Pulsatile Proptosis

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Grand Rounds

January 10th, 2020

Patient Presentation

CC

Red, proptotic eye

HPI

Consult from neurosurgery for eye injury with proptosis in a 74 yo WF who fell down a flight of stairs. She sustained many bodily fractures, facial and skull fractures, and subarachnoid hemorrhage. She is now intubated and sedated in the ICU.

History

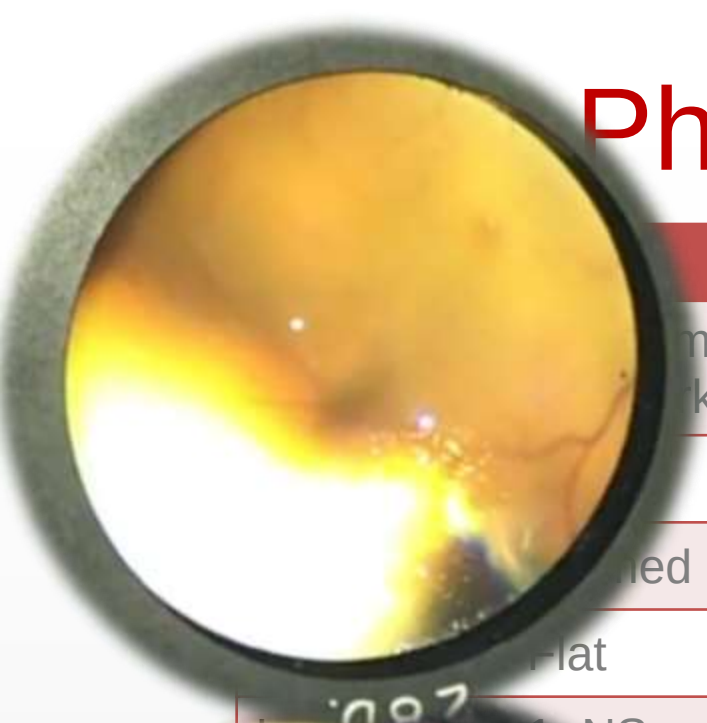
Past Medical History	Squamous Cell Carcinoma Migraine
Family Hx	Noncontributory
Meds	Sumatriptan, ASA 81mg
Allergies	Sulfa
Social Hx	-Never Smoker -1 drink per day alcohol -No illicits
RoS	Unable to obtain

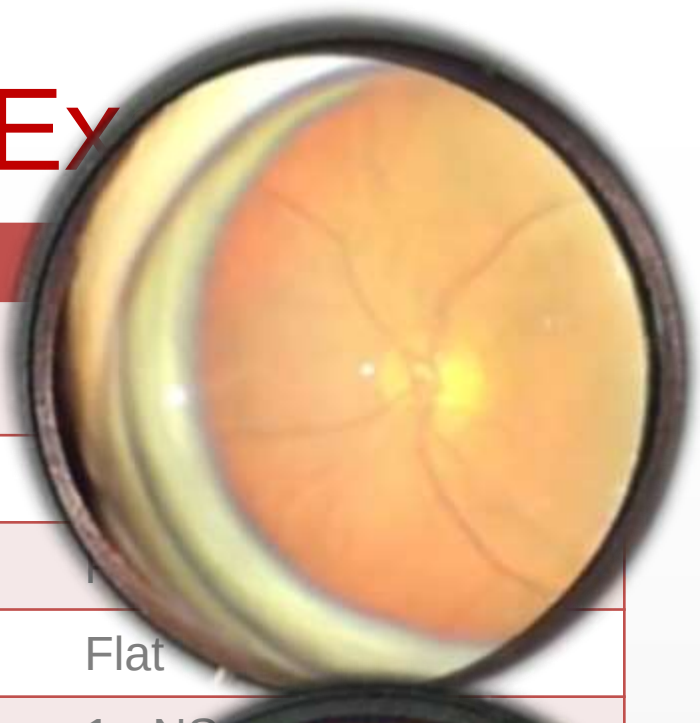
Physical Exam

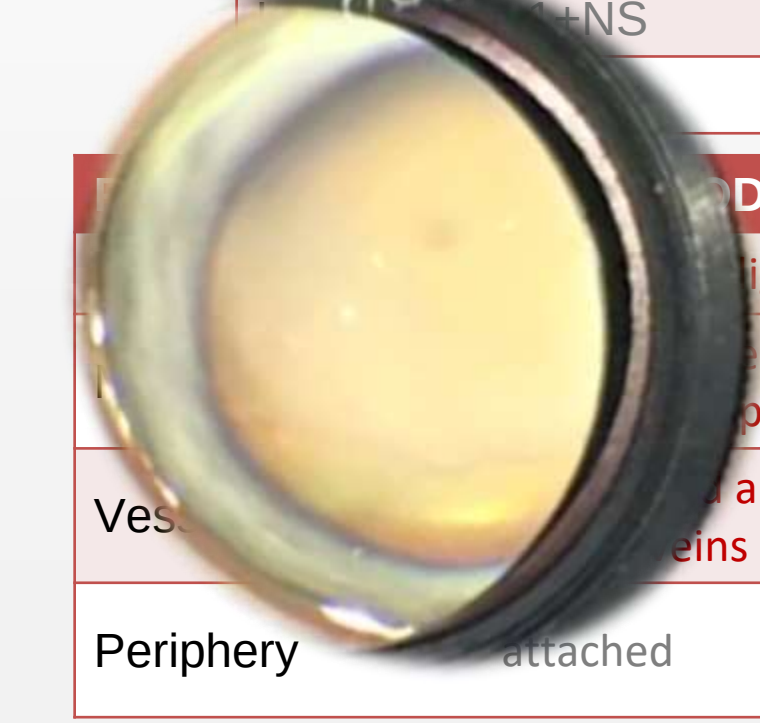
	OD	OS
VAscD	Unable to Obtain	Unable to Obtain
Pupils	**3+ RAPD**	4→3mm
IOP	**55 mmHg**	14 mmHg
EOM	Unable to Obtain	Unable to Obtain
CVF	Unable to Obtain	Unable to Obtain
Lids	**Ecchymosis, edema**	**Ecchymosis, edema**

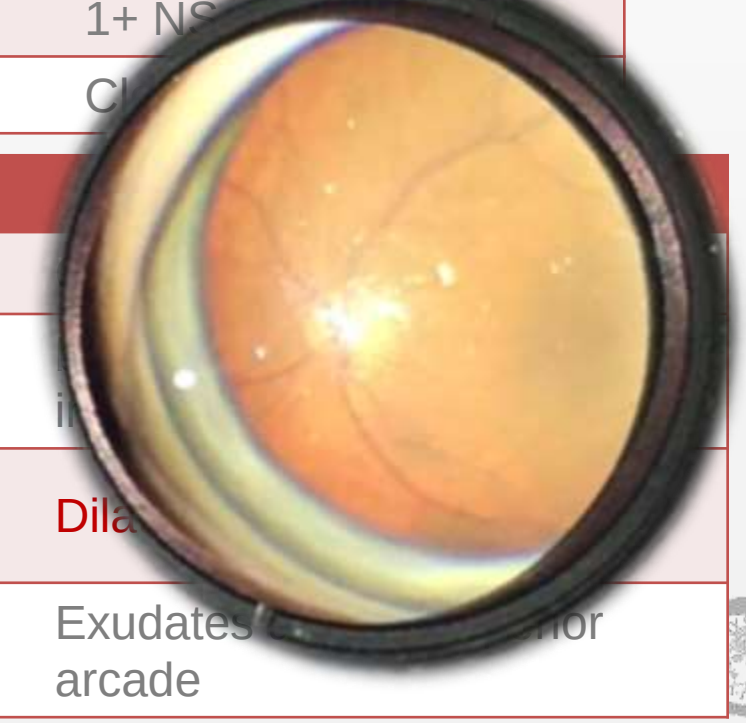
External Exam

Physical Ex



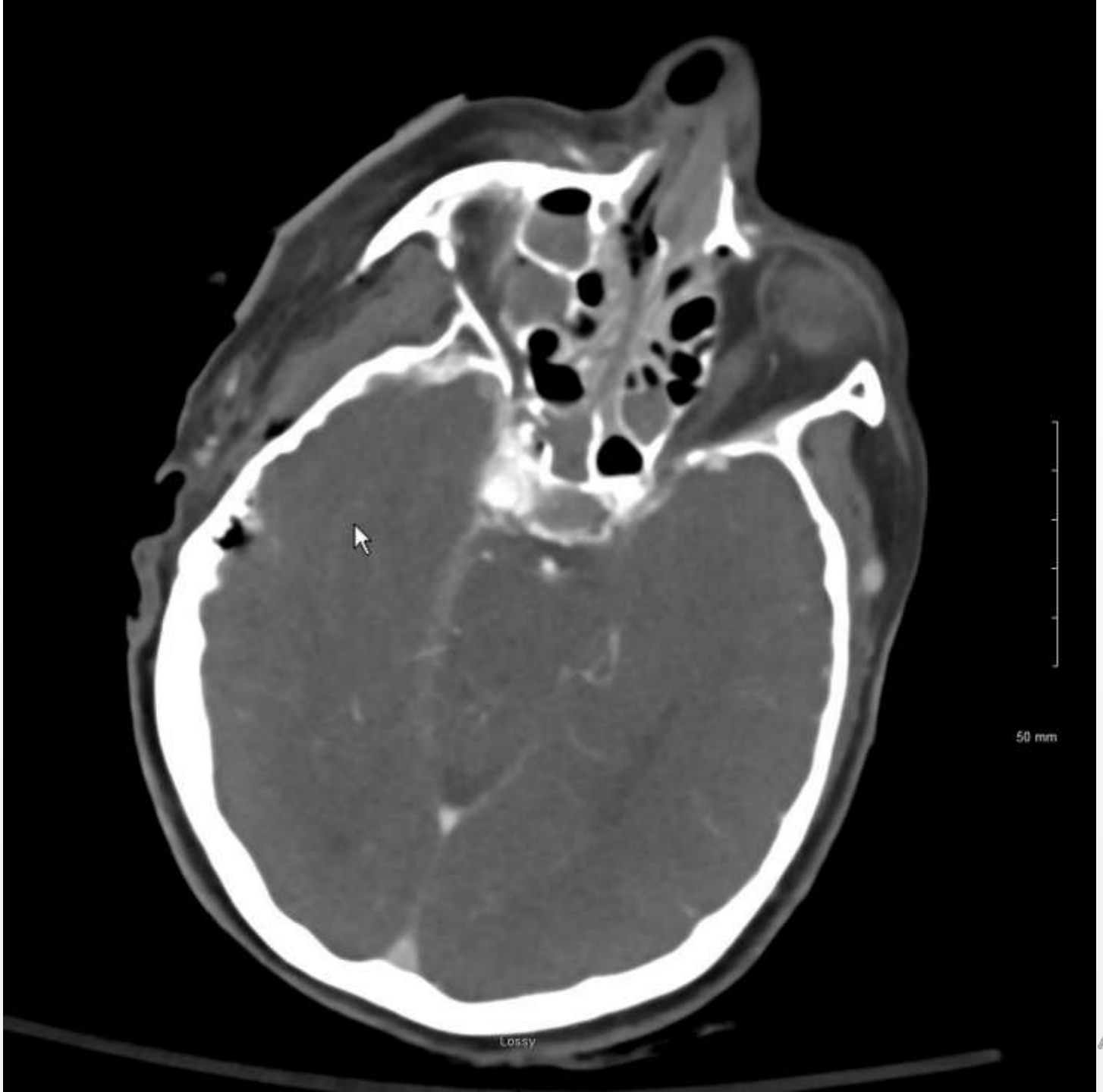






	OD	
	mosis, injection kscrew vessels	
	ied	
	Flat	Flat
	1+NS	1+ NS
		Cl

	OD	
	ight pallor	
	ening with pot	
Ves	arteries, eins	Dila
Periphery	attached	Exudates arcade


50 mm
Lossy

Assessment

- 74 yo WF intubated and sedated after a fall down flight of stairs sustaining skull and facial fractures and subarachnoid hemorrhage, now with pulsatile proptosis, severe injection and chemosis, RAPD, and retinal whitening with cherry red.

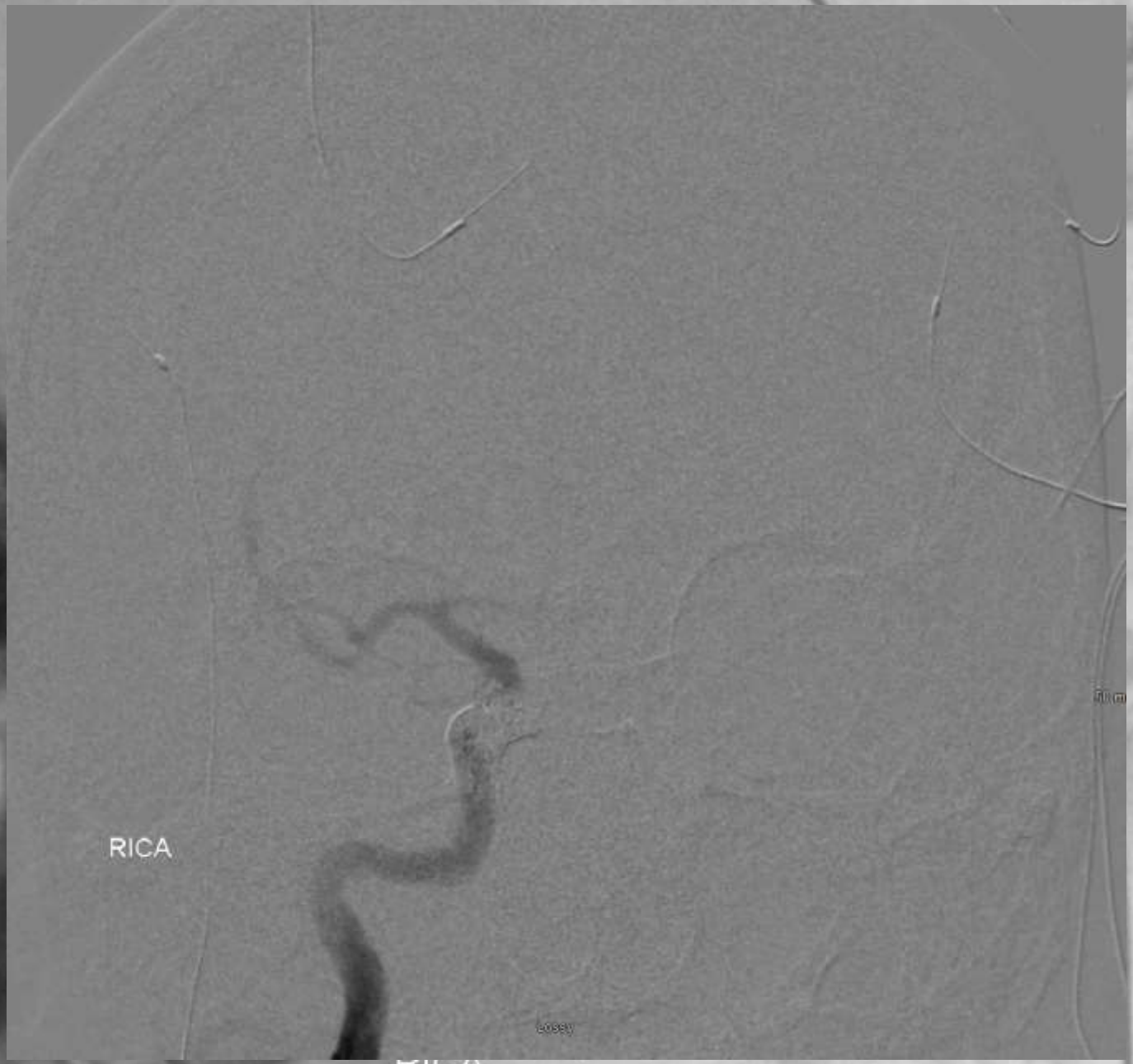
- Concerning for **Carotid Cavernous Fistula** and **Central Retinal Artery Occlusion**

- Differential Diagnosis of pulsatile proptosis
  - CCF fistula
  - Normal intracranial pulsation transmitted to the orbit due to skull base fracture

Plan

- Informed neurosurgery that clinically she appears to have C-C fistula and recommend neurosurgical intervention.

- Transcatheter embolization of a traumatic intracranial carotid-cavernous fistula with platinum coils


RICA

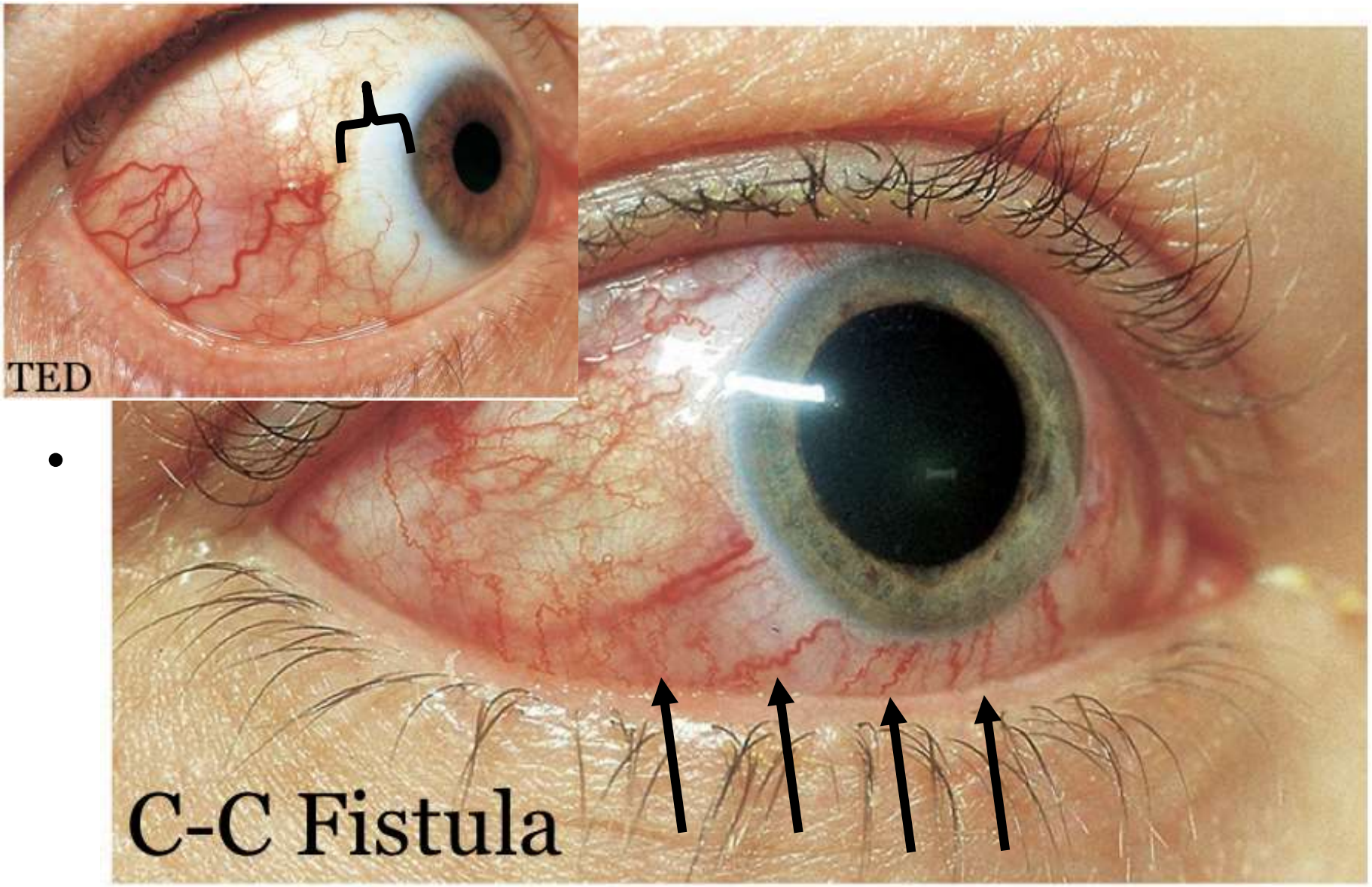
Follow Up

- Recovered well from systemic injuries
- Remains NLP
- Suffered CN III and CN VII palsies
  - Exposure keratopathy
    - tarsorrhaphy

Carotid Cavernous Fistula

- Classification
  - Anatomy: Direct (70-90%) vs Dural
  - Etiology: Traumatic vs Spontaneous
  - Velocity: High vs Low flow
- Majority are Direct Traumatic High Flow CCFs
- Dural CCFs
  - Low flow with communications from meningeal arterial branches to dural veins
  - Hypertension, Atherosclerosis, Collagen vascular disease, Childbirth

Signs and Symptoms



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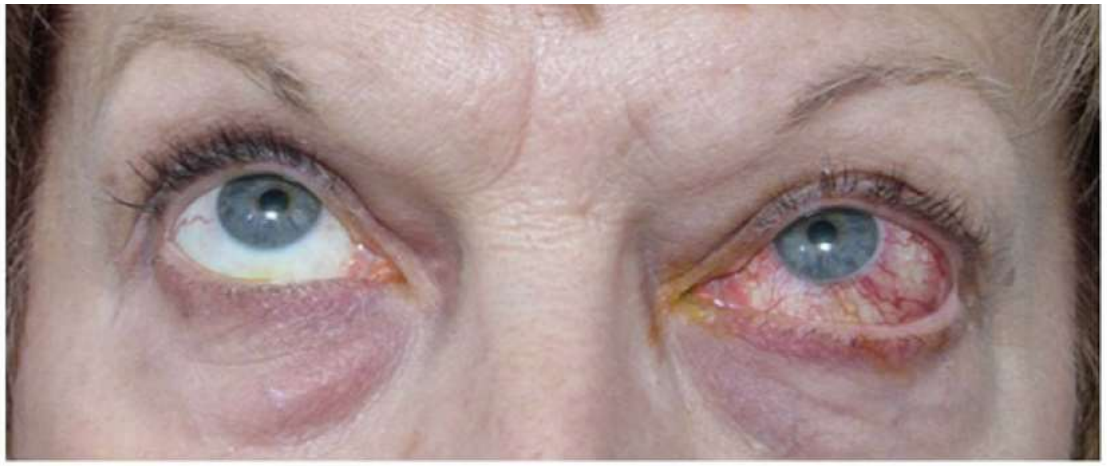
Possible Posterior Findings

- Dilation of retinal veins
- Intraretinal hemorrhages
- Optic disc swelling
- Disc hyperemia
- Retinal detachments
- Choroidal detachments



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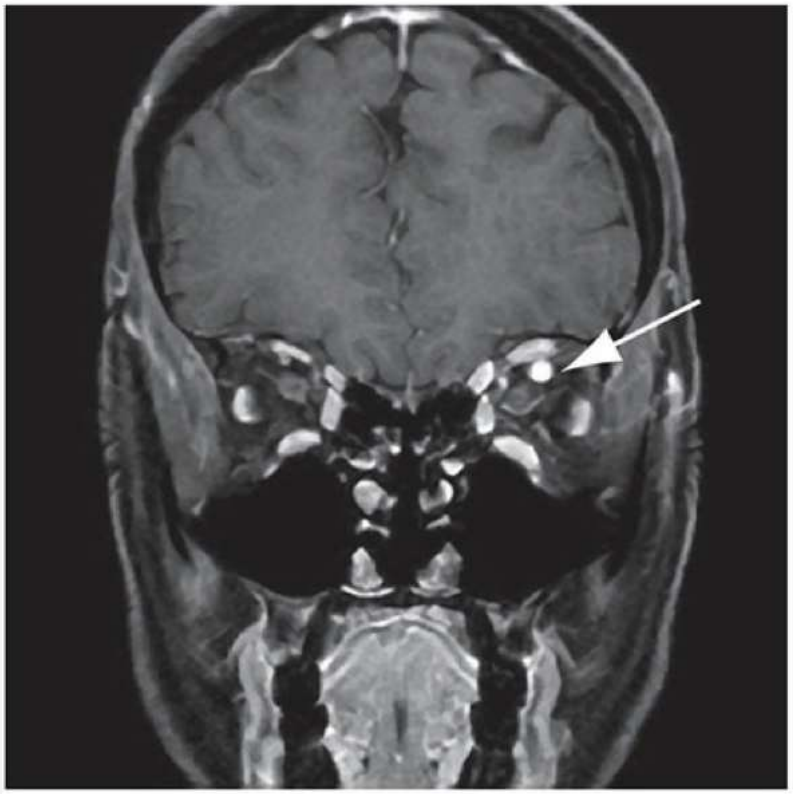

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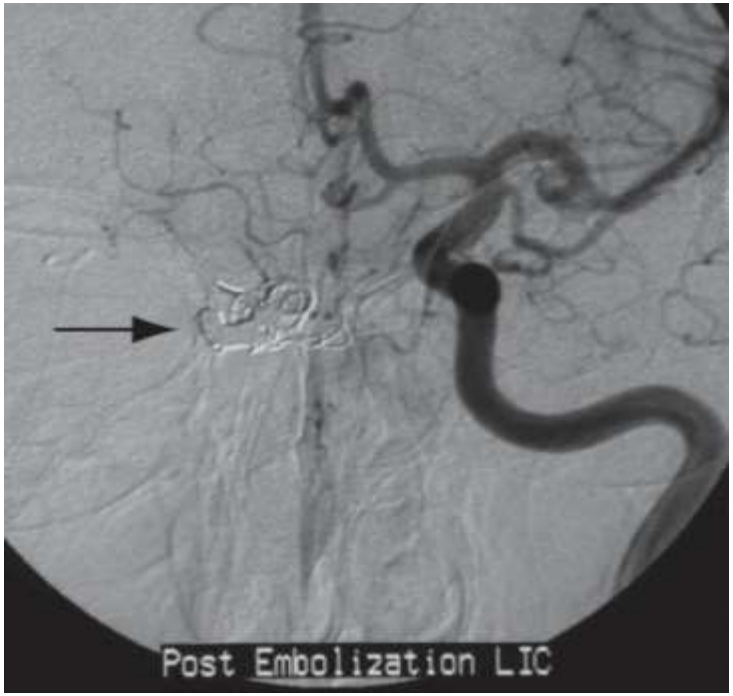
Radiologic Classification

- Type A (Direct most common)
  - Intracavernous ICA and cavernous sinus
- Type B (Dural)
  - Meningeal branch of the intracavernous ICA and cavernous sinus
- Type C (Dural)
  - Meningeal branches of the external carotid artery and cavernous sinus
- Type D = B+C

Complications

- Glaucomatous damage
- Venous stasis retinopathy
- Central retinal vein occlusion
  - Rarely CRAO
- Choroidal detachment
- Exudative retinal detachment
- Anterior Ischemic Optic Neuropathy
- Deterioration of vision
- Intolerable bruit
- Diplopia
- Proptosis leading to corneal exposure

Treatment



Direct CCF

- Endovascular techniques with Neurointerventional radiologist
  - Percutaneous transarterial embolization
  - Platinum coils
- Surgical Treatment with Neurosurgeon
  - Nonballoon embolization
  - Electrothrombosis
  - Craniotomy

Dural CCF

- Observation
  - 10-60% spontaneous closure
- *New:* Transvenous embolization as initial therapy
- Self-carotid compression
  - Compress carotids for 10-30 seconds several time per hour

Ophthalmology's role in treatment

- Elevated IOP
  - Topical antiglaucoma medications
  - Peripheral iridotomy
  - Filtering Surgery
  - Panretinal photocoagulation in case of NVG

Prognosis

- After Fistula closure
  - Immediate resolution of ocular bruits and pulsations
  - Days to Months: Conjunctival chemosis, conjunctival arterialization, eyelid edema, Venous stasis retinopathy, Disc swelling
  - Immediate to months: elevated IOP
- Dural CCFs can reform while direct usually remains closed
- Direct CCFs may not have resolution of proptosis or visual loss

Visual impairment in high flow and low flow carotid cavernous fistula

Md. Shahid Alam[1], Mukesh Jain[2], Bipasha Mukherjee[1], Tarun Sharma[2], Swatee Halbe[3], Durgasri Jaisankar[2] & Rajiv Raman[2]

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- Retrospective study
- Tertiary eye center in South India
- All CCF patients seen at center between June 2001 - June 2015
- 48 patients with DSA proven CCF

Characteristics	High flow	Low flow			P value (high vs. low)
	A(n = 8)*	*B*(n = 6)*	*C*(n = 7)*	*D*(n = 27)*	
Mean age (years)	26.6	54.0	42	55.3	**<0.0001**
Gender (M/F)†	6/2	4/2	4/3	14/13	0.440
History of trauma	7 (87.5%)	0	1 (14.3%)	1 (3.7%)	**<0.0001**
Laterality (Unilateral)	7 (87.5%)	6 (100%)	7 (100%)	24 (88.9%)	1.000

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Characteristics	High flow	Low flow			P value (high vs. low)
	A(n = 8)*	*B*(n = 6)*	*C* (n = 7)*	*D*(n = 27)*	
Retinal vein dilatation	5 (62.5%)	4 (66.7%)	4 (57.1%)	13 (48.1%)	0.710
Intraretinal Hemmorhages	3 (37.5%)	3 (50%)	1 (14.3%)	8 (29%)	1.000
Preretinal Hemmorhage	1 (12.5%)	1 (16.8%)	0	2 (7.4%)	0.189
Vitreous Hemmorhage	1 (12.5%)	0		0	0.167
Macular edema	0	1 (16.8%)	0	3 (11.1%)	0.594
Choroidal detachment	0	0	0	1 (3.7%)	1.000
Retinal detachment	0	0	0	1 (3.7%)	1.000
Disc Hyperaemia	1 (12.5%)	3 (50%)	1 (14.3%)	5(18.5%)	1.000
Disc pallor	1 (12.5%)	0	0	1 (3.7%)	0.671

Disease	High flow	Low flow			P value (high vs. low)
	A (n = 8)*	*B* (n = 6)*	*C*(n = 7)*	*D* (n = 27)*	
CRVO†	0	0	0	2 (7.4%)	1.000
Glaucomatous cupping	1 (12.5%)	2 (33.3%)	1 (14.3%)	3 (11.1%)	1.000
CRAO‡	1 (12.5%)	0	0	0	0.167
ION	1 (12.5%)	0	0	1 (3.7%)	
TON	2 (25.0%)	0	0	0	

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- Direct or Dural more common?
  - Traditionally thought Direct 70-90%
  - This study shows Dural:
  - Why?
    - Decreased head injuries from improved traffic regulations
    - Greater sensitivity of modern imaging finding undiagnosed dural CCFs
  - Most Durals were type D -> C -> B
  - Late presentation of Visual impairment in Dural CCFs, requires high index of suspicion

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Mechanism of Visual Impairment:

Direct CCF

- Immediate
  - Traumatic optic neuropathy
  - Vitreous Hemorrhage
  - Central retinal artery occlusion
- Delayed
  - Ischemic Optic Neuropathy
  - Glaucomatous optic damage

Dural CCF

- Delayed d/t Chronic hypoxia-induced retinal dysfunction
  - Stasis retinopathy
  - Central retinal vein occlusion
  - Ischemic optic neuropathy
  - Glaucomatous optic damage
  - Combined retinal and choroidal detachment

Visual impairment in high flow and low flow carotid cavernous fistula

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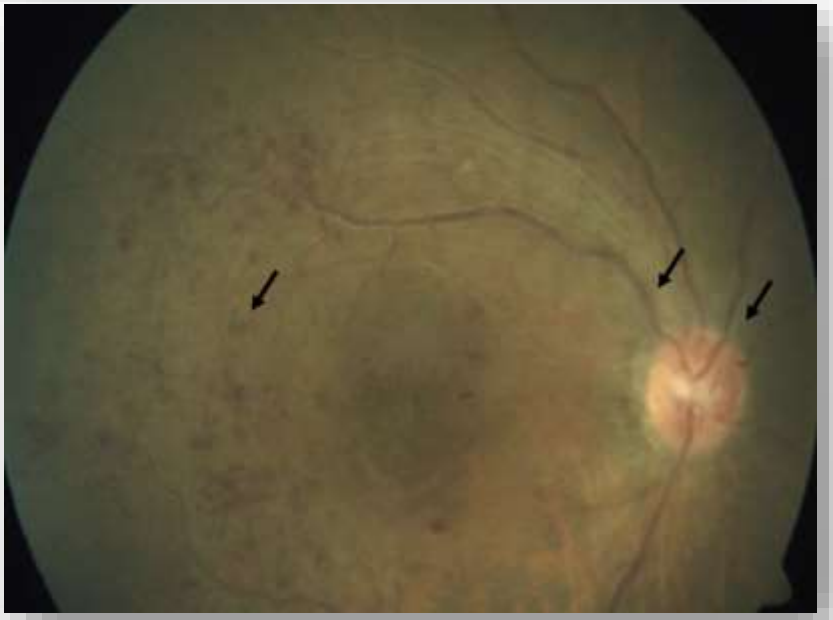
"3 point sign"

In Dural CCF:

- predict moderate visual Impairment

- Retinal vein dilatation
- Intra-retinal hemorrhages
- Disc hyperemia



Visual Impairment†	High flow	Low flow			p value (high Vs low)
	A(n=8)*	*B*(n=6)*	*C* (n=7)*	*D*(n=27)*	
Mild	2 (25%)	4 (66.7%)	5 (71.4%)	21 (77.7%)	*0.017*
Moderate	1 (12.5%) *ION††*	2 (33.3%) *Stasis retinopathy*	1 (14.3%) *Stasis retinopathy*	2 (7.4%) *Stasis retinopathy*	
Severe	1 (12.5%) *GON‡‡*	0	1 (14.3%) *GON‡‡*	1 (3.7%) *ION††*	
Blindness	4 (50%) *[2-TON^;1-TS$ 1- CRAO**]*	0	0	3 (11.1%) *[2-CRVO‡; 1-RD+CD#]*	

Conclusions

- Consider Direct CCF in trauma setting
- Ophthalmology may be the provider to catch the dx
- Keep low index of suspicion for dural CCF as they may be subtle and present to clinic with “red eye”
- Consider the “3 point sign” – venous dilation, intraretinal heme, disc hyperemia
- Don’t throw away your stethoscope

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