

# GRAND ROUNDS

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# History

HPI: 38F with intermittent L-sided pulsatile headaches and L eye redness and bulging x 2 months

- No changes in visual acuity, no double vision
- Headache not associated with activity, time of day, position
- No fever or recent illness
- No whooshing or ringing sound reported
- No history of head trauma

# History

- PMH: none
- POH: none
- Meds/Gtts: none
- FH: non-contributory
- SH: denies smoking, etoh/IVD use
- All: NKDA

# Examination

- BP: 112/70
- nVa<sub>sc</sub>: 20/20 ou
- Pupils:
  - Light: 3.5mm od, 2.5mm os
  - Dark: 7mm od, 4mm os
  - Hippus ou, dilation lag os, ?slight APD os
- EOMs: full ou, no pain, no diplopia
- CVF: full ou
- Tapp Day 1: 11/20 @ 8:45pm. No pulsatile proptosis appreciated during appplanation.
  - Tapp Day 3: 11/16 @ 10:50am (after initiation of brimonidine o/2)

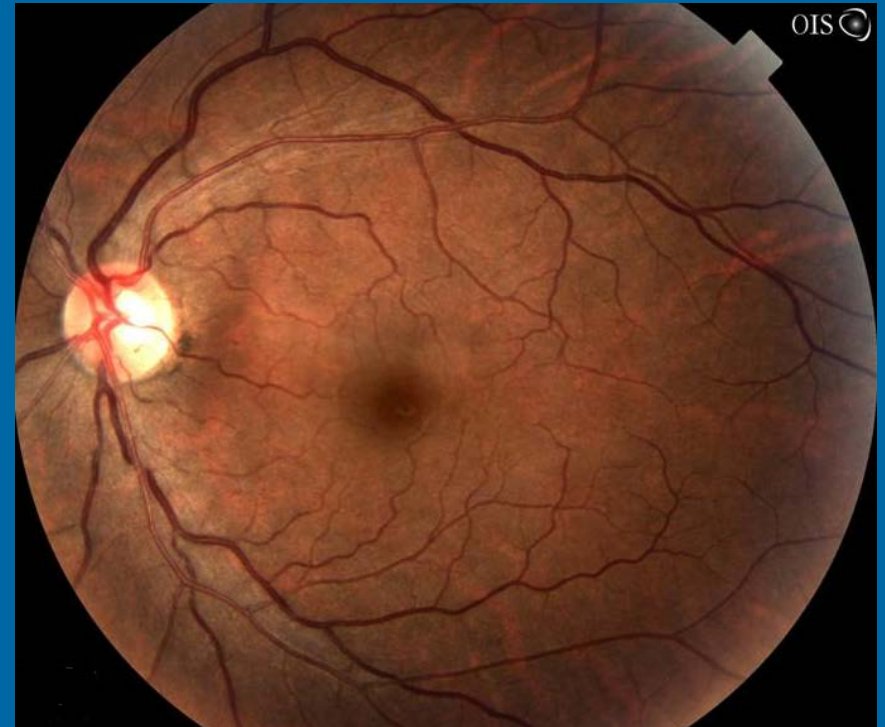
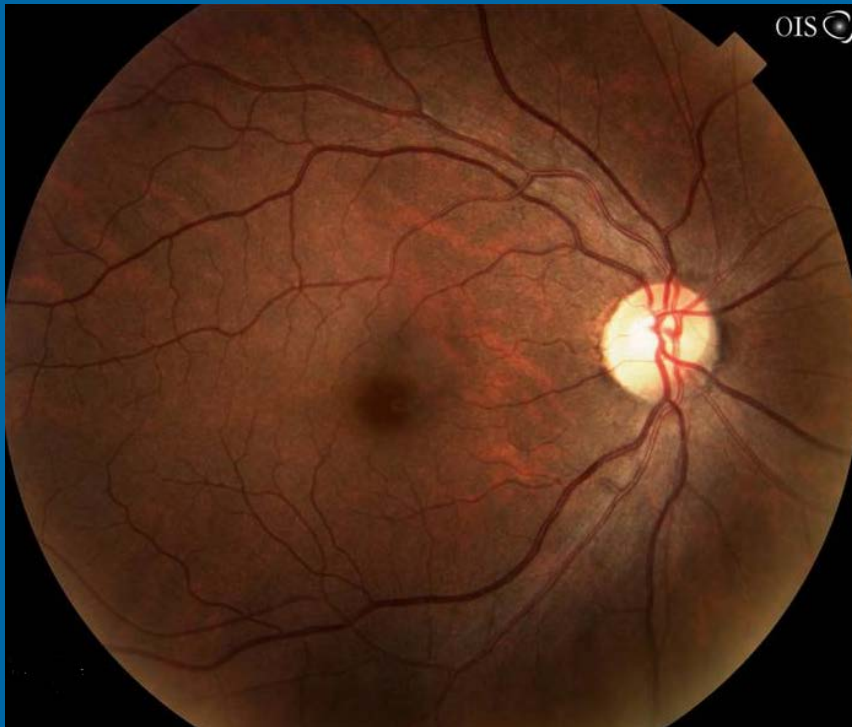
# External Exam



# Examination

- SLE
  - LLA: wnl od; proptosis os, no ptosis ou
  - C/S: w+q od; dilated and tortuous arterialized conjunctival and episcleral vessels os
  - K: clear ou
  - AC: d+q ou
  - I/P: r+r ou
  - L: cl ou
- Hertel: 19/23, base 118

# DFE



# DFE

- Vit: cl ou
- ON: 0.35/0.5, s+p ou
- M: flat ou
- V/P: wnl od; dilated, tortuous veins os



# Examination

- Color plates: 16/16 ou
- Cranial Nerve exam: CN II-VIII grossly intact.  
+corneal sensation ou, no hypoesthesia ou
- No bruit appreciated on auscultation of the globe ou

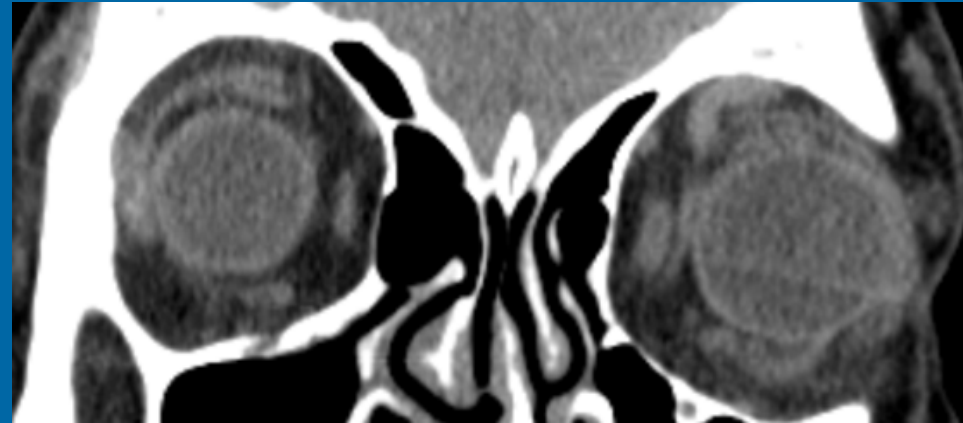
# Differential Diagnosis?

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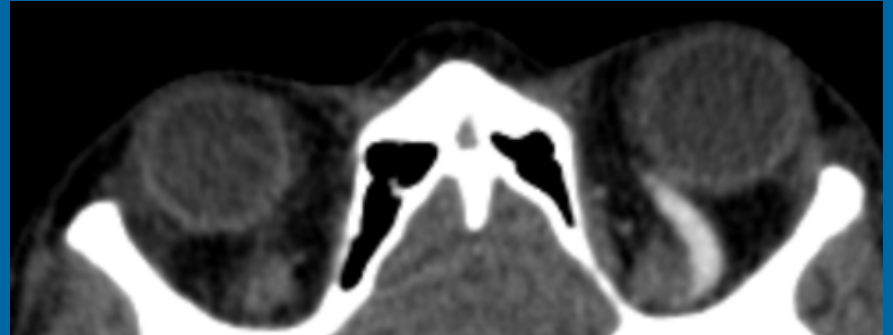
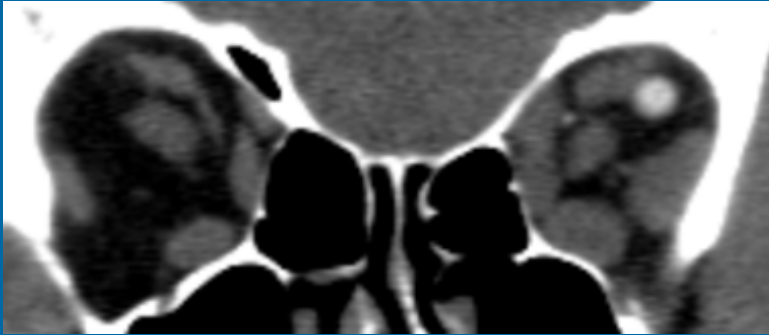
- Increased episcleral venous pressure
  - Carotid cavernous sinus fistula (direct vs. indirect)
  - Orbital venous malformation (orbital varix)
  - Cavernous sinus congenital AV malformation
  - Cavernous sinus thrombosis
- Proptosis
  - Idiopathic orbital inflammation/Tolosa-Hunt Syndrome
  - Thyroid eye disease/Graves ophthalmopathy/dysthyroid orbitopathy
  - Orbital tumor (cavernous hemangioma, meningioma, etc)
  - Cavernous sinus tumor (primary vs. metastatic)
  - Orbital hemorrhage/hematoma
- Red eye
  - Conjunctivitis

# Next Step?

# CT orbits w/o contrast



# CTA intra/extracranial

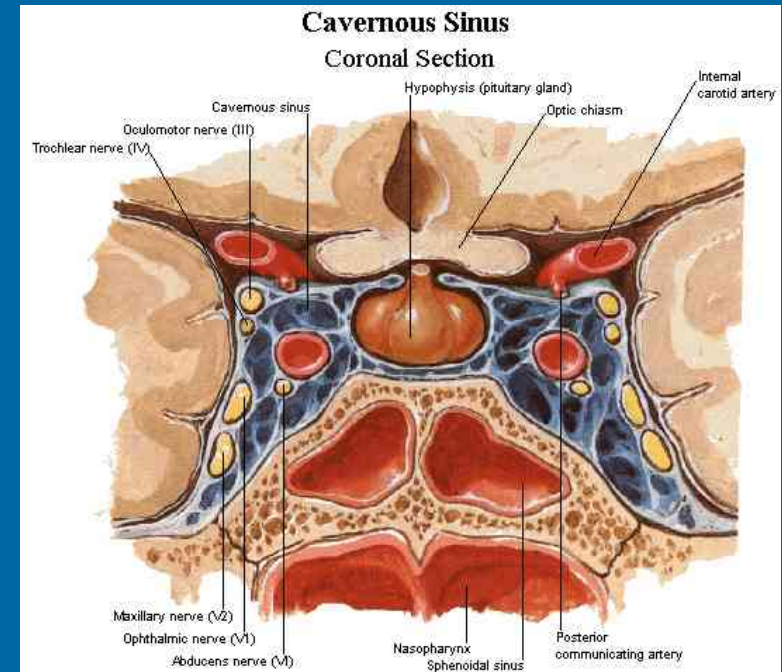


# Objectives

- To review the different types of CCFs
- To review the typical presentation and sequelae of CCFs
- To review the diagnostic and treatment strategies for CCFs

# Carotid Cavernous Sinus Fistula

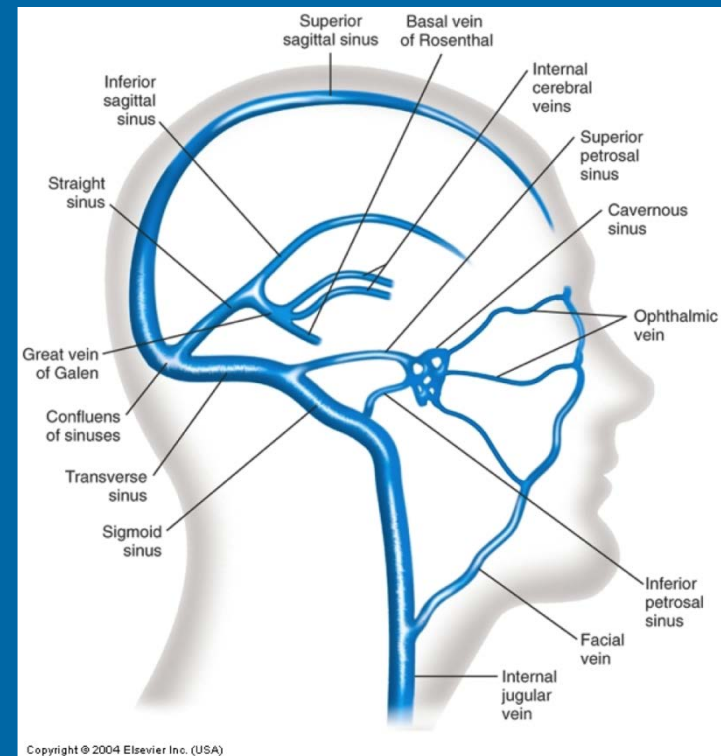
- Abnormal connection between the carotid artery (or its branches) and the cavernous sinus
- High arterial pressures transmitted into the low-pressure venous circulation of the cavernous sinus





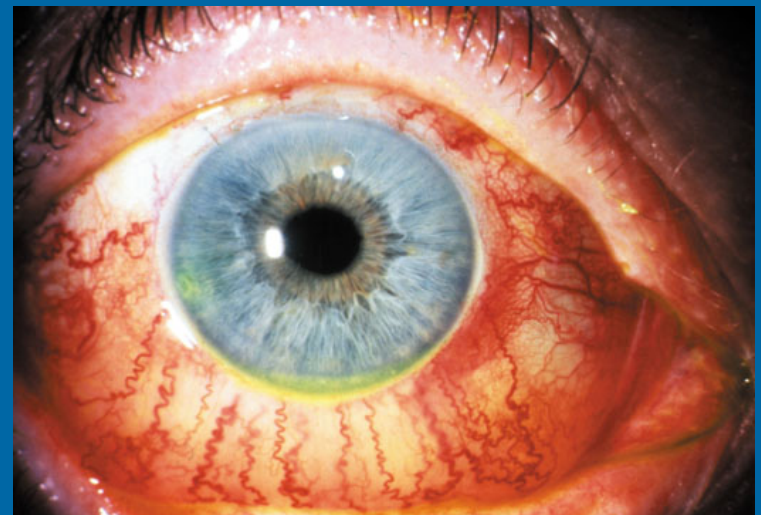
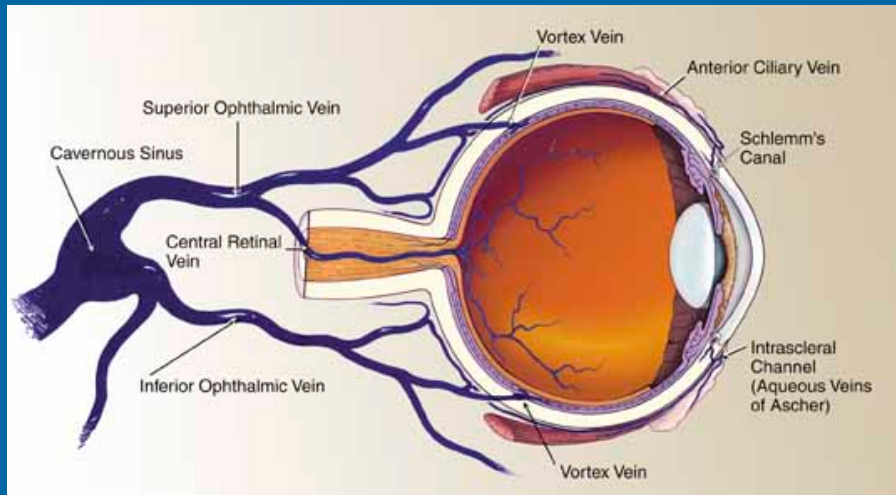
# Cavernous Sinus

- Receives blood from the superior and inferior ophthalmic veins (and some cerebral venous outflow)
- Drains into the superior and inferior petrosal sinuses
- Increased pressure in the sinus → reversal blood flow in the superior ophthalmic vein → orbital venous congestion
- Diversion of arterial blood → ischemia



# Ocular Venous Outflow

- Retinal veins → central retinal vein  
Choroidal veins → vortex veins  
Episcleral venous plexus → ciliary veins
- All three primarily drain into the superior ophthalmic vein
- Venous congestion in CCF leads to the classic initial sign of dilated, corkscrew, arterialized conjunctival and episcleral vessels.



# Classification

- Pathogenesis: traumatic vs. spontaneous
- Hemodynamics: high-flow vs. low-flow
- Angiography: direct vs. indirect

# Classification

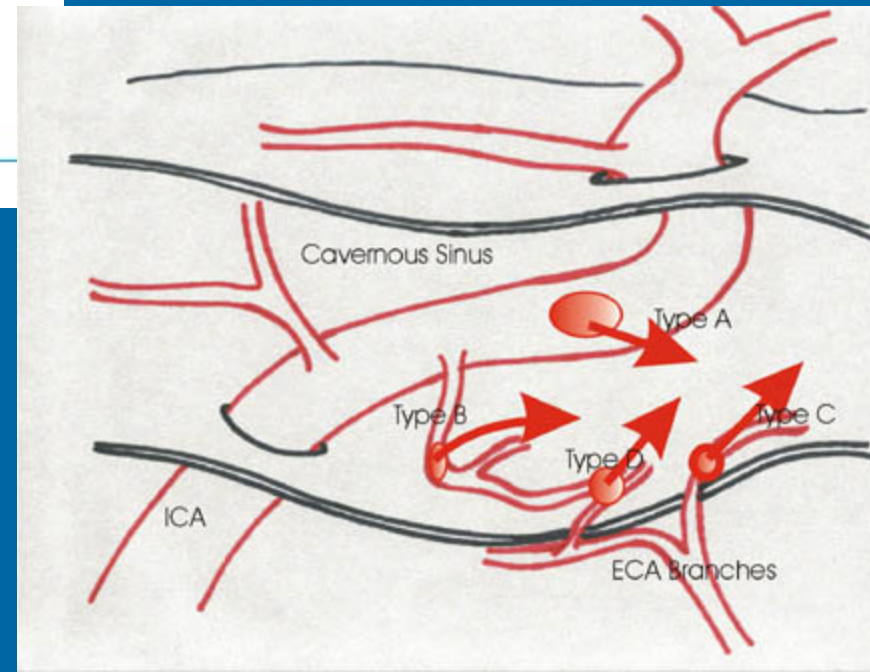
- Direct, high-flow fistula: internal carotid artery and the cavernous sinus
  - Most commonly after severe head trauma (basal skull fracture), which causes a tear in the cavernous portion of the ICA
  - Sometimes from rupture of cavernous ICA aneurysm
- Indirect, “dural”, low-flow fistula: small meningeal branches off the internal and/or external carotids and the cavernous sinus
  - Most often occur spontaneously, exact mechanism unclear
  - More common in middle aged and older women, often with history of HTN and atherosclerosis.
  - Other associated factors: ruptured aneurysm, pregnancy, minor trauma, straining/Valsalva, collagen vascular disease

# Classification

- Barrow et al. Classification

Type	Pathogenesis	Arterial supply	Hemodynamics
A	Head trauma/ aneurysm rupture	ICA	High flow
B	Spontaneous	ICA dural branches	Low flow
C	Spontaneous	ECA dural branches	Low flow
D	Spontaneous	ICA and ECA dural branches	Low flow

ICA: Internal carotid artery, ECA: External carotid artery



# Presentation

- Direct
  - Classic triad: pulsatile proptosis, conjunctival injection (dilated, corkscrew, arterialized episcleral/conjunctival vessels and chemosis), ocular bruit
  - Rapid, dramatic onset
  - Pain, headache
  - Visual loss (multifactorial)

# Presentation

- Cavernous sinus congestion
  - EOM palsies, diplopia (compression of CNs in cavernous sinus; CN VI most common). EOMs also restricted from enlarged muscles secondary to orbital congestion.
  - APD (compression of ON)

# Presentation

- Orbital congestion
  - Dilated episcleral vessels
  - Dilated retinal vessels, retinal hemorrhage, disc edema, CRVO, CRAO, serous RD
  - Choroidal effusion → secondary angle-closure glaucoma
  - Increased IOP (increased episcleral venous pressure) → secondary glaucoma



# Presentation

- Ischemia
  - Retinal ischemia → iris neovascularization → secondary glaucoma
  - Ischemic optic neuropathy
  - Anterior segment ischemia → corneal edema, cell/flare
- Proptosis
  - Dry eyes/exposure keratopathy (if proptosis is significant)

# Presentation

- Indirect
  - Less dramatic, insidious onset
  - No bruit
  - Dilated, corkscrew, arterialized episcleral/  
conjunctival vessels
  - Mild proptosis and pain
  - Increased IOP → secondary glaucoma (more  
chronic)

# Presentation

- Isolated CN VI palsy in context of quiet eye → compression on nerve by enlarged inferior petrosal sinus that drains the fistula
  - Pulsatile tinnitus often present

# Presentation

J Med Assoc Thai. 2008 Jun;91(6):852-8.

**Dural carotid cavernous sinus fistula: ocular characteristics, endovascular management and clinical outcome.**

Preechawat P, Narmkerd P, Jiarakongmun P, Poonvathalang A, Pongpech SM.

- 80 patients with dural CCF (50F, 30M)
- Arterialization of conjunctival vessels: 93%
- Proptosis: 84%
- Cranial nerve palsy: 52%
- Elevated intraocular pressure: 51%
- Decreased vision: 43% (ranging from 20/40 to NLP)
- Chemosis: 42%
- Optic neuropathy: 13%

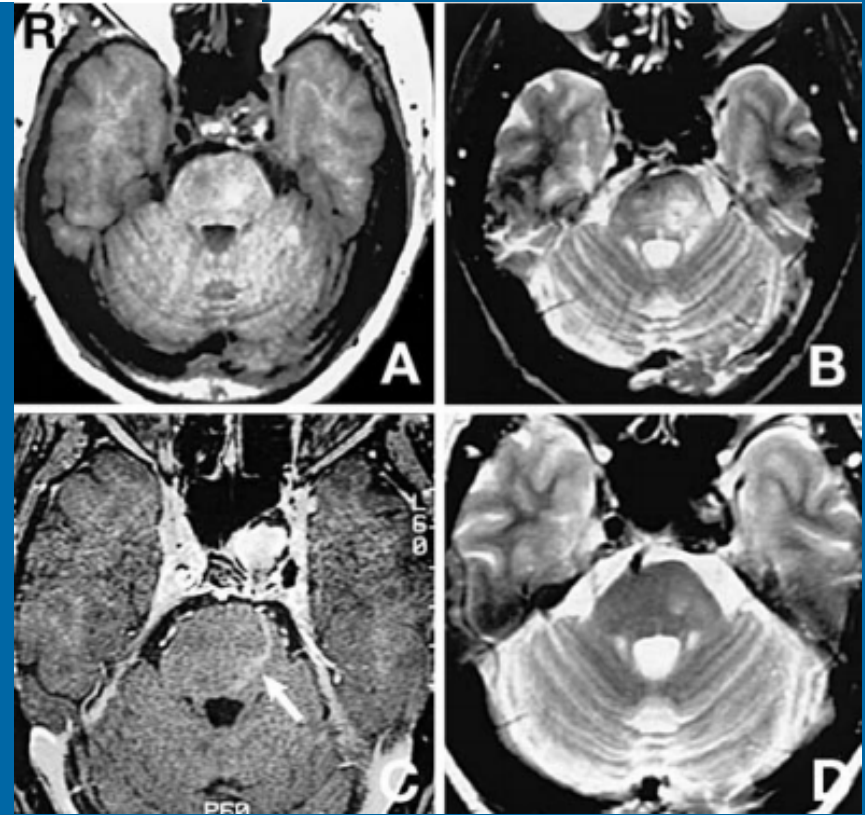
# Rare Presentation

Neurol Med Chir (Tokyo). 2003 May;43(5):255-8.

## Brainstem congestion caused by direct carotid-cavernous fistula--case report.

Murata H, Kubota T, Murai M, Kanno H, Fujii S, Yamamoto I.

- 41F tinnitus L ear, HA, diplopia, L cheek pain.
- L high-flow, direct CCF
- Few days later, sudden R hemiparesis, dysarthria, ocular conjugate deviation to the right, then somnolence
- Angiography: spontaneous occlusion of the superior petrosal sinus with engorgement of veins in the surrounding brainstem with subsequent small pontine hemorrhage and edema
- Brainstem congestion caused by CCF is very rare, but can be life-threatening
- Symptoms resolved after treatment of CCF



A, B: T1, T2 MRI: small pontine hemorrhage and edema

C: T1 MRI with contrast on day 1: engorgement of perforating vein within pontine parenchyma, coinciding with subsequent hemorrhage and edema

D: T2 MRI 4 weeks after: improvement

# Rare Presentation

Chin J Traumatol, 2004 Oct;7(5):317-20.

**Hemiparesis in carotid cavernous fistulas (CCFs): a case report and review of the literature.**

Wang HX, Bai RL, Huang CG, Lu YC, Zhang GJ.

- Hemiparesis from cerebral infarction caused by total steal of blood flow by the CCF
- Recovery after treatment of CCF

# Rare Presentation

Neurosurgery, 2010 Oct;67(4):E1160-3; discussion E1163-4. doi: 10.1227/NEU.0b013e3181edb148.

**Brainstem compression caused by bilateral traumatic carotid cavernous fistulas: case report.**

Gierthmuehlen M, Schumacher M, Zentner J, Hader C.

- 30M s/p motorbike accident with untreatable epistaxis
- Angiography: bilateral traumatic CCFs and bleeding of L sphenopalatine artery
- 3 months s/p treatment: severe headache, dilated R pupil and somnolence
- CT: retroclival mass compressing the brainstem
- Angiography: re-perfused left CCF with huge dilation of the retroclival cavernous sinus
- Symptoms resolved after repeated treatment of CCF

# Radiological Evaluation

- Angiography required for definitive diagnosis, characterization, and treatment planning
- CT/CTA, MRI/MRA often used in initial work-up
  - Proptosis
  - Dilation of superior ophthalmic vein (SOV)
  - Enlargement of EOMs
  - Enlargement of ipsilateral cavernous sinus
- Color Doppler can assist in diagnosis and follow-up
  - Increased velocity of blood flow
  - Reversal of the direction of blood flow
  - Arterial pulsations
  - Dilation of SOV



# Indications for Emergent Intervention

Angiographic findings	Clinical signs and symptoms
Pseudoaneurysm → SAH	Increased intracranial pressure
Large varix of the cavernous sinus → SAH	Rapidly progressive proptosis (may signify spontaneous orbital venous outflow thrombosis)
Venous drainage to cortical veins → hemorrhagic venous infarction	Decreased visual acuity
Thrombosis of distant venous outflow pathways → increased ICP, hemorrhage	Intracerebral, subarachnoid and external hemorrhage
	Transient ischemic attack (cerebral ischemia secondary steal phenomenon)

# Treatment

- Conservative treatment for indirect, dural fistulas
  - Observation for small asymptomatic/mildly symptomatic fistulas: reassurance, education
    - Spontaneous closure can occasionally occur within days to months after presentation
    - Risk of intracranial hemorrhage due to venous arterialization

# Treatment

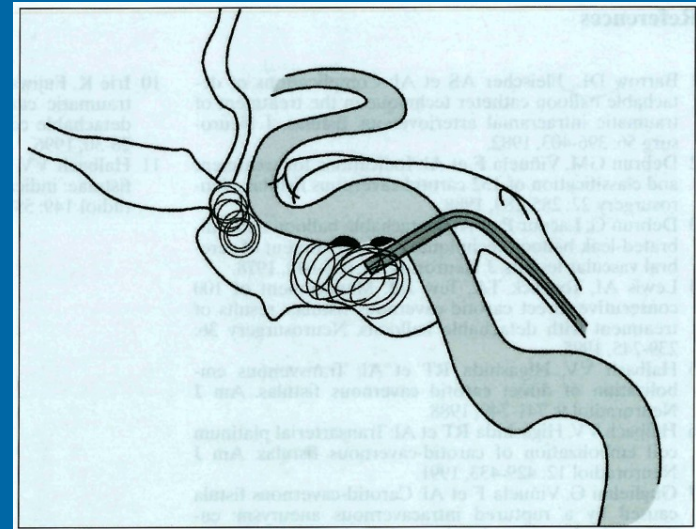
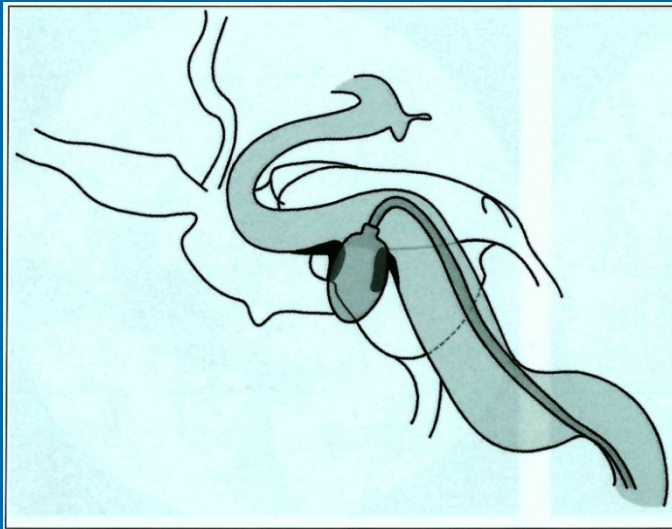
- Conservative
  - Manual external carotid-jugular compression therapy
    - Patient compresses carotid artery and jugular vein with contralateral hand for 10 s, 4-6x/hour
    - Decrease arterial inflow while increased outflow pressure, promoting thrombosis within the fistula
    - Contralateral hand will fall away if cerebral ischemia develops
    - Has been reported to result in spontaneous resolution in up to 30% of patients

# Treatment

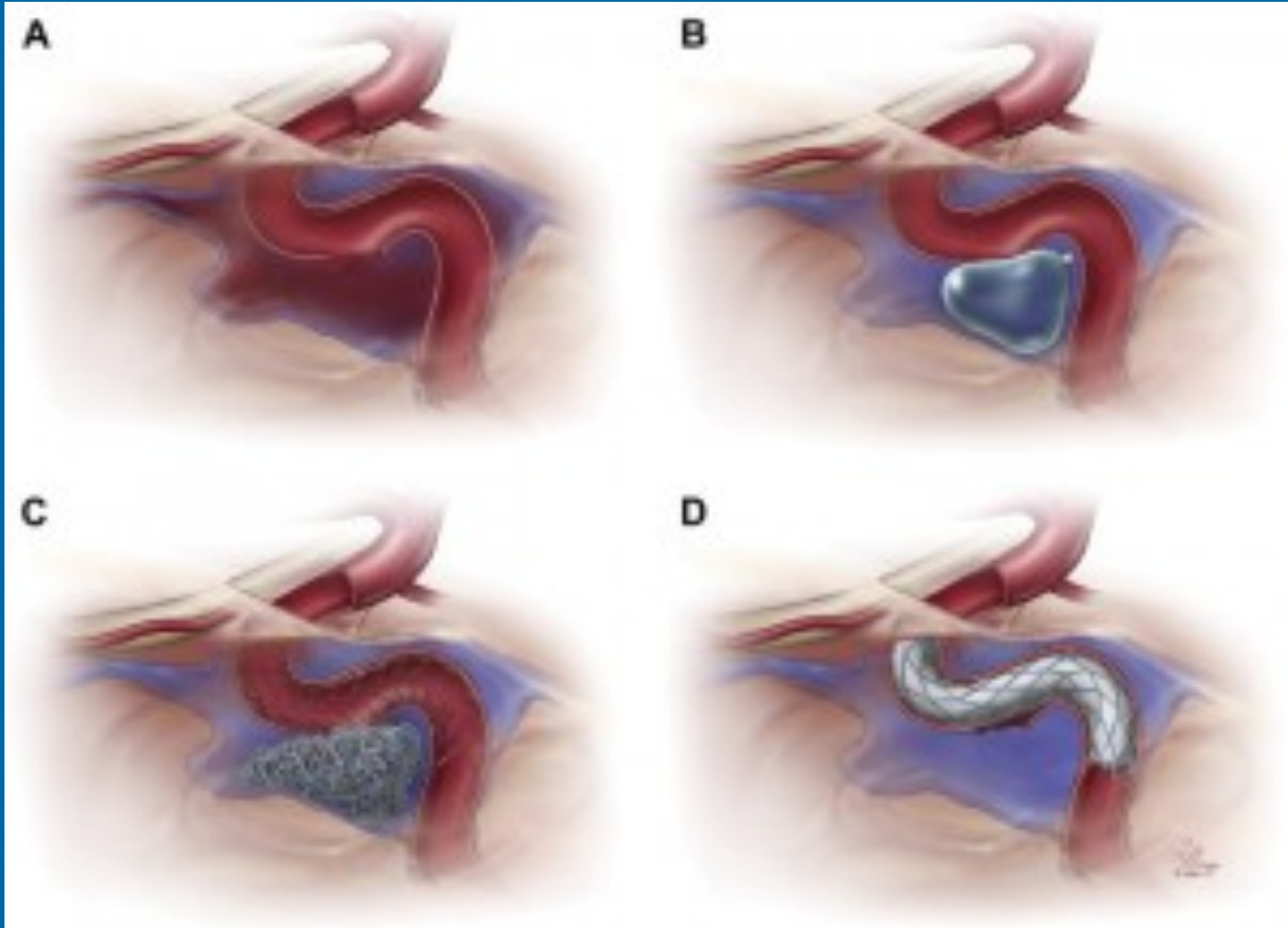
- Direct Fistulas
  - Surgical ligation/trapping
  - Radiosurgery
  - Endovascular embolization
    - Primary treatment option for direct, as well as for the indirect fistulas that are not appropriate for conservative management

# Treatment

- Endovascular embolization (coils, balloons, liquid embolic agents, stents)
  - Direct
    - Previous use of detachable balloons; now replaced by coil embolization, via transvenous or transarterial access. Sometimes combined with balloon or stent placement for protection of the parent vessel.
    - Transvenous access: inferior petrosal sinus, superior ophthalmic vein, femoral access



# Endovascular Embolization of Direct CCF



# Treatment

- Indirect
  - Transvenous embolization more common, but transarterial embolization of the arterial branches supplying the fistula also performed.
  - Can be complicated because many smaller vessels often involved

# Back to our patient

- CTA findings discussed with neuro-radiologist at KCHC.
- Patient determined to likely have a left-sided dural CCF, with possible early Horner's Syndrome (asymmetric pupils) and possible ON involvement (possible slight APD os)
- Referred to Bellevue for angiography with embolization by interventional neuroradiology.





# Back to our patient

- Angiogram: Fistula in the anterior portion of the L cavernous sinus with arterial supply from both the external (mainly a branch from the maxillary artery) and the internal carotid arteries.
  - Barrow's Type D Fistula
- Patient underwent embolization of the maxillary artery branch around the foramen rotundum.
- Patient reported improvement of symptoms after the surgery.

# Review Questions

- What is the classic triad on presentation of a direct CCF?
- What is the primary mechanism of glaucoma associated with CCFs?
- What are the common findings on CT/CTA suggestive of a CCF?

# Reflective Practice

This case allowed me to learn about the pathophysiology, presentation, classification, diagnosis and treatment of carotid cavernous fistula.

It also gave me the opportunity to communicate with and coordinate the timely treatment of my patient with the members of the neurosurgery and interventional neuroradiology teams at Bellevue Hospital.

# Core Competencies

Patient Care: The case involved thorough patient care and timely and appropriate recommendation for treatment. Follow-up was accomplished only via telephone, though patient was instructed and encouraged to come to clinic for follow-up on several occasions.

Medical Knowledge: This case allowed me to learn about the pathophysiology, presentation, classification, diagnosis and treatment of carotid cavernous fistula.

Practice-Based Learning and Improvement: This presentation included a literature search of common and rare presentations of carotid cavernous fistulas, as well as the current treatment methods.

Interpersonal and Communication Skills: Every effort was made to communicate clearly with the patient, as well as with the neurosurgery, interventional radiology and ophthalmology teams at Bellevue Hospital to coordinate appropriate and timely treatment of the patient.

Professionalism: The patient was diagnosed and referred for treatment in a timely and appropriate manner.

Systems-Based Practice: The ophthalmology service at Kings County Hospital and the neurosurgery and interventional radiology services at Bellevue Hospital worked together to appropriately treat the patient.

# References

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- Korkmazer, B., et al. Endovascular treatment of carotid cavernous sinus fistula: A systematic review. *World J Radiol.* 2013 April 28; 5(4): 143-155.
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- Preechawat, P., Dural carotid sinus fistula: ocular characteristics, endovascular management and clinical outcome. *J Med Assoc Thai.* 2008 Jun; 91(6): 852-8.
- Section 5: Neuro-Ophthalmology. Basic and Clinical Science Course. American Academy of Ophthalmology. 2013-2014.
- Section 7: Orbit, Eyelids, and Lacrimal System. Basic and Clinical Science Course. American Academy of Ophthalmology. 2013-2014.
- Wang, HX, et al. Hemiparesis in carotid cavernous fistulas (CCFs): a case report and review of literature. *Chin J Traumatol.* 2004 Oct; 7(5): 317-20.

# Thank you

- Patient
- Dr. Elmalem
- Dr. Shinder
- Interventional neuroradiology, neurosurgery and ophthalmology teams at Bellevue Hospital.