, and gait intact. 2+ patellar reflexes bilaterally.

Despite low likelihood of ICH or other significant head injury based on normal exam, she obtained a CT head at an OSH which was unremarkable:

“No acute intra-axial or extra-axial hemorrhage. No intra-axial mass, significant mass effect or midline shift. Cerebral ventricles: Tiny 2mm hyperdensity at the roof of the 3rd ventricle, probably incidental choroid plexus vs. questionable colloid cyst. No hydrocephalus. No skull fractures, No significant sinusitis. Unremarkable soft tissue or mastoid air cells.”

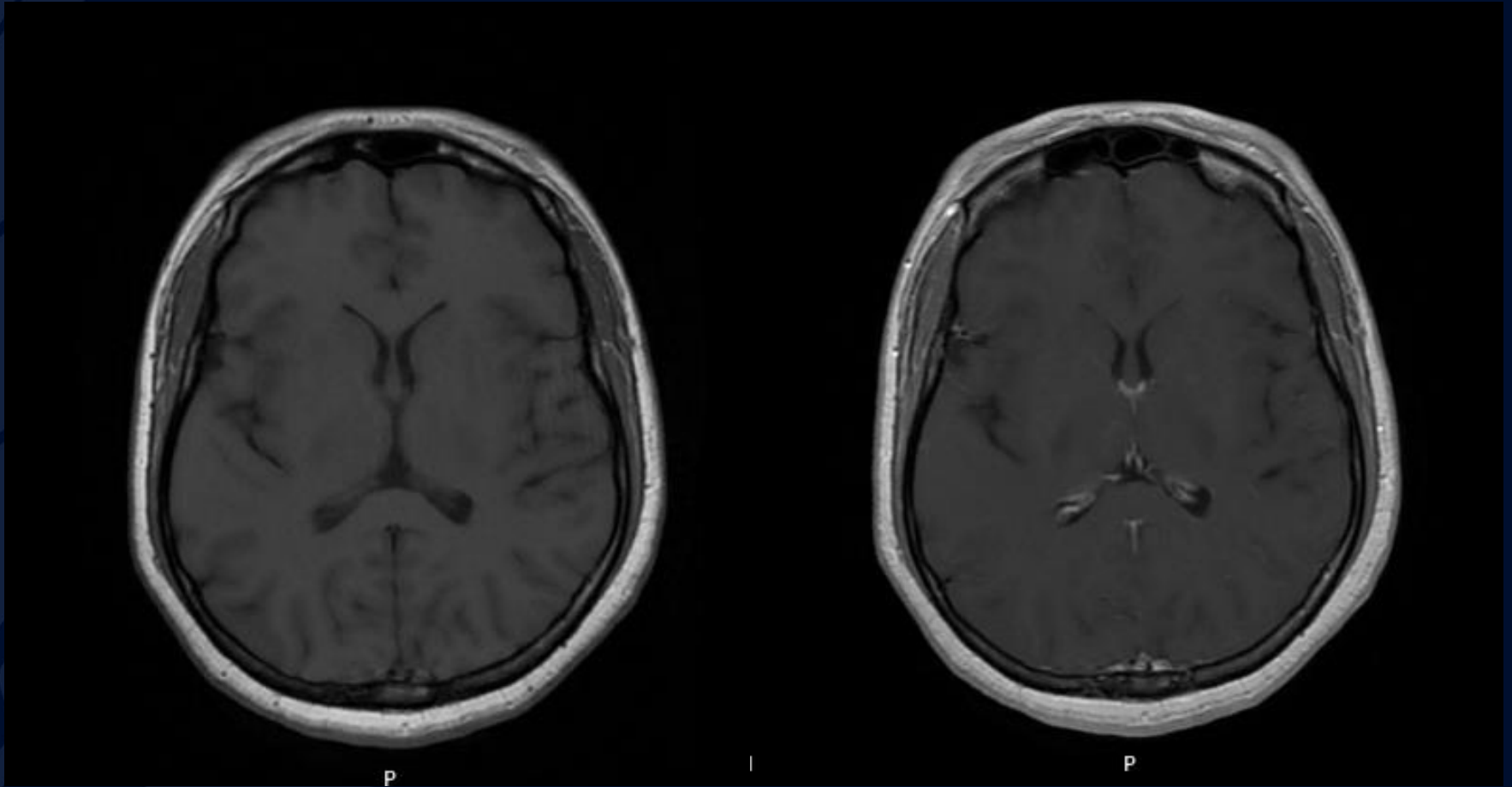
Interim History

Patient was seen by her PCP, who refers her to a neurosurgeon given her persistent symptoms of tinnitus and posterior head and neck pain since the fall incident, which were interfering with her sleep. She also was now reporting *occasional* LUE numbness upon waking.

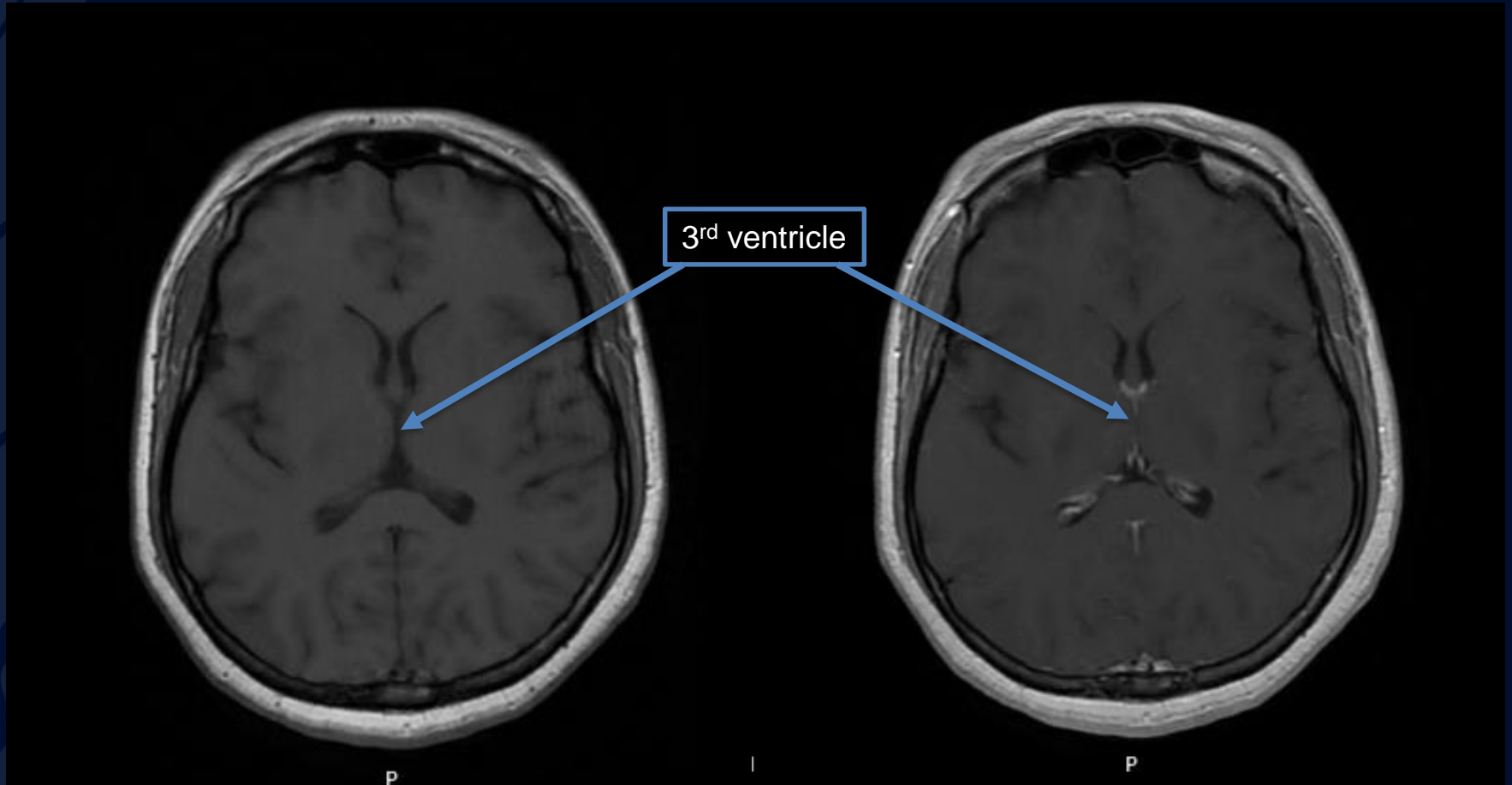
On evaluation by neurosurgery, pertinent positive findings of 3+ DTRs throughout, limited neck extension were noted. Primary diagnosis for the encounter was cyst of brain, but for further evaluation plan was made for:

- MRI Brain w w/o contrast
- Physical therapy

T1 pre and post IV contrast

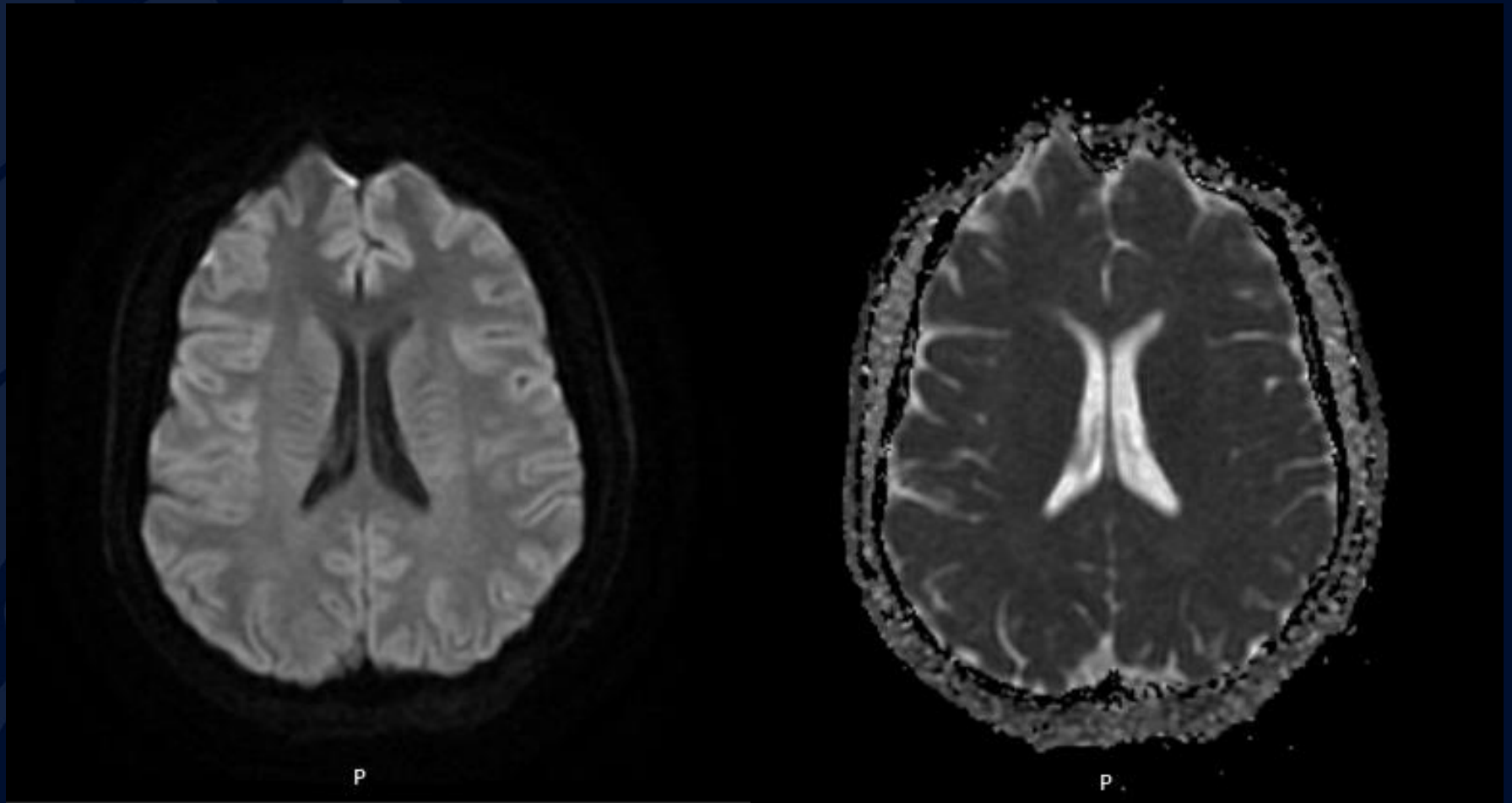


T1 pre and post IV contrast

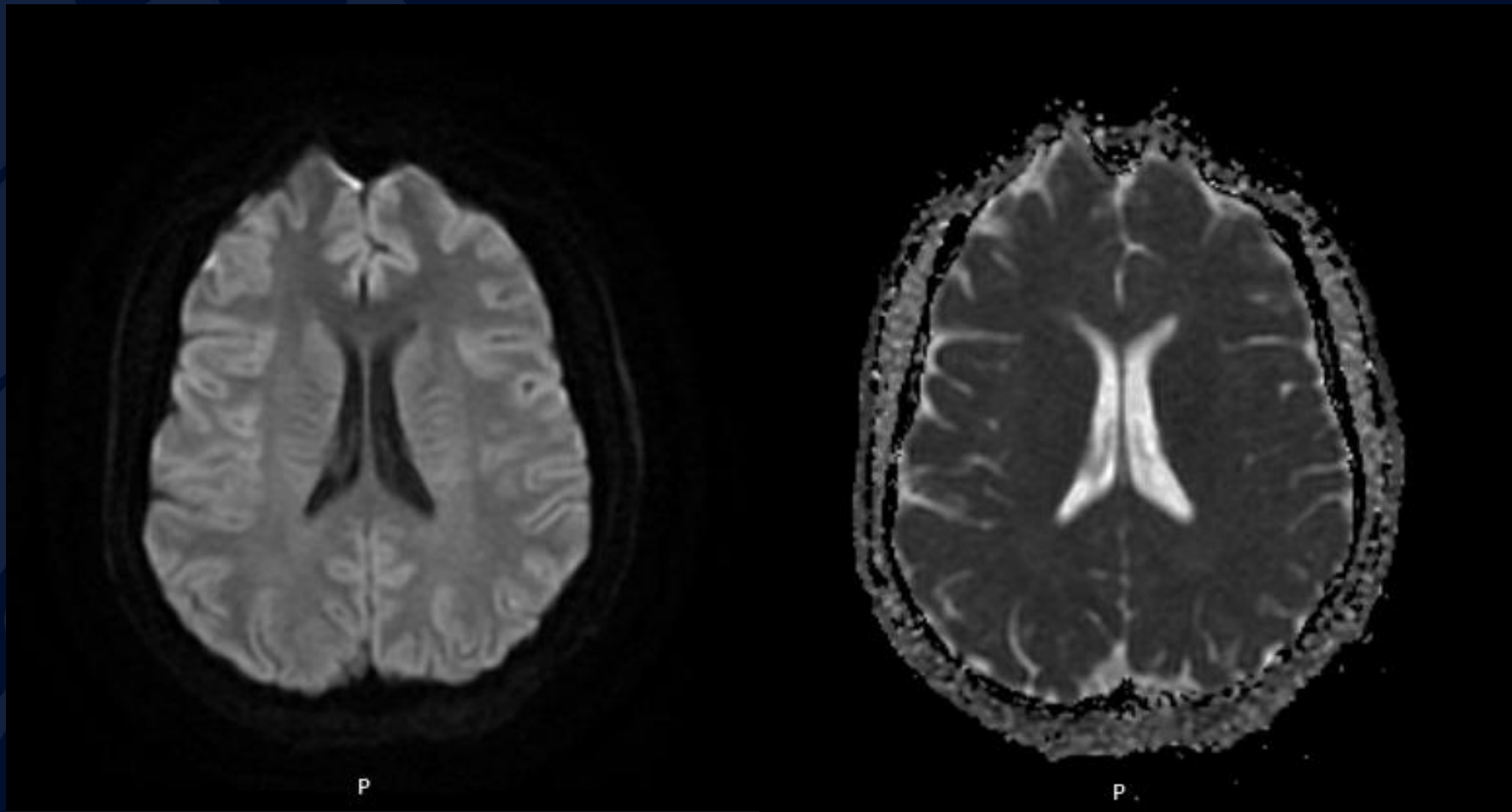


No nodule, signal abnormality, or abnormal enhancement in the 3rd ventricle. A small calcification can remain occult on MRI. No evidence of recent or remote hemorrhage in the parenchyma. No mass lesion or parenchymal enhancement noted.

DWI and ADC

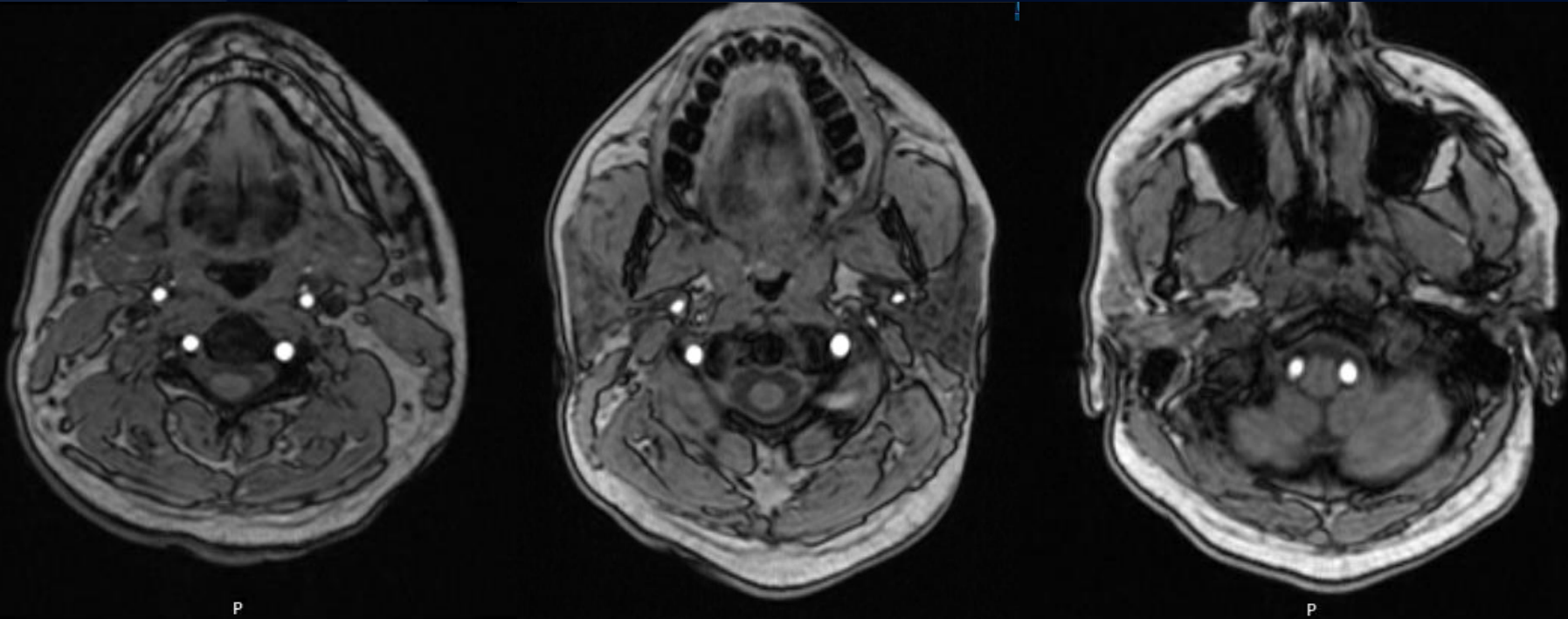


DWI and ADC

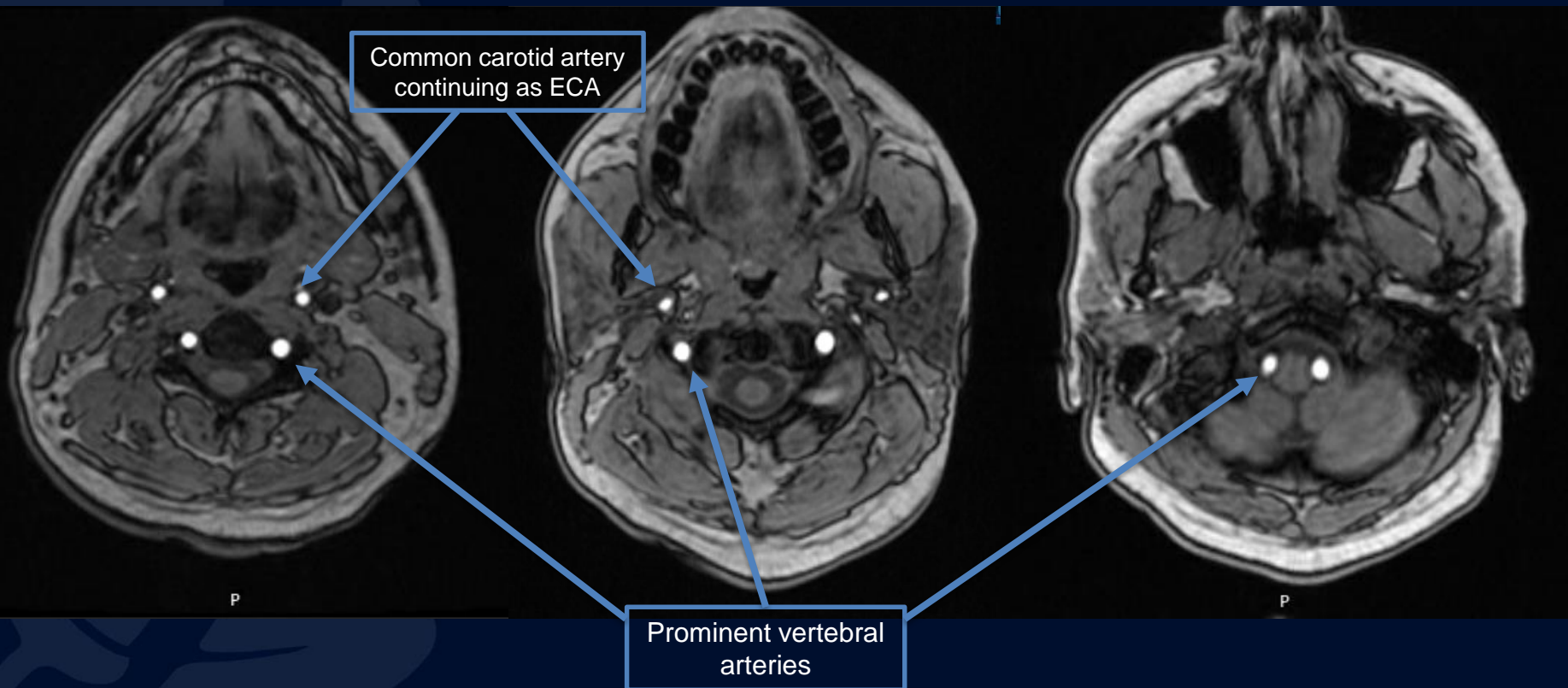


No focal areas of restricted diffusion indicating acute changes.

MRA of neck



MRA of neck

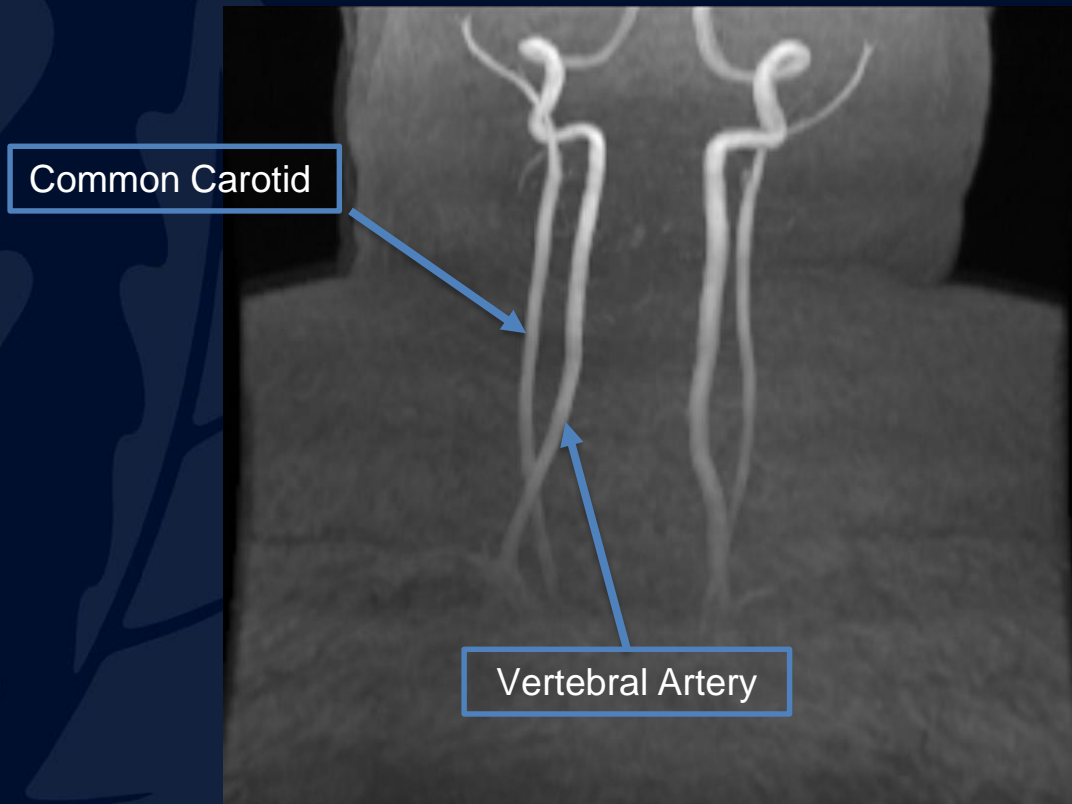


The common carotid arteries do not bifurcate on either side, continuing as the external carotid arteries in the upper neck. There is a co-dominant vertebral system.

MRA Neck 3D MIP

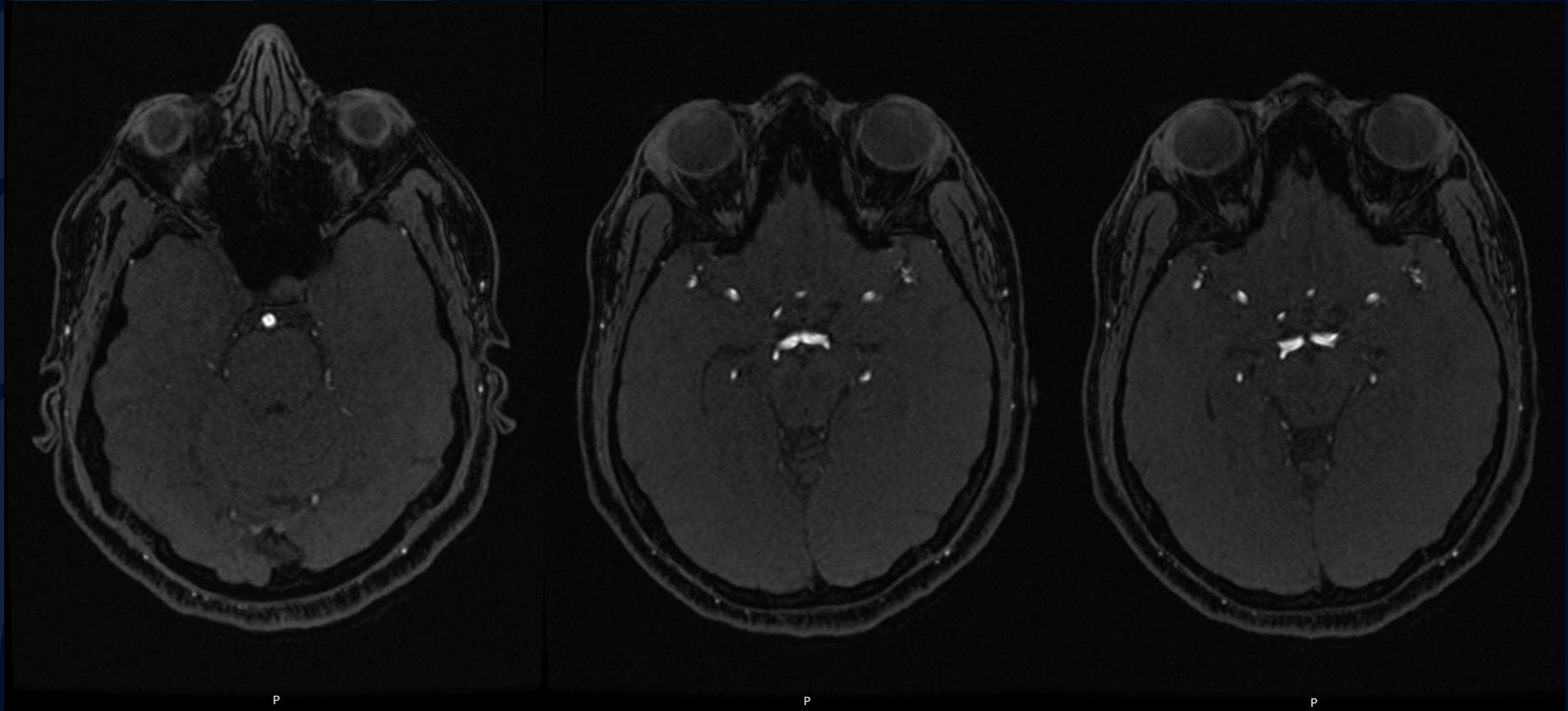


MRA Neck 3D MIP

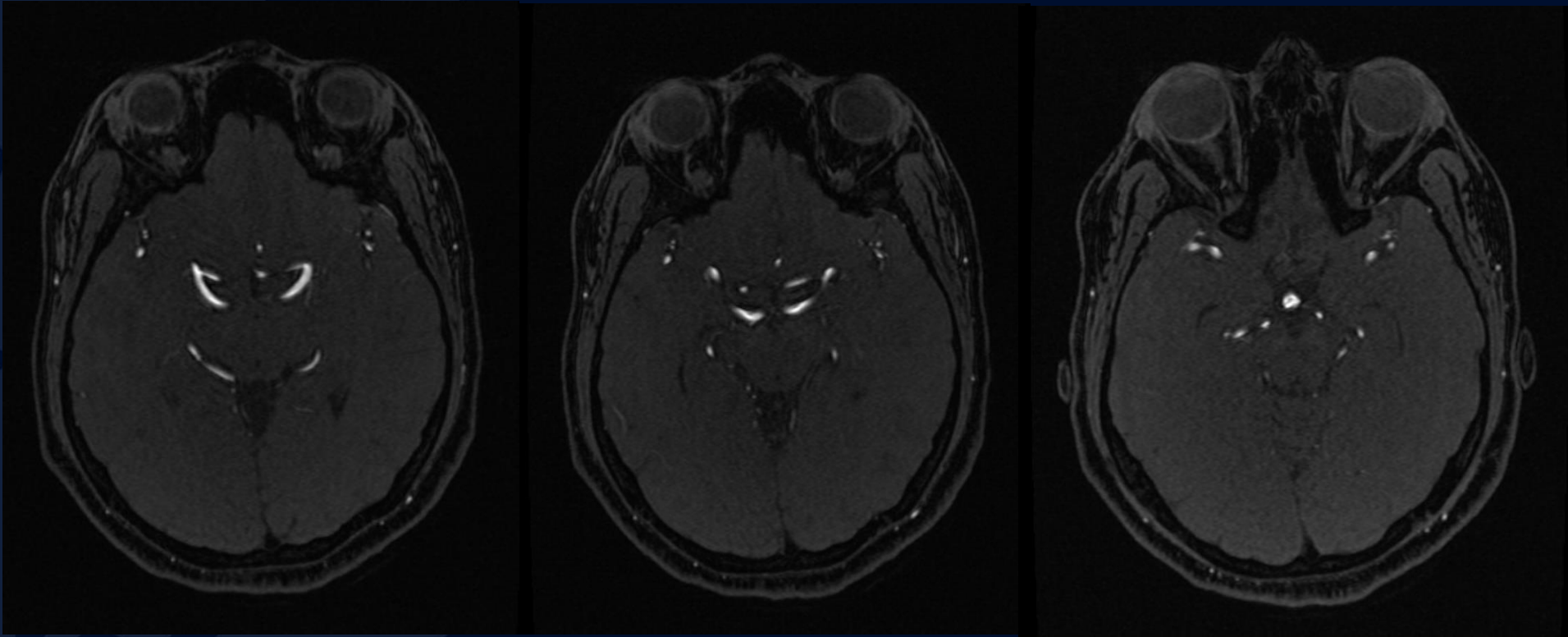


The common carotid arteries do not bifurcate either side, continuing as the external carotid arteries in the upper neck. No evidence of aneurysm or focal stenosis.

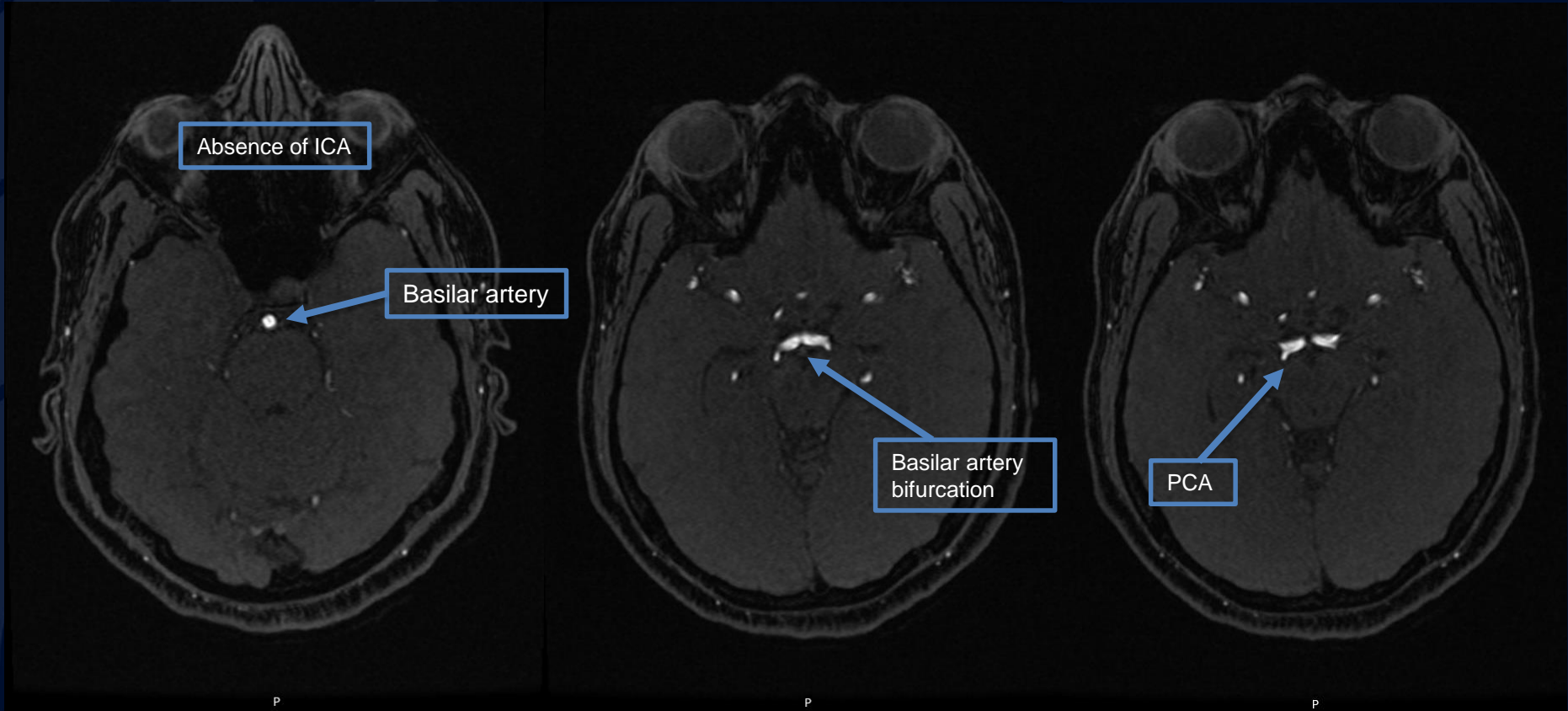
MRA of Head Axial Sequence



MRA of Head Axial Sequence 2

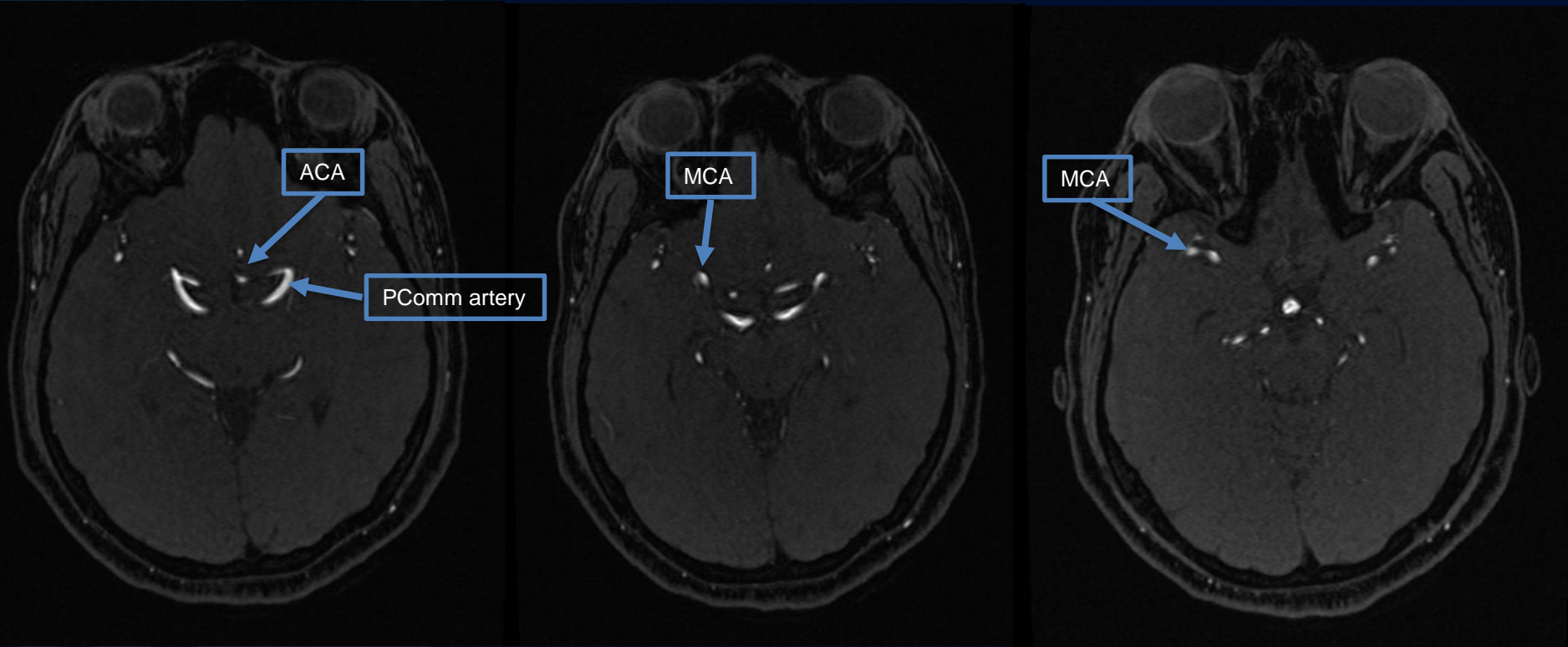


MRA of Head Axial Sequence



The internal carotid arteries are congenitally absent.

MRA of Head Axial Sequence 2



A robust vertebrobasilar system with robust posterior communicating arteries supply the anterior circulation as well as the posterior circulation. No diffuse luminal irregularity is seen within the small intracranial vessels.

MRA Head 3D

Rotation: 9.0 deg.



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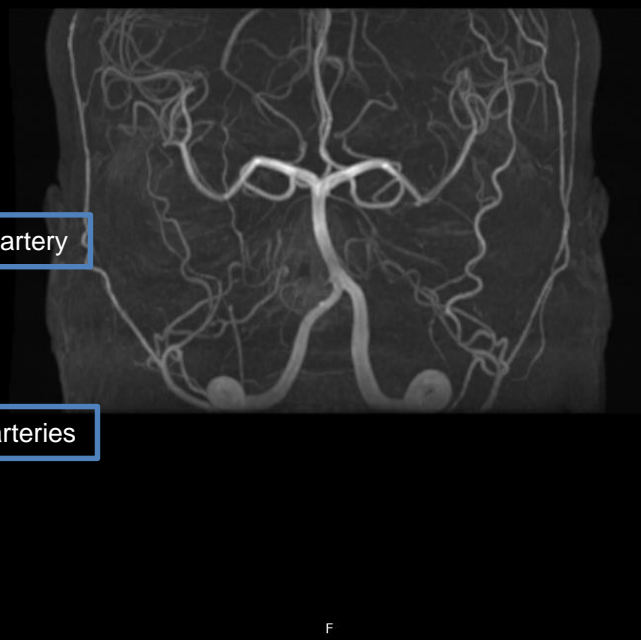
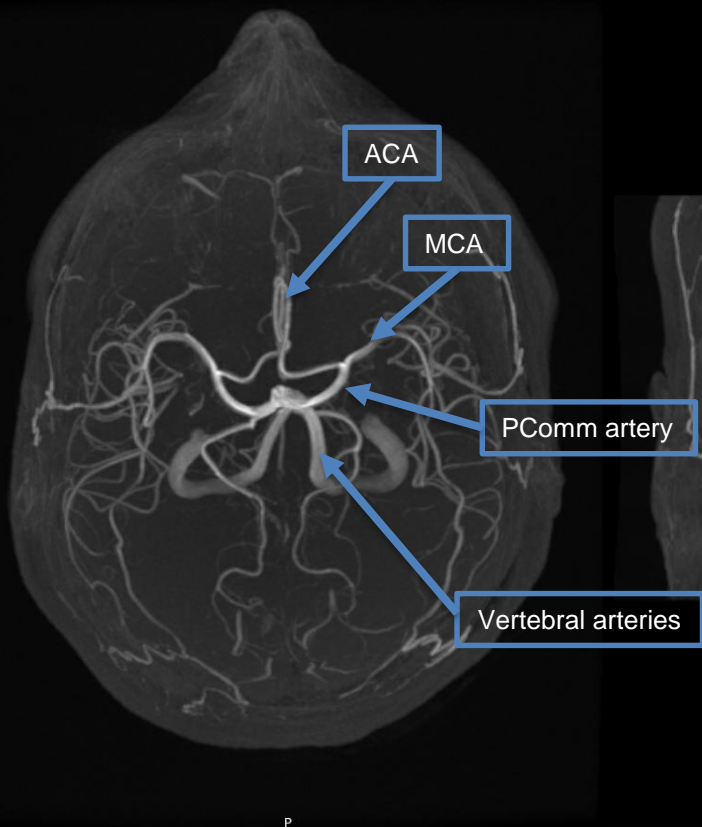
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MRA Head 3D

Rotation: 9.0 deg.

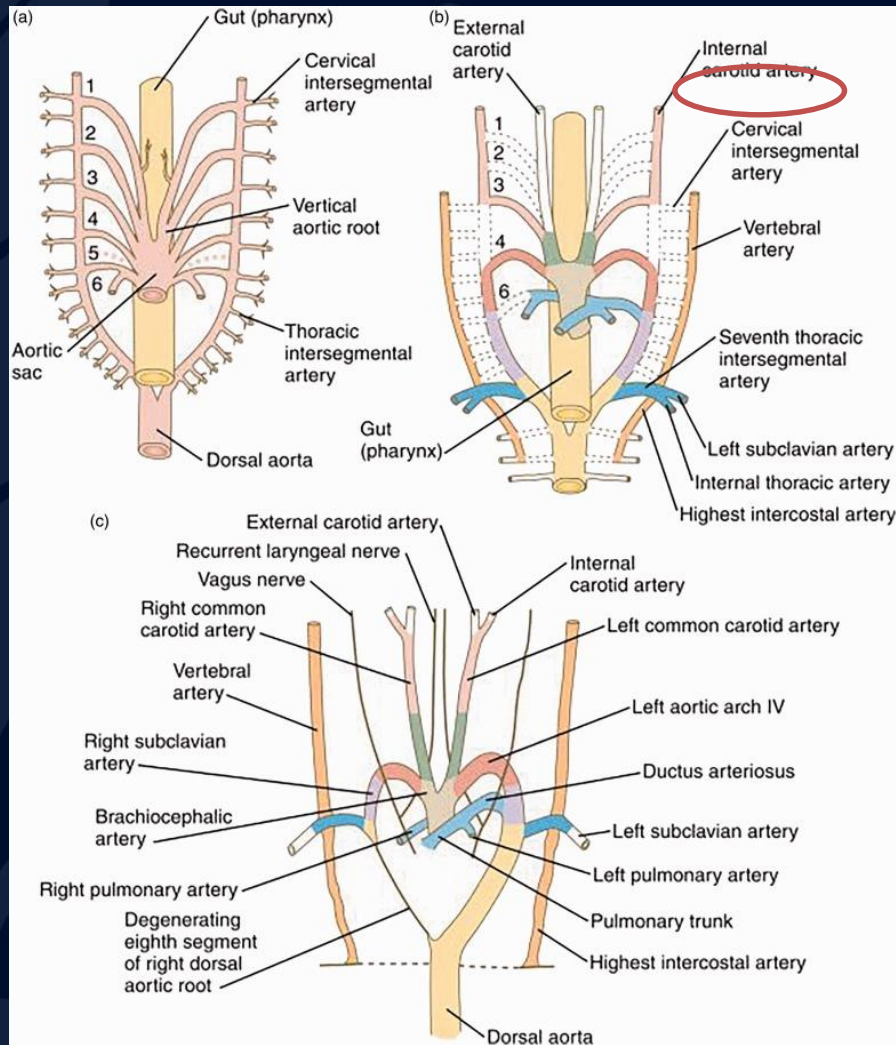


Differential Diagnosis?

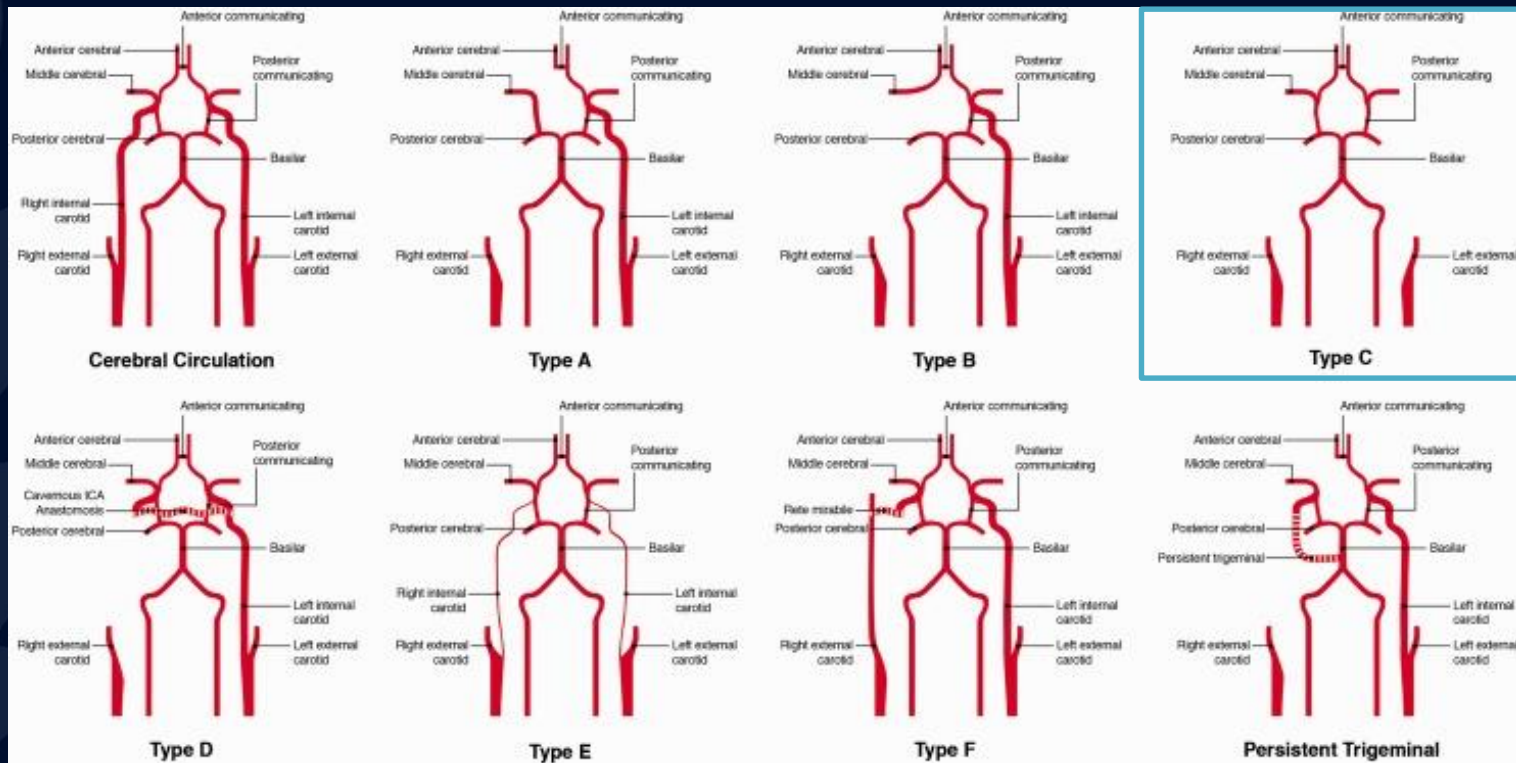
- Primary headaches especially migraine considering her phonophobia, nausea and unilateral headache
- Third ventricle cyst
 - Considered given the reported finding of a possible cyst in the third ventricle on CT head
 - Colloid cyst in third ventricle can suddenly impede the flow of cerebrospinal fluid, causing obstructive hydrocephalus. Majority of patients also experience an acute onset headache (which can be in the occipital area as in our patient) with resolution, lasting from seconds to a day. About one third of patients have associated nausea and vomiting.
- Vascular
 - Considered given family history of TIA in mother associated with a vascular malformation.
 - These may include developmental anomalies of arteries, veins, and capillaries.
 - Aneurysm can also cause headache.
 - Reversible cerebral vasoconstriction syndrome (RCVS)
 - Cerebral Venous Thrombosis
- Neoplasm
 - Acoustic neuroma causing progressive tinnitus, left facial discomfort and headache. Can cause a fall with cerebellar involvement, though her cerebellar exams are normal.
- Meningeal irritation from infectious or inflammatory process given her headache and neck stiffness, though less likely.

Bilateral Congenital Internal Carotid Artery Agenesis

Congenital Bilateral ICA Agenesis



- Rare, affecting less than 0.01% of the population
Currently around 200 reported cases in the literature.
- Intracranial circulation is maintained by collateral circulation from the vertebrobasilar system through the posterior communicating artery
- Most of the time patients are asymptomatic due to the collaterals, but patients can present with headache, seizures, or TIA.
- Aneurysms are also reported to be associated with ICA agenesis.
- ICA agenesis occurs due to abnormal regression of the 1st and 3rd aortic arch, with unclear etiology.



Li, Shuo & Hooda, Kusum & Gupta, Nishant & Kumar, Yogesh. (2017). Internal carotid artery agenesis: A case report and review of literature. The Neuroradiology Journal. 30. 197140091769216. 10.1177/1971400917692162.

- Several types of circulation to the ACA and MCA on the side of carotid agenesis have been described. Three categories of collateral circulation include flow through the circle of Willis, persistent fetal circulation, and through skull base arteries from the ECA.
- Our patient belongs to Type C circulation, where there is bilateral carotid artery agenesis with supplies to the ACAs and MCAs through the posterior circulation via the Posterior Communicating arteries.

Management

- Given the asymptomatic and congenital nature of carotid agenesis, there is no guideline for the management of internal carotid artery agenesis.
- However, given its association with cerebral aneurysms, screening for aneurysm using non-ionizing imaging modalities like MR angiogram has been suggested in patients with ICA agenesis.

Sources

1. Li S, Hooda K, Gupta N, Kumar Y. Internal carotid artery agenesis: A case report and review of literature. *Neuroradiol J*. 2017;30(2):186-191. doi:10.1177/1971400917692162
2. Oz II, Serifoglu I, Yazgan O, Erdem Z. Congenital absence of internal carotid artery with intercavernous anastomosis: Case report and systematic review of the literature. *Interv Neuroradiol*. 2016 Aug;22(4):473-80. doi: 10.1177/1591019916641317.
3. Schwedt T, Swanson J et al. Overview of Thunderclap Headache. Uptodate. 2021. https://www.uptodate.com/contents/overview-of-thunderclap-headache?search=3rd%20ventricle%20cyst&source=search_result&selectedTitle=1~150&usage_type=default&display_rank=1