

AMSER Case of the Month

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74 year-old female with headache, blurry vision, and
right CN III palsy

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Patient Presentation

- **HPI:** 74yoF with PMHx of HTN, HLD, and DM2 presented with 10-day of progressive R-sided headache, drooping R eyelid, and blurry vision. Patient reported abrupt onset of blurry vision upon waking up and foreign body sensation in R eye since onset of symptoms. She denied double vision, trauma, provoking factors, history of stroke or seizures, focal weakness, or altered sensorium.
- **PE:** Normal vitals. Neuro exam is pertinent for asymmetric but equally reactive pupils, R mydriasis, anisocoria ($R > L$ eye), impaired extra-ocular moment in both eyes (L eye with reduced vertical upgaze, not able to abduct; R eye with intact abduction and inferomedial gaze but restricted in other directions); R ptosis; R eye at rest is “down and out”. The rest of the neuro exam and mental exam is unremarkable.

Pertinent Labs

- **CMP:** Glu 153 (H), Na 138, K 3 (L)
- **CBC:** WBC 8.3, Hb 15, Pt 299
- Troponin 7, A1C 6.5, TSH 1.35, T4 1.52
- **Coags:** INR 1.1, PT 11.9, aPTT 33
- **Inflammatory labs:** ESR 12, CPR 0.37, ACE 82
- **Infectious labs:** negative for HIV, syphilis, TB, *Borrelia burdorferi*

What Imaging Should We Order?

Select the applicable ACR Appropriateness Criteria

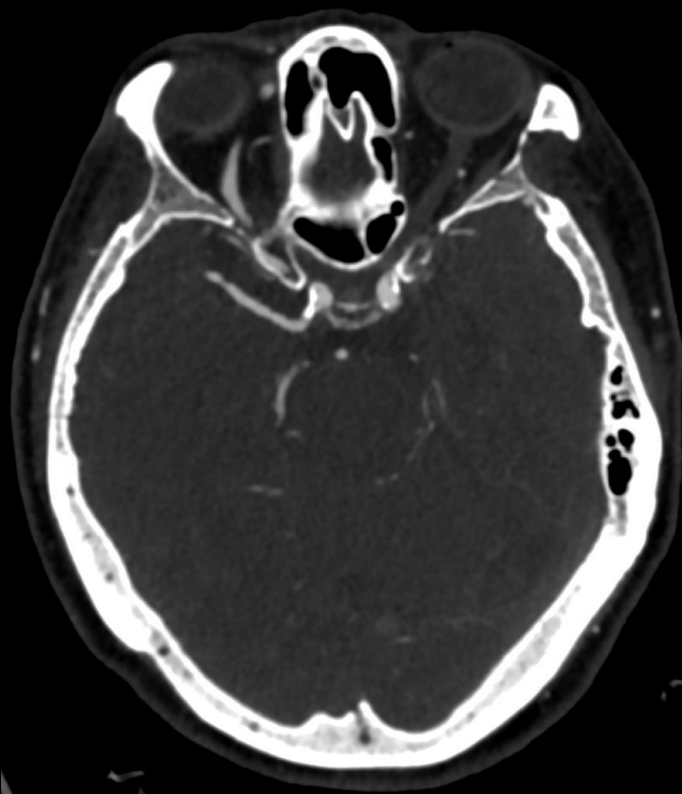
Variant 1:

Ophthalmoplegia or diplopia. Initial imaging.

Procedure	Appropriateness Category	RRL
MRI head without and with IV contrast	Usually Appropriate	○
MRI orbits without and with IV contrast	Usually Appropriate	○
CT orbits with IV contrast	Usually Appropriate	⊗⊗⊗
MRI orbits without 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	○
MRA head and neck without IV contrast	May Be Appropriate	○
MRI head without IV contrast	May Be Appropriate	○
CT head with IV contrast	May Be Appropriate	⊗⊗⊗
CT head without IV contrast	May Be Appropriate	⊗⊗⊗
Arteriography cervicocerebral	Usually Not 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	⊗

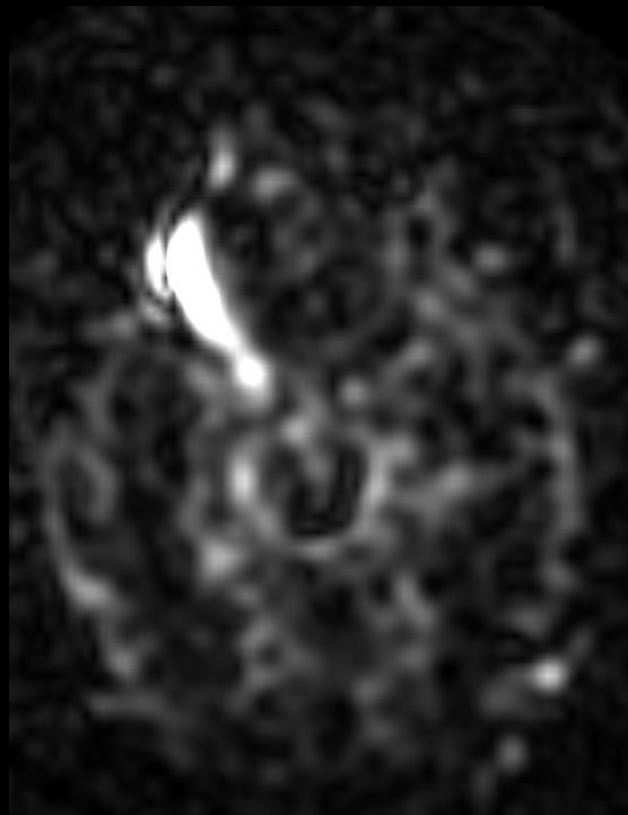
These imaging modalities were ordered by the ER physician

Findings: CTA (unlabeled)

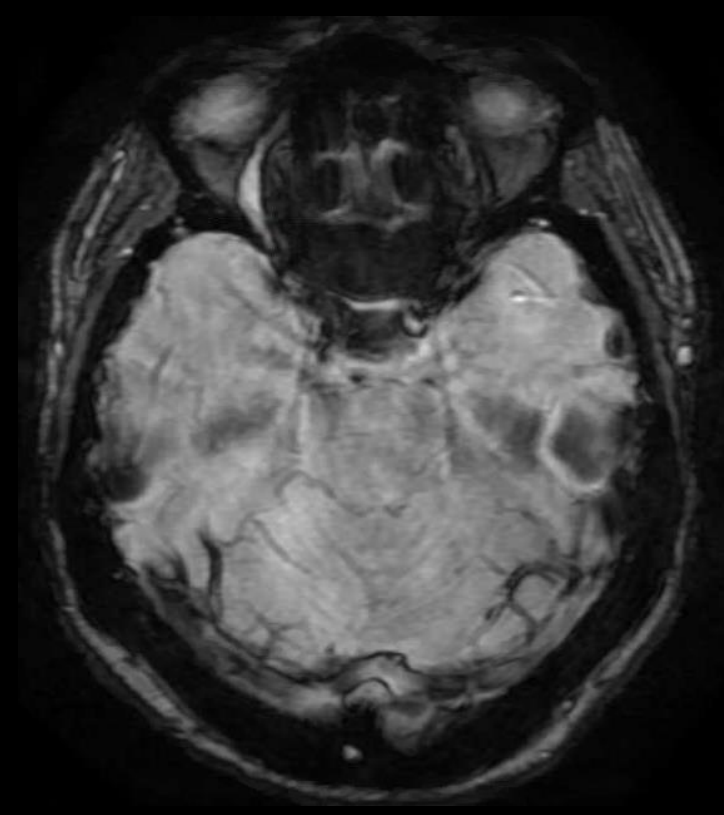
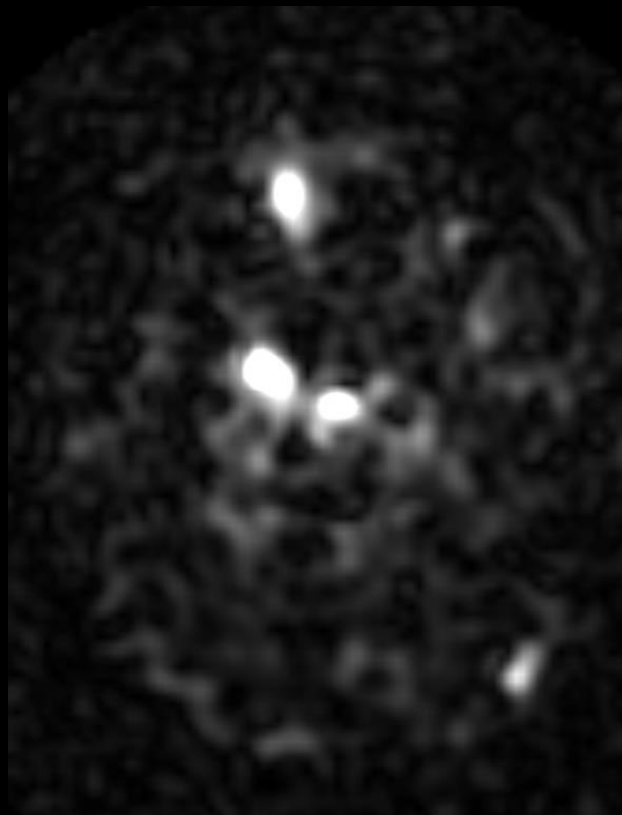


Findings: MRI (unlabeled)

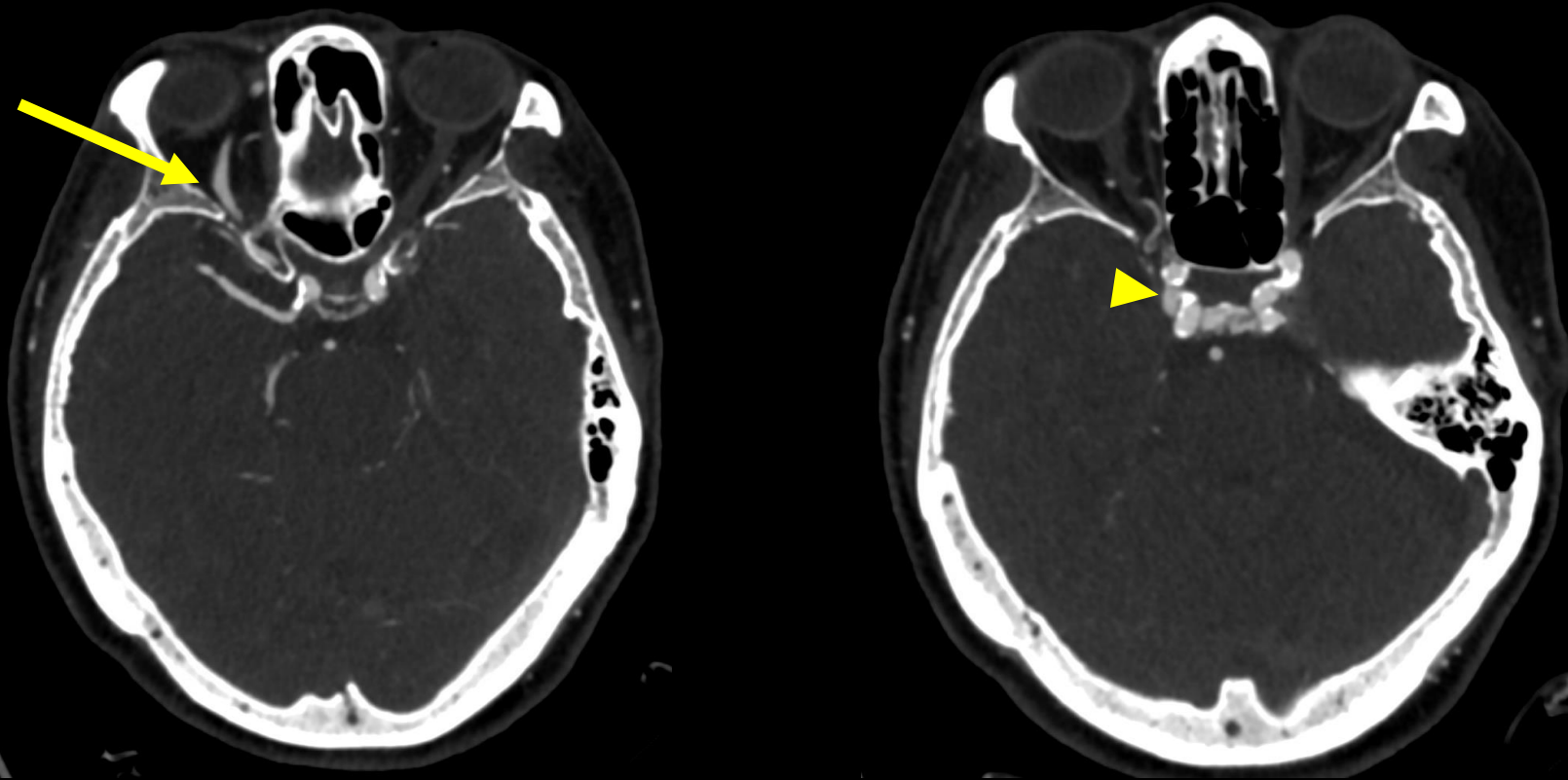
ASL



SWAN



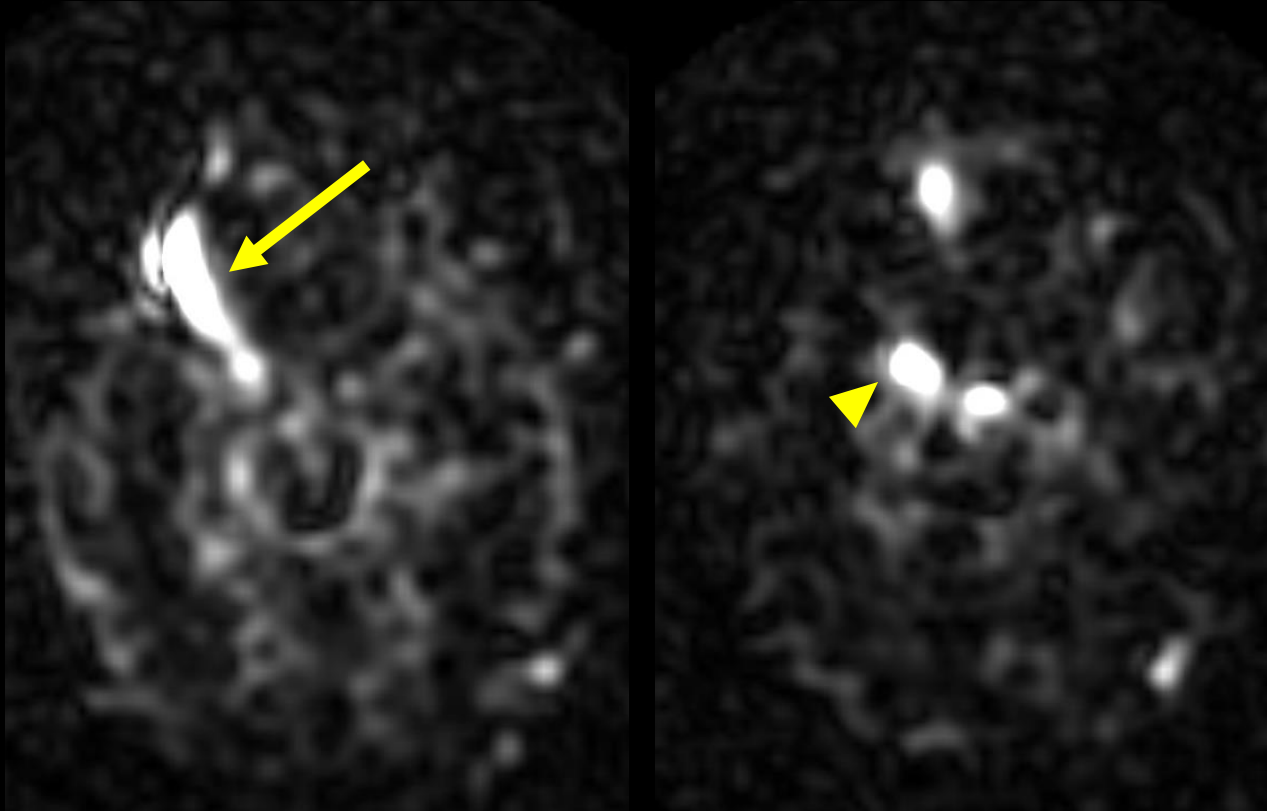
Findings: CTA (labeled)



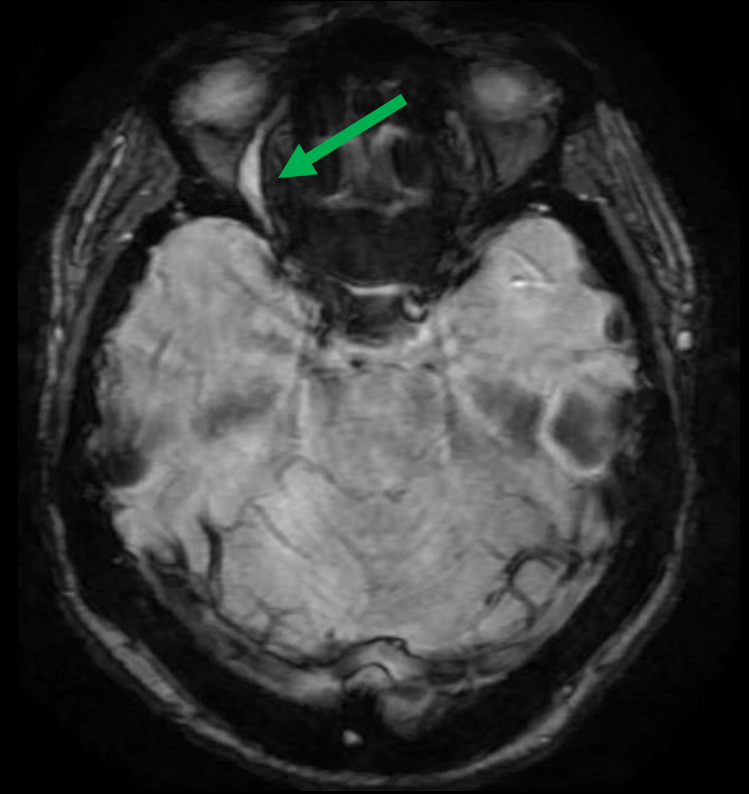
CTA shows abnormal arterial enhancement of the **right superior ophthalmic vein (arrow)** and **right cavernous sinus (arrowhead)**.

Findings: MRI (labeled)

ASL



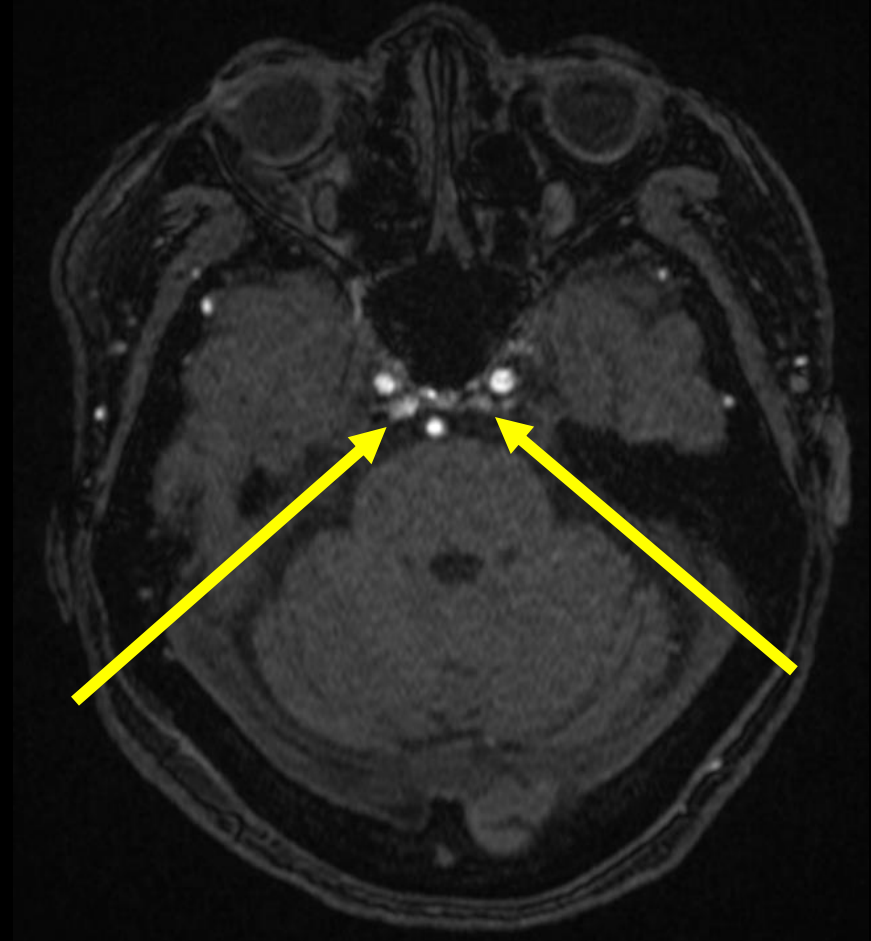
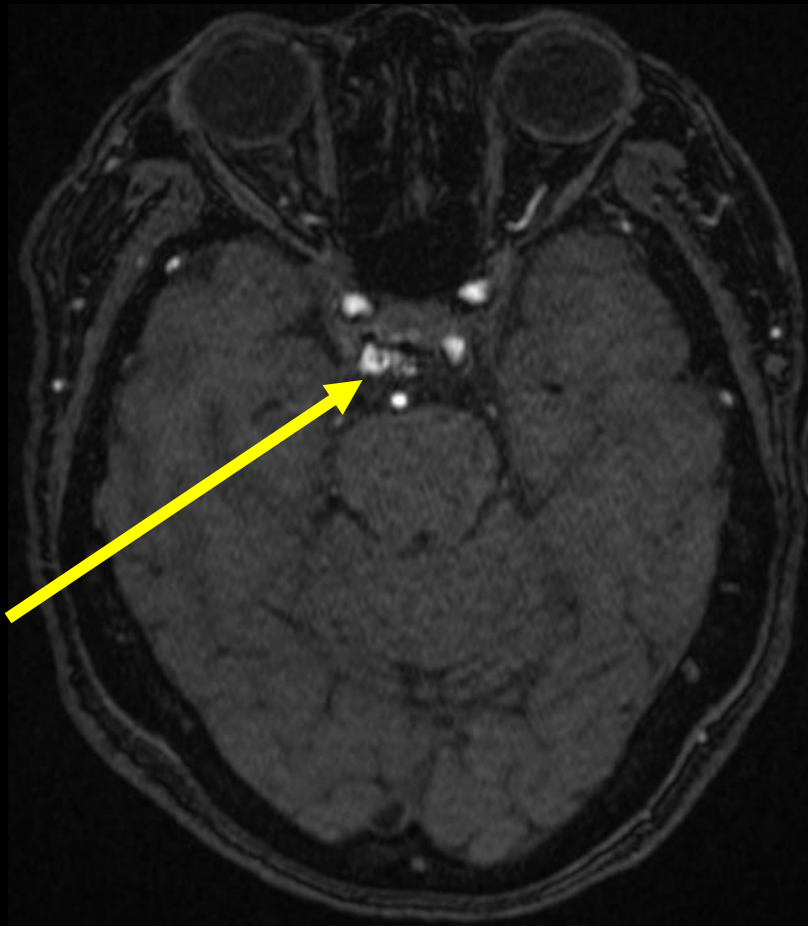
SWAN



ASL sequence shows elevated signal within **right superior ophthalmic vein (arrow)** and **right cavernous sinus (arrowhead)**, suggesting arteriovenous shunting.

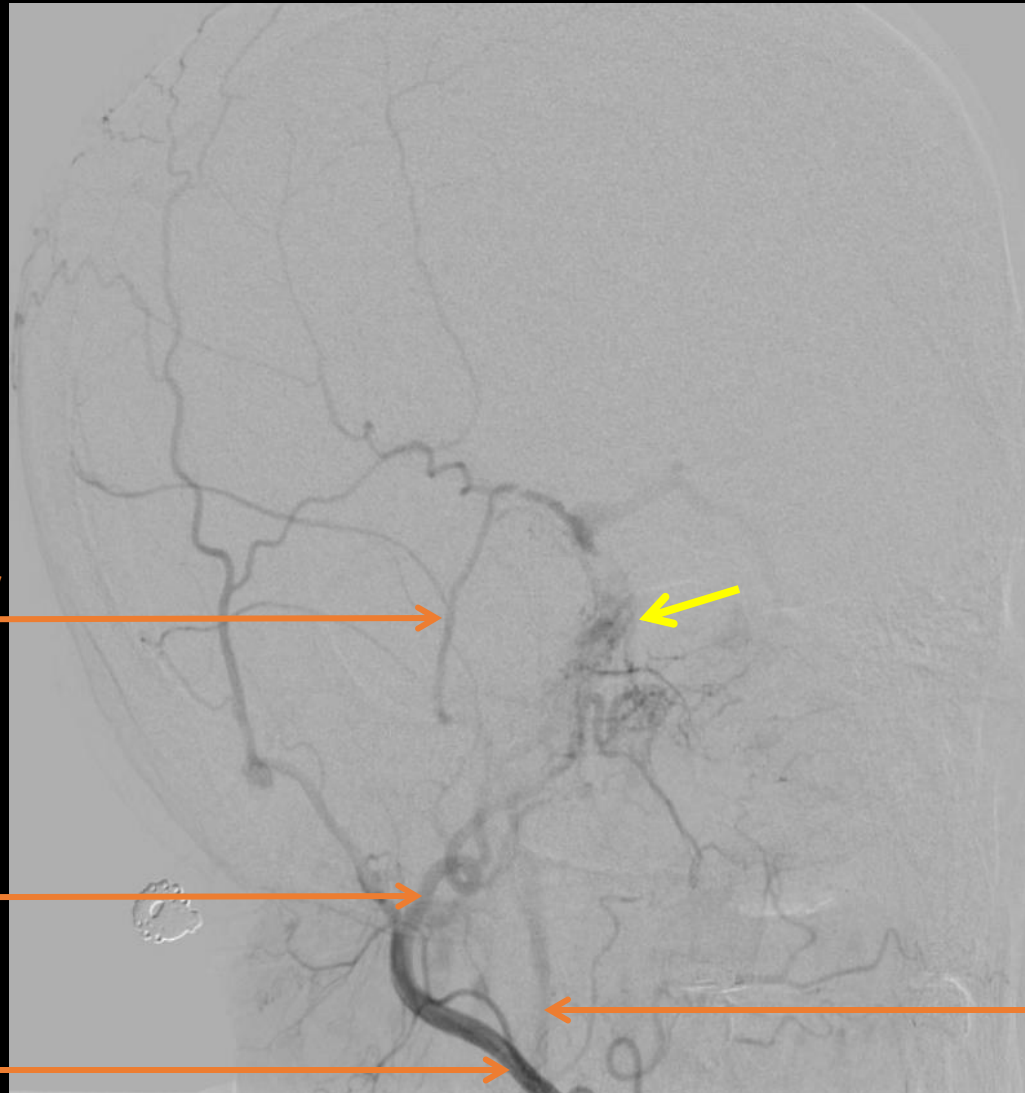
SWAN sequence shows corresponding high signal in the right superior ophthalmic vein (**green arrow**), suggesting elevated oxyhemoglobin in the setting of shunting.

Follow-up MRA



MRA also shows **abnormal flow-related signal** in the **right-greater-than-left cavernous sinuses**, providing additional evidence of arteriovenous shunting.

Follow-up diagnostic cerebral angiogram



Angiogram of the **right** extracranial circulation showing:

- **Barrow type C carotid cavernous fistula (arrow)** supplied primarily by the right internal maxillary artery and middle meningeal artery

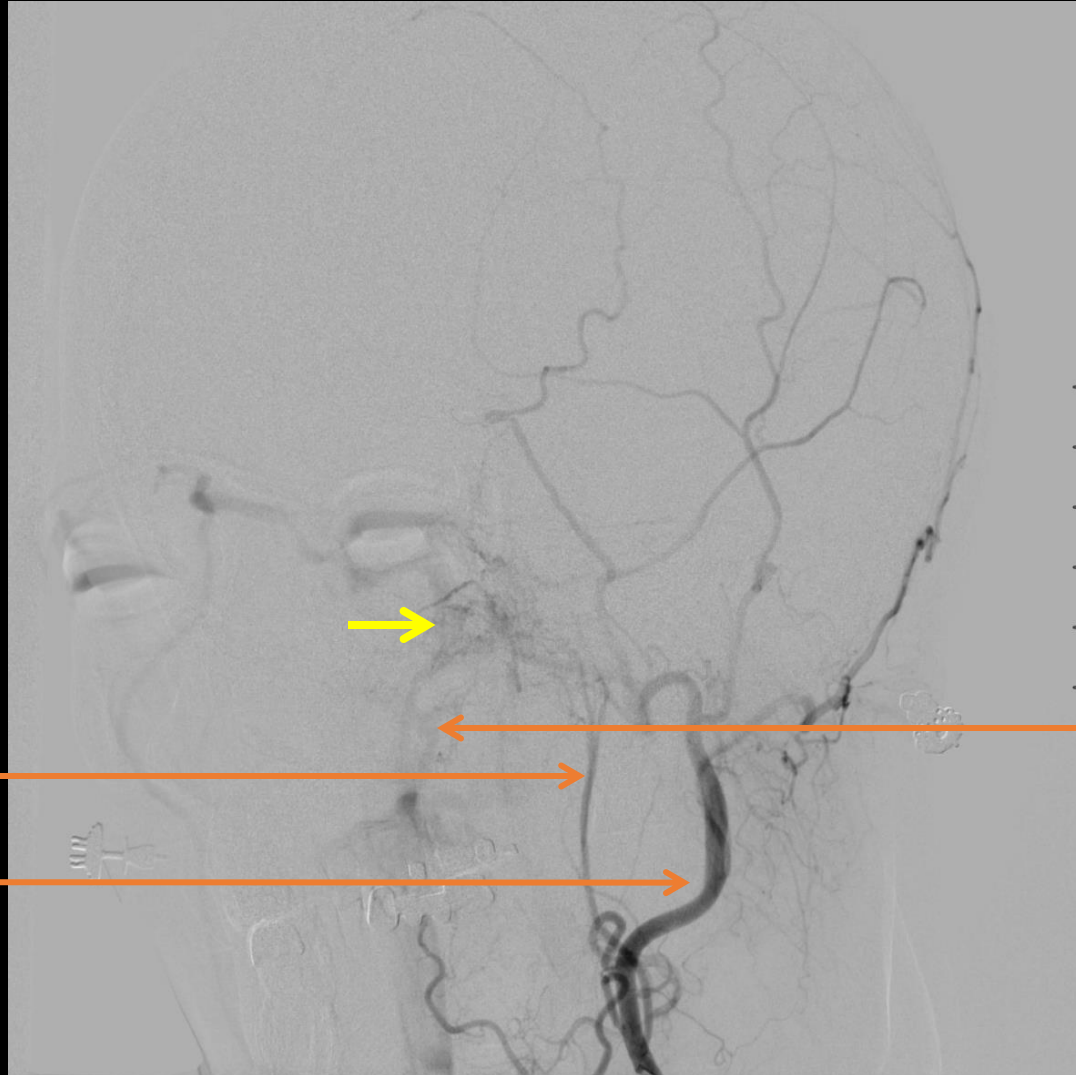
Middle meningeal artery

Internal maxillary artery

External carotid artery

Early venous drainage into the right inferior petrosal sinus

Follow-up diagnostic cerebral angiogram



Angiogram of the **left** extracranial circulation showing:

- **Barrow type C carotid cavernous fistula (arrow)** supplied primarily by the left ascending pharyngeal artery

Ascending pharyngeal artery

External carotid artery

Early venous drainage into the left inferior petrosal sinus

Final Dx:

Bilateral carotid-cavernous fistulas (CCF)

Case Discussion

- CCF is formed by abnormal connection between the internal and/or external carotid arteries and cavernous sinus ¹
- Symptoms ^{1,2}: depending on involvement of CN III, IV, V1, V2, and VI, can cause pulsatile exophthalmos, chemosis, tinnitus, proptosis, restricted ocular mobility, diplopia, elevated intraocular pressure, and secondary glaucoma
- Etiologies ¹:
 - Direct connection between the intracavernous internal carotid artery and cavernous sinus (Direct; Barrow Type A): caused by trauma, ruptured aneurysm, or connective tissue disorder
 - Dural shunt between intracavernous branches of the internal carotid and/or meningeal branches of the external carotid artery and cavernous sinus (Indirect; Barrow Types B, C, D): related to cavernous sinus thrombosis with subsequent revascularization (similar to dural arteriovenous fistula)
 - CCF diagnosis is often delayed due to nonspecific, insidious symptoms, with an average diagnostic delay of about 7 months⁴

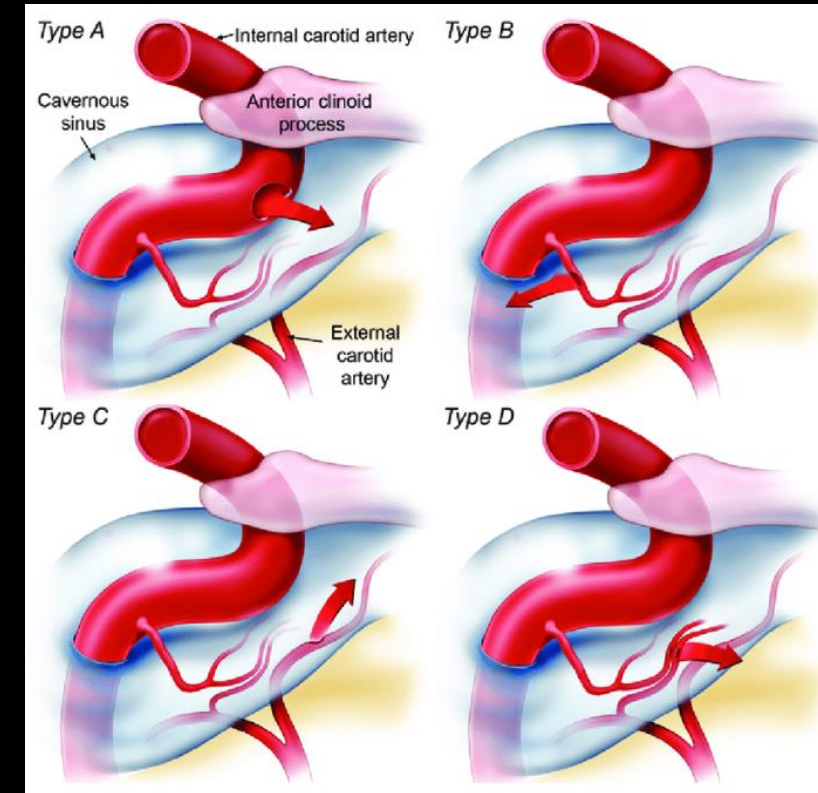


Image adaptive from *Ellis et al.* ³ **The Barrow classification of CCF:** Type A: direct fistula, Type B,C,D: indirect from ICA (B), from ECA (C), from both ICA & ECA (D)

Case Discussion

- Imaging – crucial for CCF diagnosis ^{1,3}:
 - Cerebral angiography: gold-standard for diagnosis of CCF
 - CTA, MRA: usually obtained as initial imaging, common findings includes early enhancement/abnormal flow-related signal involving cavernous sinus and superior ophthalmic vein, proptosis, extraocular muscle enlargement, superior ophthalmic vein dilation
 - MRI: ASL perfusion may show elevated signal in cavernous sinus and/or superior ophthalmic vein
 - Ultrasound: Doppler assessment may show dilated arterialized superior ophthalmic veins
- Management options ¹:
 - Endovascular embolization: first-line with cure rate > 80%
 - Surgery (suturing/clipping fistula, packing cavernous sinus, ligating the ICA) : 31-79% success rate
 - Compression of cervical carotid for 4-6 weeks: effective in 30% indirect, low-flow cases
 - Spontaneous closure: occur in 20-60% of indirect, low-flow cases
- Potential complications with endovascular embolization ²:
 - Transient aggravation of symptoms: increased proptosis, elevation of IOP, choroidal detachment, venous stasis retinopathy
 - Visual acuity loss or ophthalmoplegia (from central retinal vein obstruction, ophthalmic artery occlusion, cerebral infarction), neovascular glaucoma, uncontrolled bleeding

Case Discussion

- Our case is unique in that the patient was diagnosed with spontaneous, non-traumatic bilateral indirect carotid-cavernous fistulas—a presentation that is exceedingly rare.
- Our patient subsequently underwent successful transvenous coil embolization of CCFs with resolution of neurological symptoms at 1-month follow-up.

References:

1. Kohli GS, Patel BC. Carotid Cavernous Fistula. [Updated 2023 Apr 3]. In: *StatPearls*. Treasure Island (FL): StatPearls Publishing; 2025 Jan. Available from: <https://www.ncbi.nlm.nih.gov/books/NBK535409/>
2. Chaudhry IA, Elkhamry SM, Al-Rashed W, Bosley TM. Carotid cavernous fistula: ophthalmological implications. *Middle East Afr J Ophthalmol*. 2009;16(2):57-63. doi:10.4103/0974-9233.53862
3. Ellis JA, Goldstein H, Connolly ES Jr, Meyers PM. Carotid-cavernous fistulas. *Neurosurg Focus*. 2012;32(5):E9. doi:10.3171/2012.2.FOCUS1223
4. Sharma R, Ponder C, Kamran M, et al. Bilateral Carotid-Cavernous Fistula: A Diagnostic and Therapeutic Challenge. *J Investig Med High Impact Case Rep*. 2022;10:23247096221094181. doi:10.1177/23247096221094181