



大外科晨會

SPC

莊銘榮 / 黃棣棟主任

Chief Complain

- 17 y/o male
- CC: Progressive headache since 2 weeks ago
- Character of his headache :
 - => Abrupt onset with dull pattern over right frontal region.
 - => There was no aura.
 - => The occurrence was at **night time** more often.
 - => No obvious nausea and vomiting were noted.

Present illness

- Initial symptoms included rhinorrhea and nasal obstruction.
- Upper respiratory infection was diagnosed initially at LMD.



- His headache became worse after one week later.
Throbbing pain (+), **Right eye photophobia (+)**
- The headache usually happened during sleep and made his awake.
- **Nausea and vomiting** were noted.

Present illness

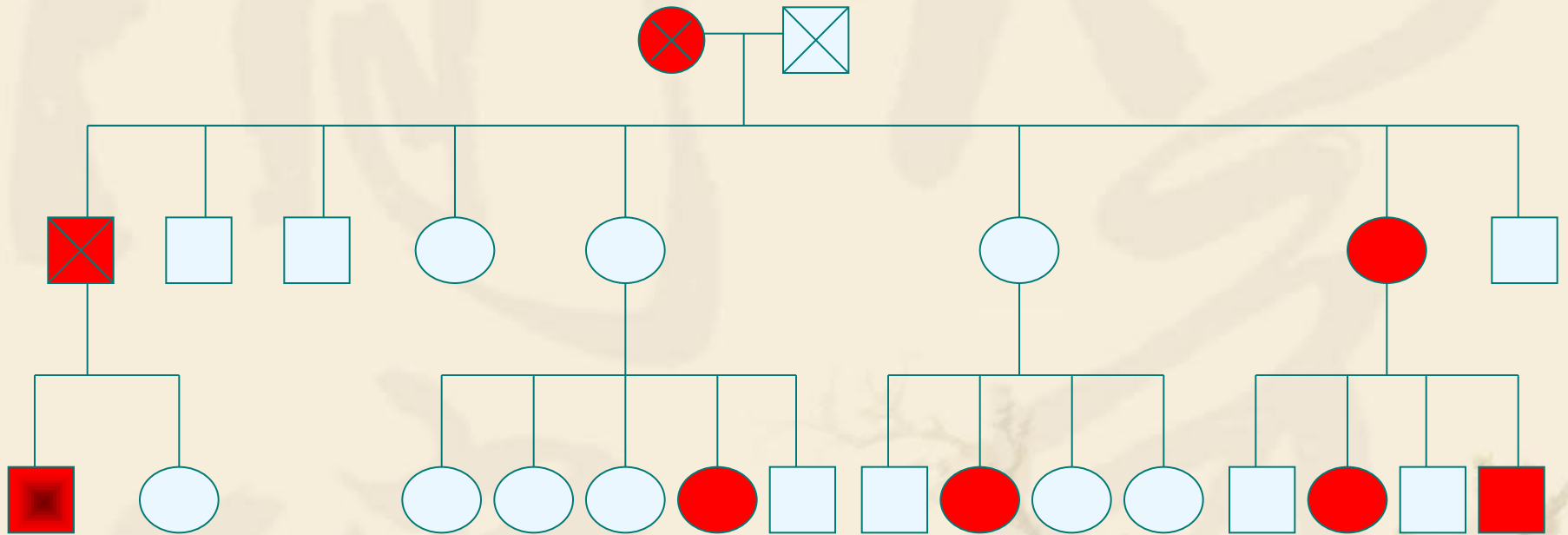


- Therefore, he was admitted at 亞東 hospital on 08-28 and arranged for MRI.
- Due to image findings, he was transferred to our hospital on 08/31.

History

- Past history:
=> Asthma history when infant. (last attack since 4 y/o)
- Birth history:
=> BW=1600 at 32th+ week via C/S(保溫箱10+天)
- Development : His development milestone was normal
- Allergy history :
=> Unknown 感冒藥 allergy with systemic skin rash

Family history



Neurological exam

- NE/PE:

Awaked and alert with good mentality

No obvious IICP sign, Supple neck

EOM: Full EOM without parinaud's sign or sunset sign.

Mild left facial palsy, central type, House grading II

No other cranial nerve dysfunctions

No motor weakness or sensory deficit

Initial Lab data

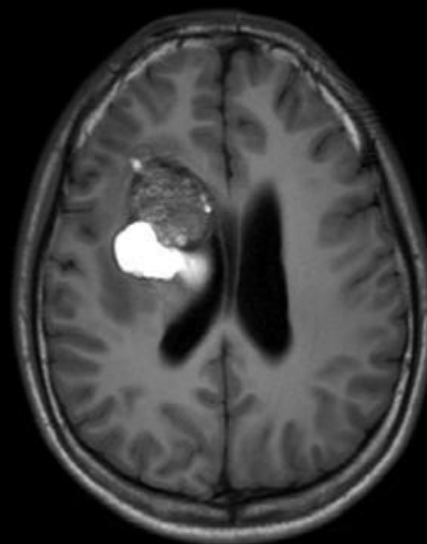
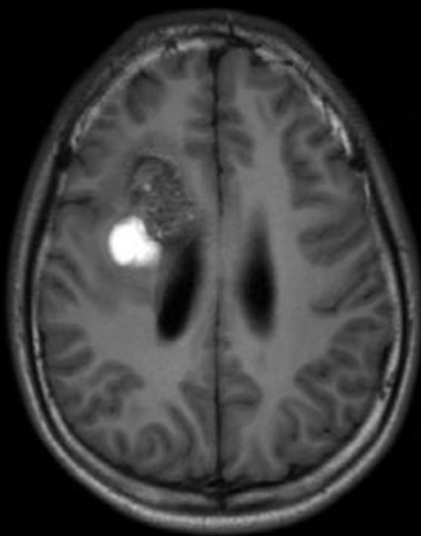
報告日期：2012/08/31

檢驗報告結果列表

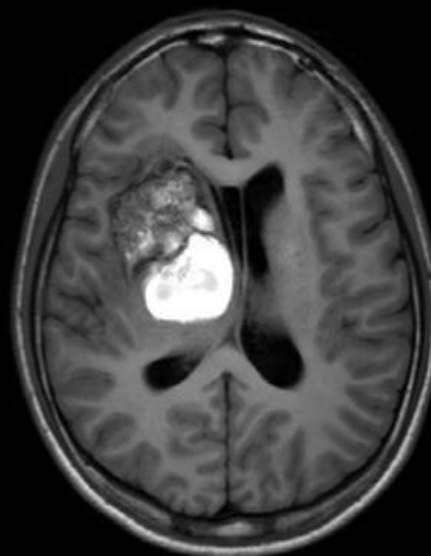
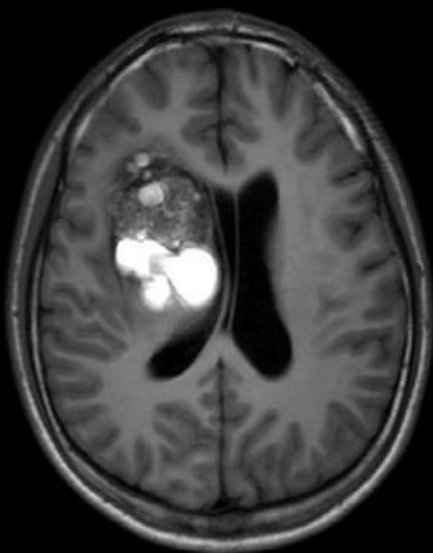
歷史資料	項目名稱	檢驗報告	單位	正常值(Low)	正常值(High)
趨勢圖	Glucose AC	114	mg/dL	70.000	110.000
趨勢圖	Creatinine	0.55	mg/dL	0.440	1.270
趨勢圖	ALT	12	IU/L	5.000	50.000
趨勢圖	Na	140	mmol/L	136.000	144.000
趨勢圖	K	3.7	mmol/L	3.600	5.100

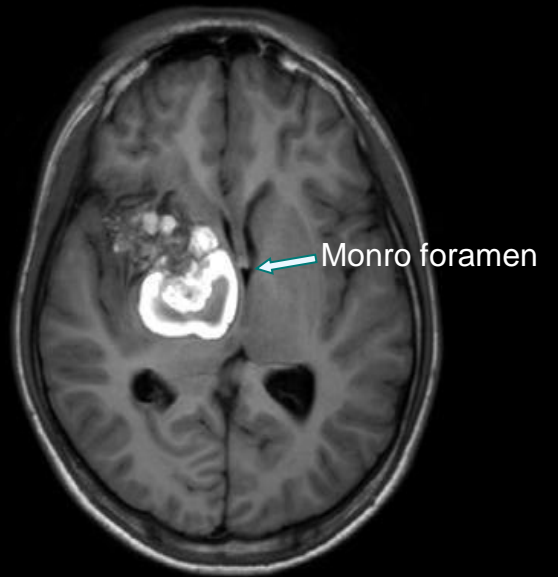
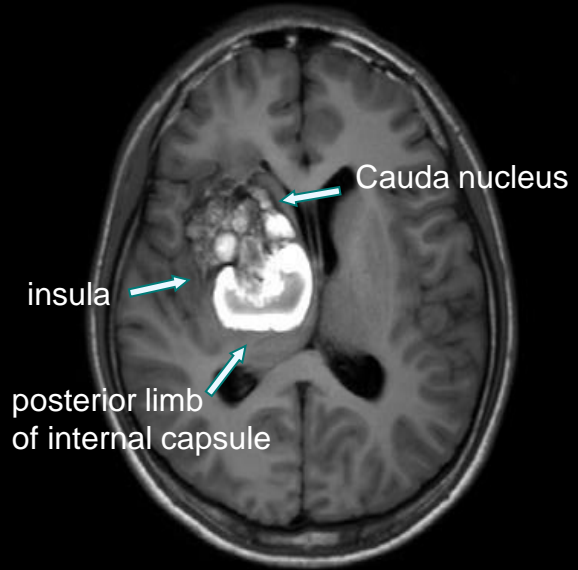
The image features a traditional Chinese ink wash painting of a plum blossom branch. The branch is dark and gnarled, with small, delicate flowers and buds. The background is a light, textured wash. The painting is framed by a decorative border at the top and bottom, consisting of a repeating geometric pattern of triangles and circles. The text "Brain MRI (C+/-)" is centered over the painting in a bold, blue font.

Brain MRI (C+/-)



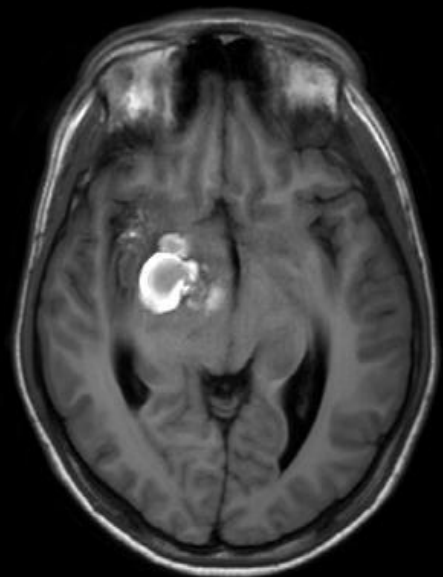
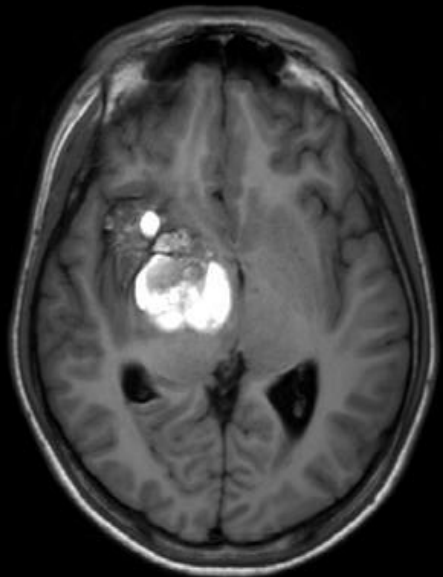
T1WC-





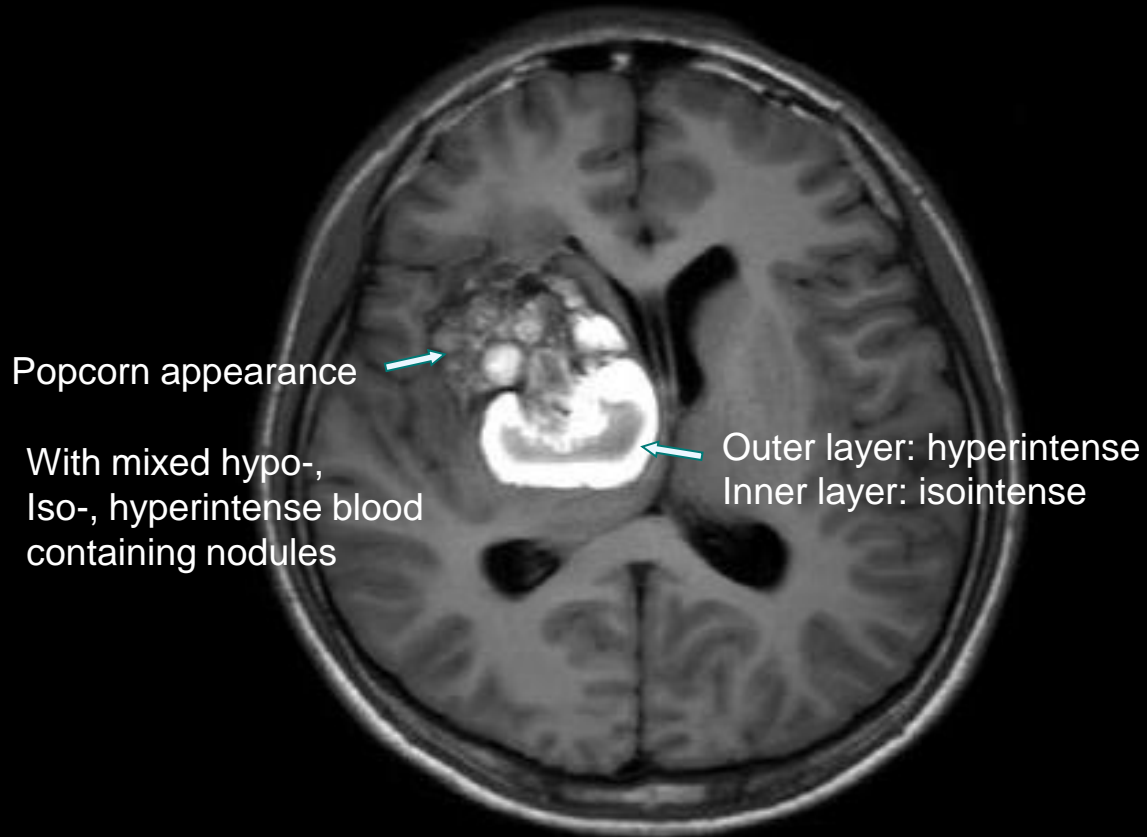
Location : Right basal ganglion

T1WC-

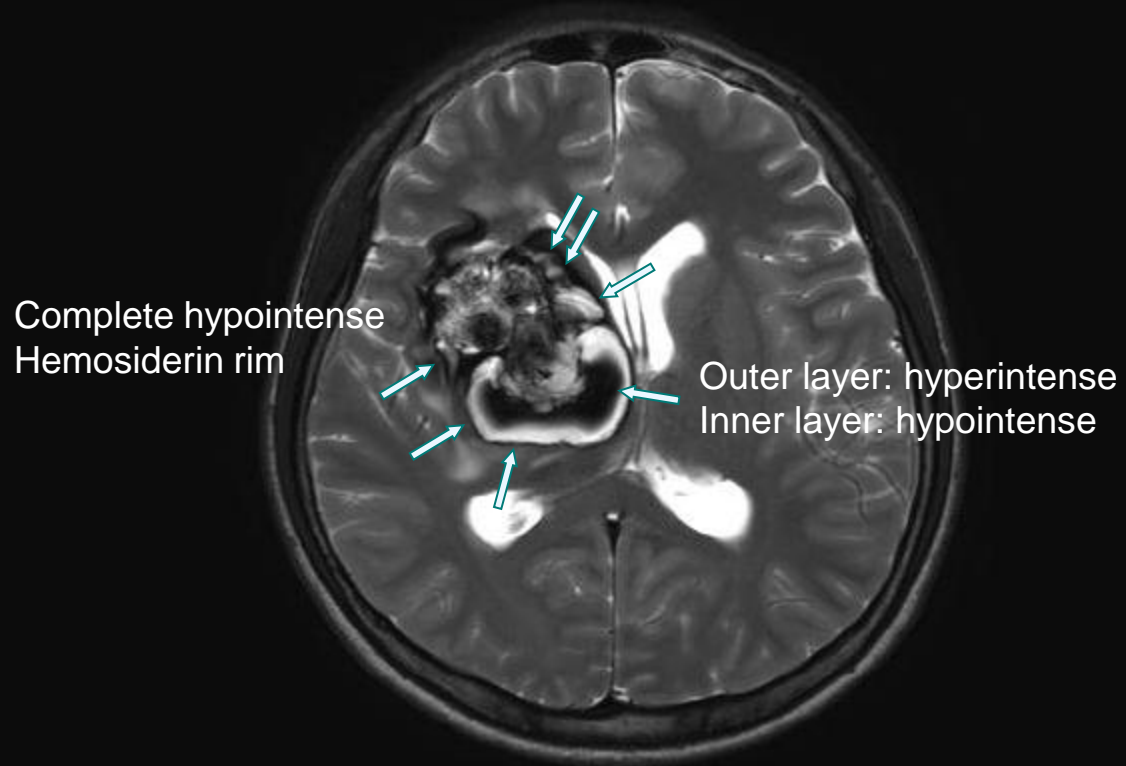


Hydrocephalus (-)
Midline deviation (+)

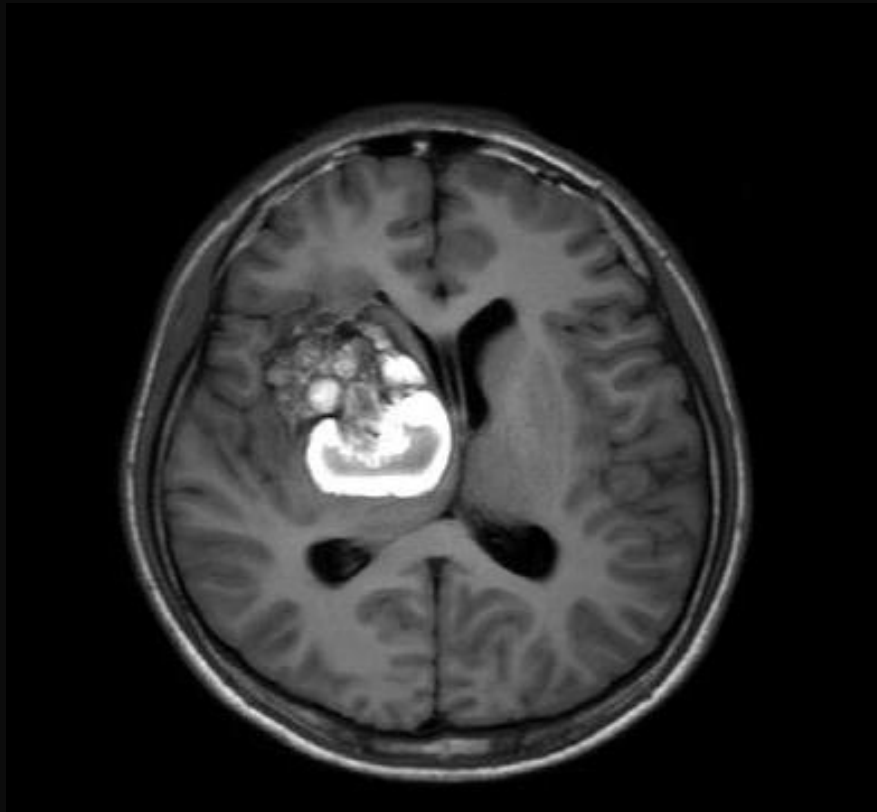
T1WC-



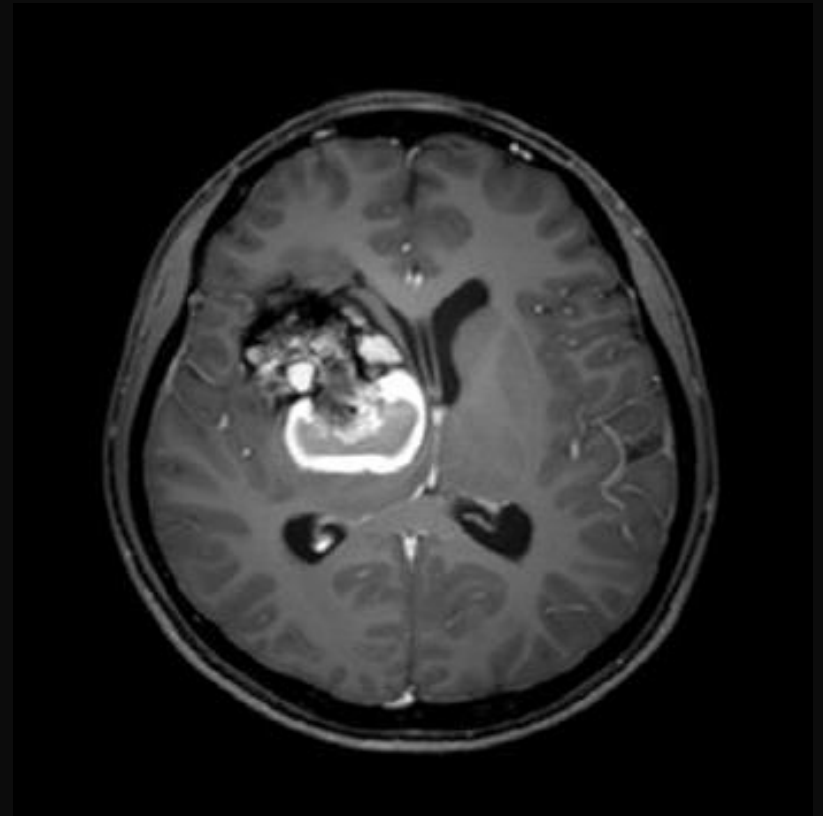
T2W



T1WC-



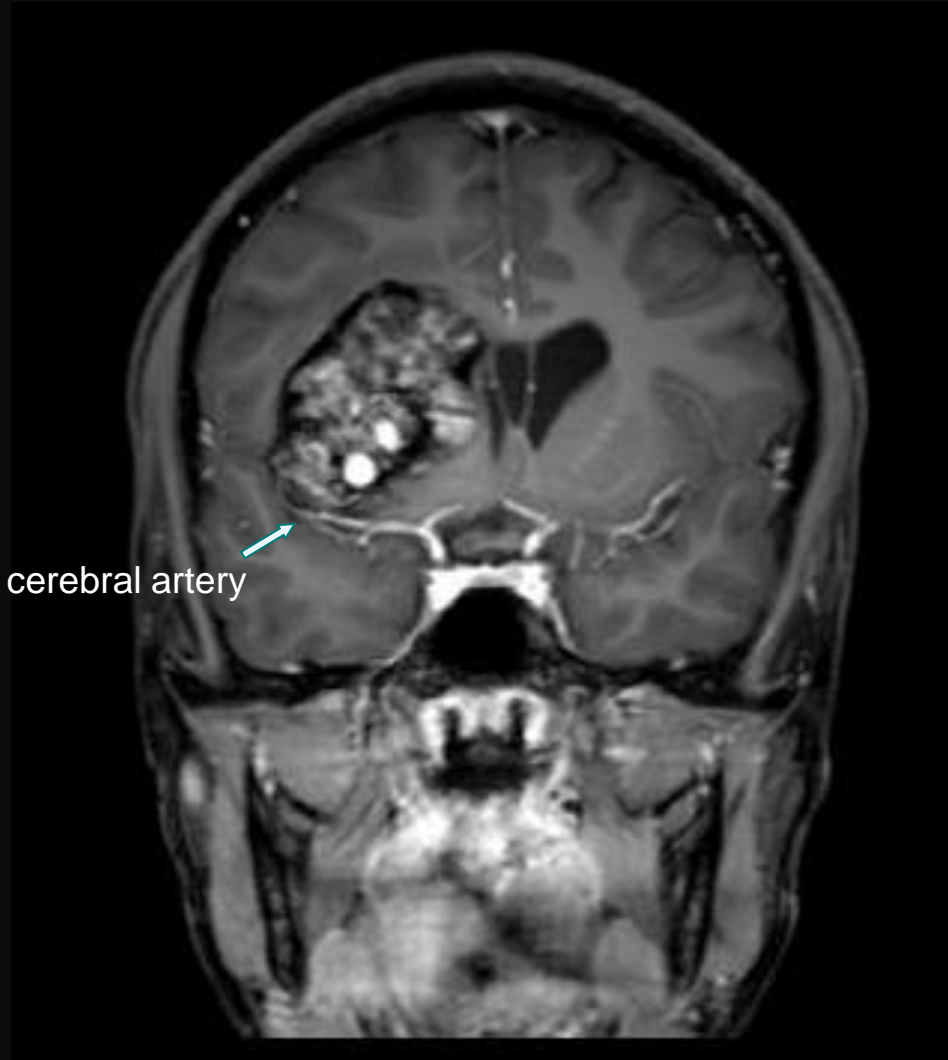
T1WC+



Minimal or no enhancement

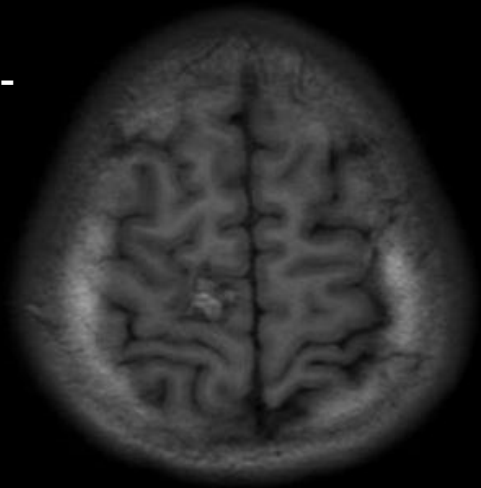
T1WC+

Right middle cerebral artery

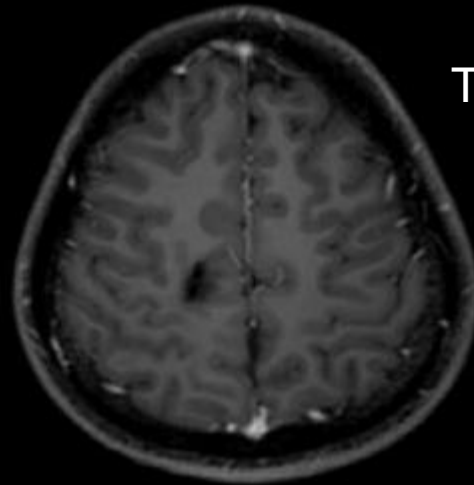


Another small lesion

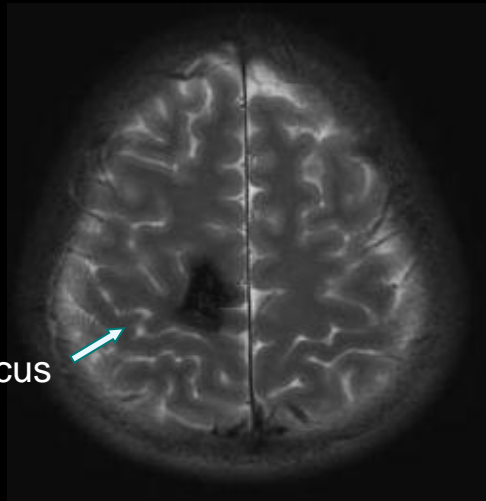
T1WC-



T1WC+

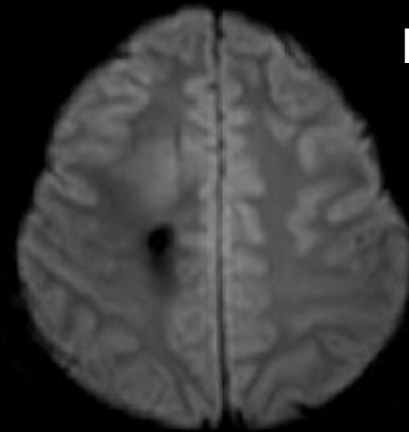


T2W

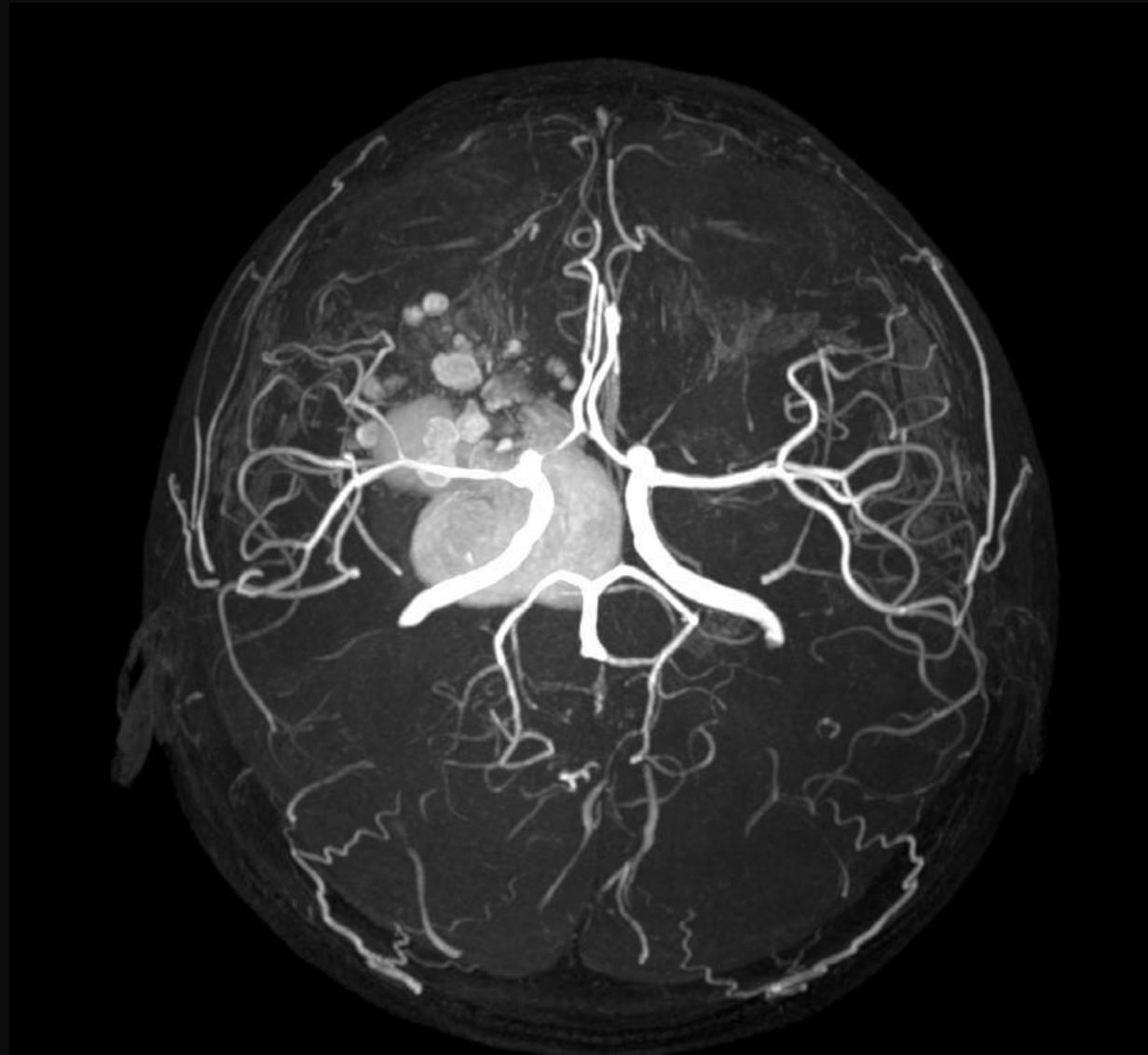


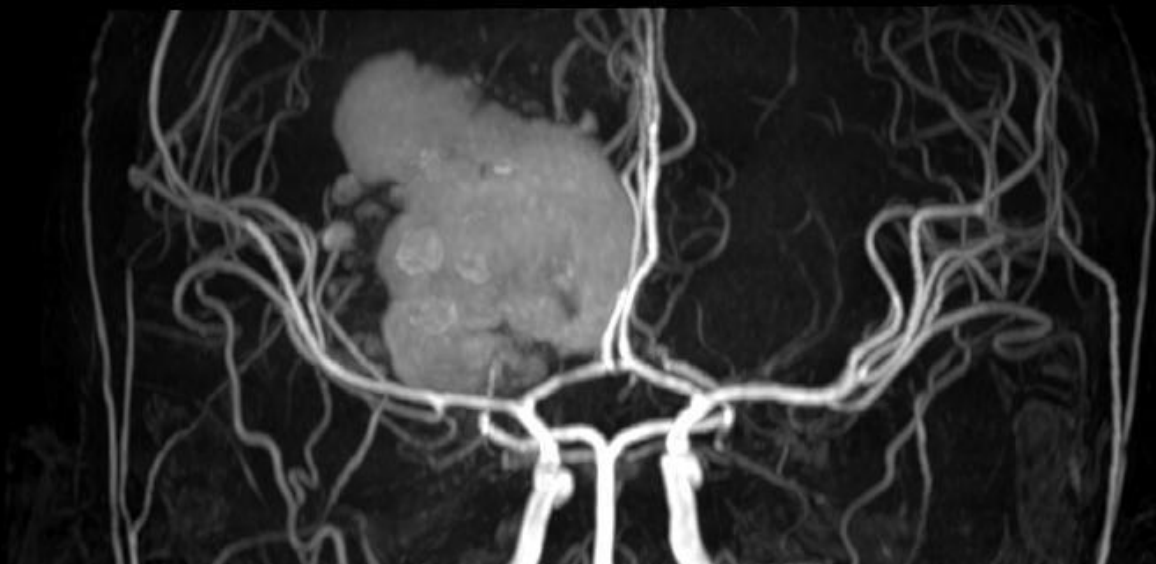
Central sulcus

DWI



MRA





R't ICA Lat.



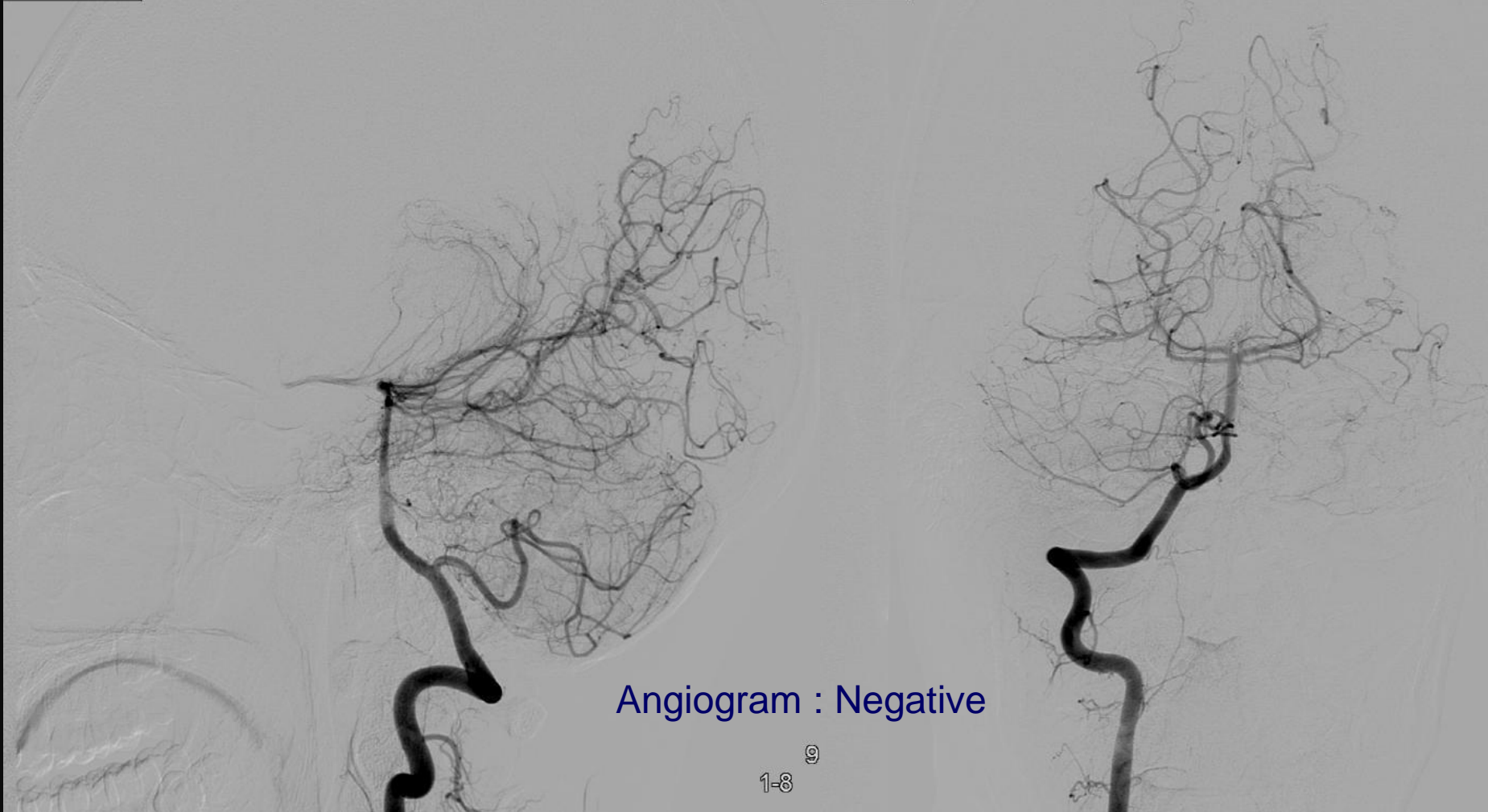
R't ICA



Angiogram : Negative

R't VA Lat.

R't VA



Angiogram : Negative

1-8⁹

Differential Diagnosis

Popcorn ball lesion:

1. Vascular malformation

- (a) Vascular malformation with AV shunting : Arteriovenous Malformation
- (b) Vascular malformation without AV shunting : Cavernous Malformation

Strong enhancement

Minimal/No enhancement

2. Hemorrhagic neoplasm

Strong enhancement

3. Calcified neoplasm (i.e. oligodendroglioma)

Moderate enhancement

Differential Diagnosis

Popcorn ball lesion:

1. Vascular malformation

- (a) Vascular malformation with AV shunting : Arteriovenous Malformation
- (b) Vascular malformation without AV shunting : Cavernous Malformation

98% solitary

75% solitary, 10-30% multiple, familial type

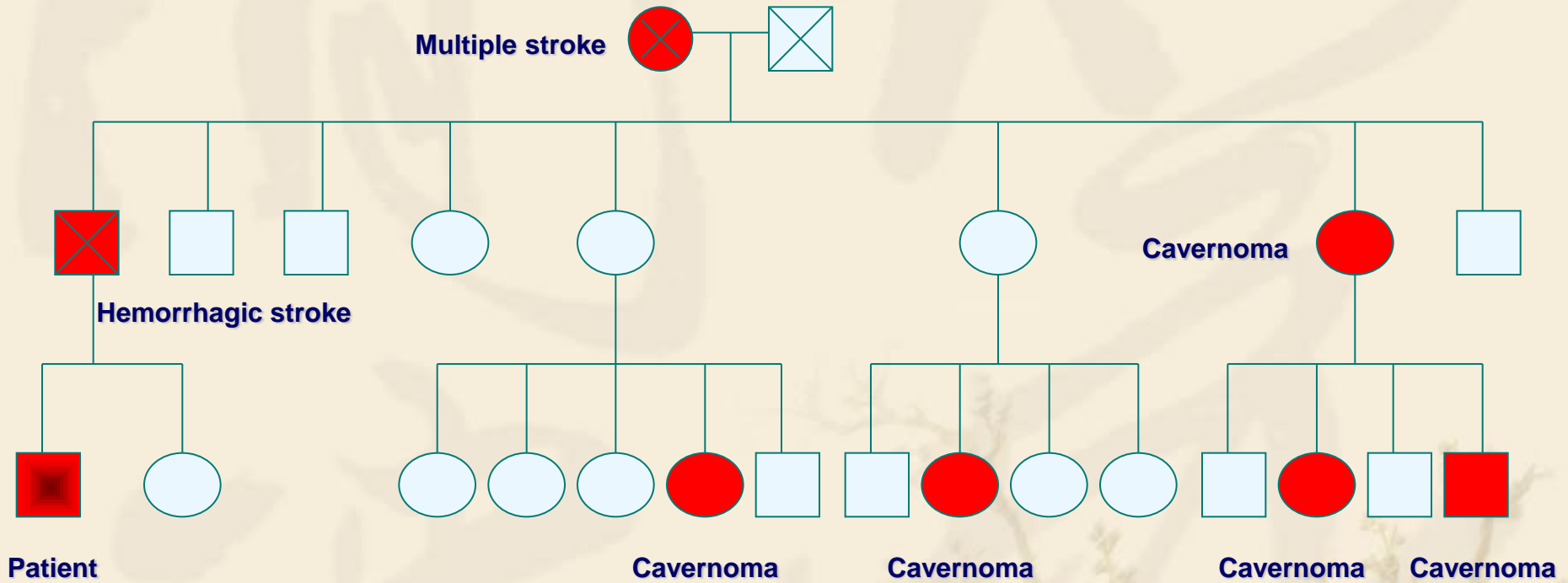
2. Hemorrhagic neoplasm

Almost solitary

3. Calcified neoplasm (i.e. oligodendroglioma)

Almost solitary

Family history of Cavernoma



Impression

1. Right basal ganglion vascular malformation with recent bleeding, suspect cavernous malformation, D/D Thrombosed Arteriovenous Malformation
2. Familial Cavernous Malformations
3. Asthma history and Preterm history



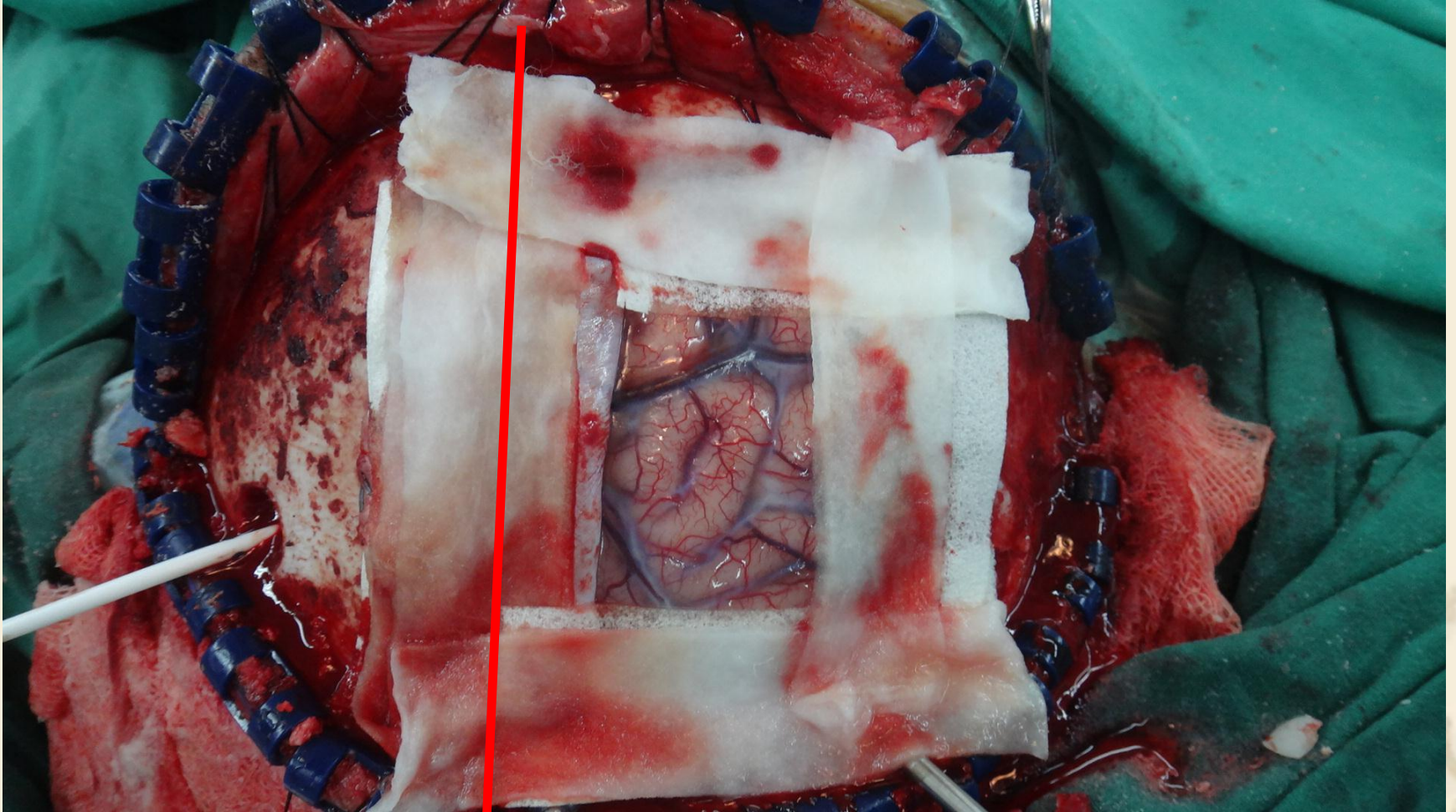
Surgery

Surgery

Op Methods:
Right frontal craniotomy with
tumor gross total removal under
navigation system



Surgery



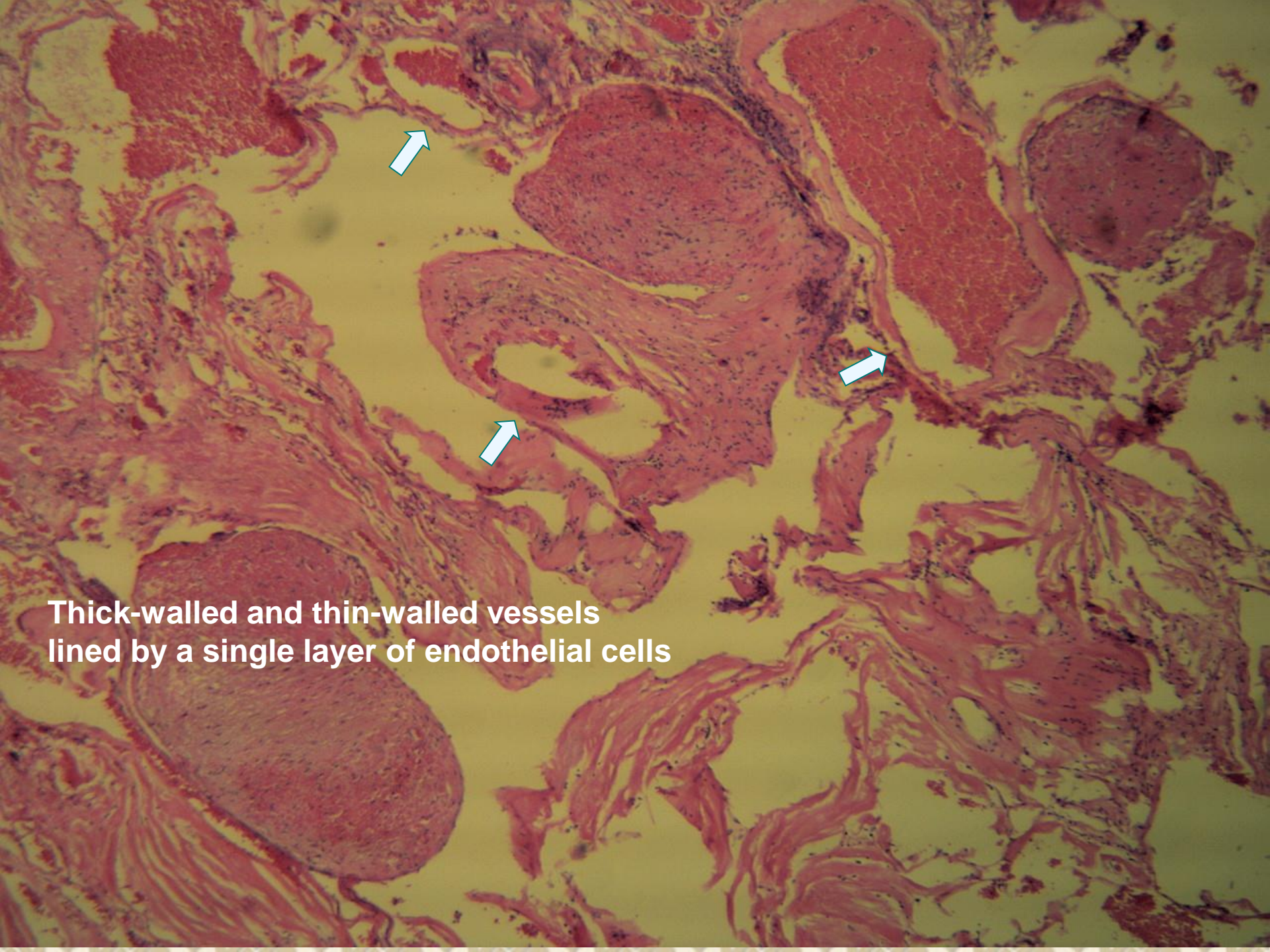
Midline

Post-op clinical course

- 9/03: Surgery
- 9/04: Extubation
- 9/08: Remove EVD
- 9/09: Transfer to ordinary ward
- 9/14: Post-op MRI
- 9/16: Discharge



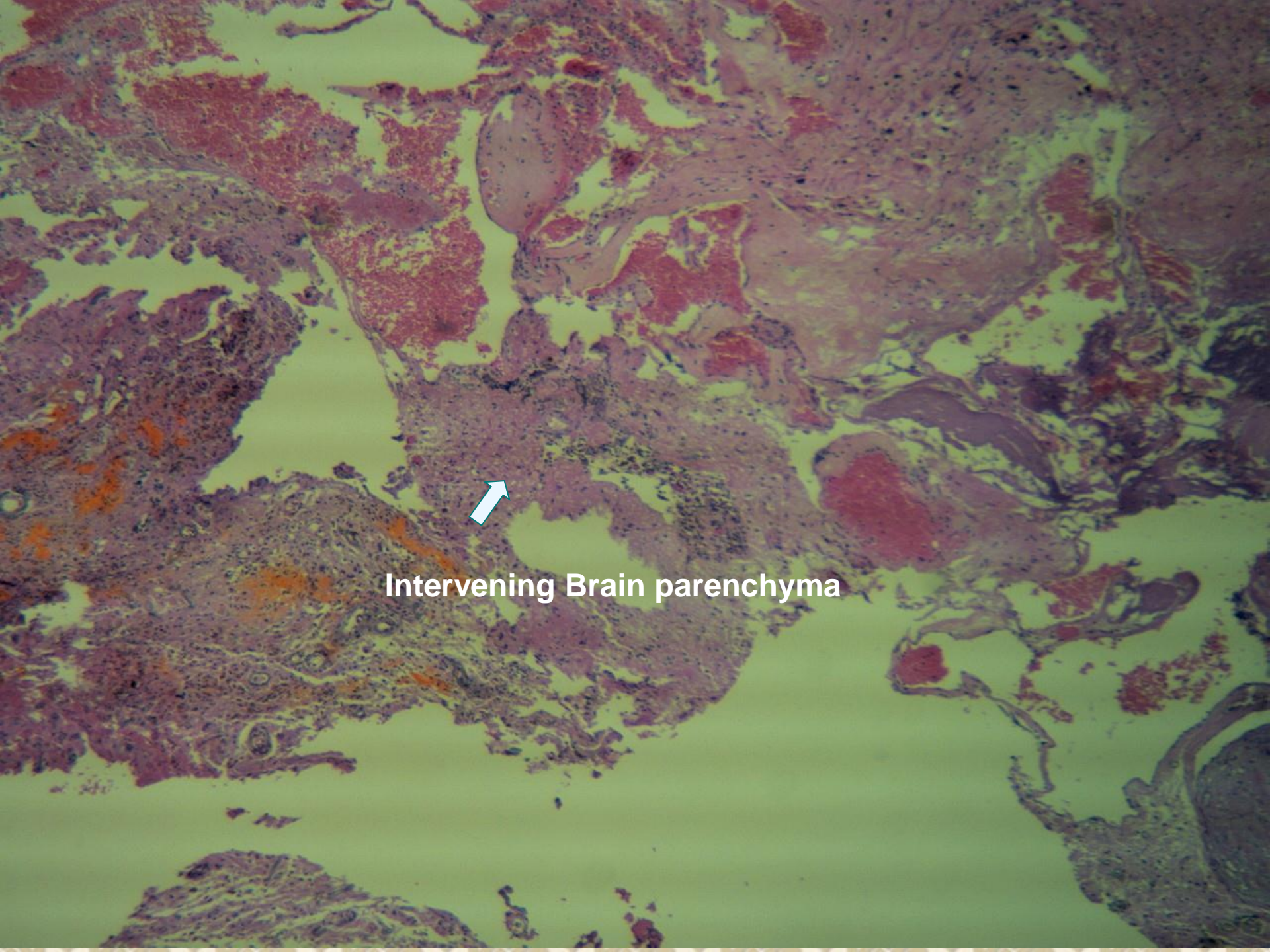
Pathology



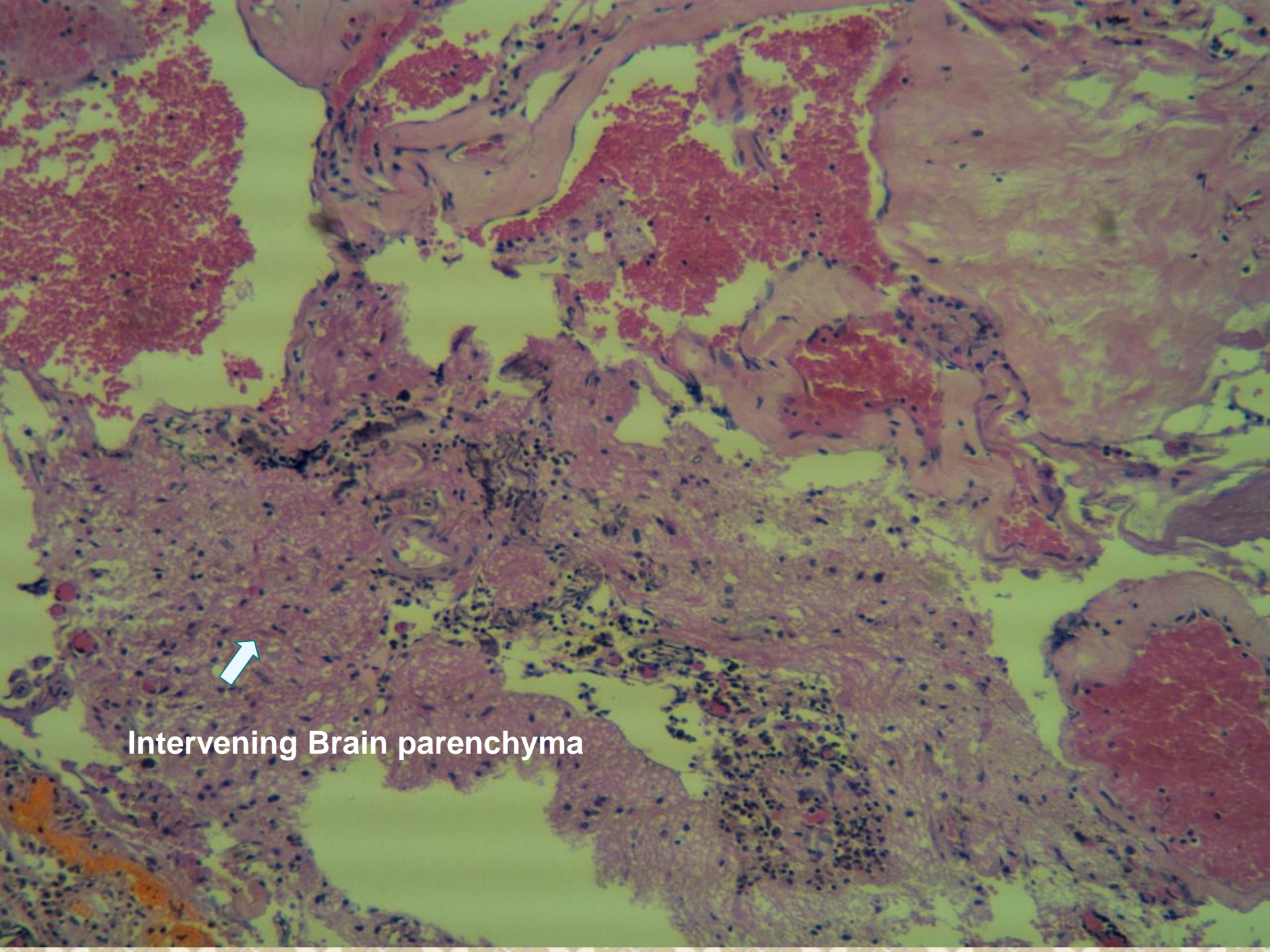
Thick-walled and thin-walled vessels lined by a single layer of endothelial cells



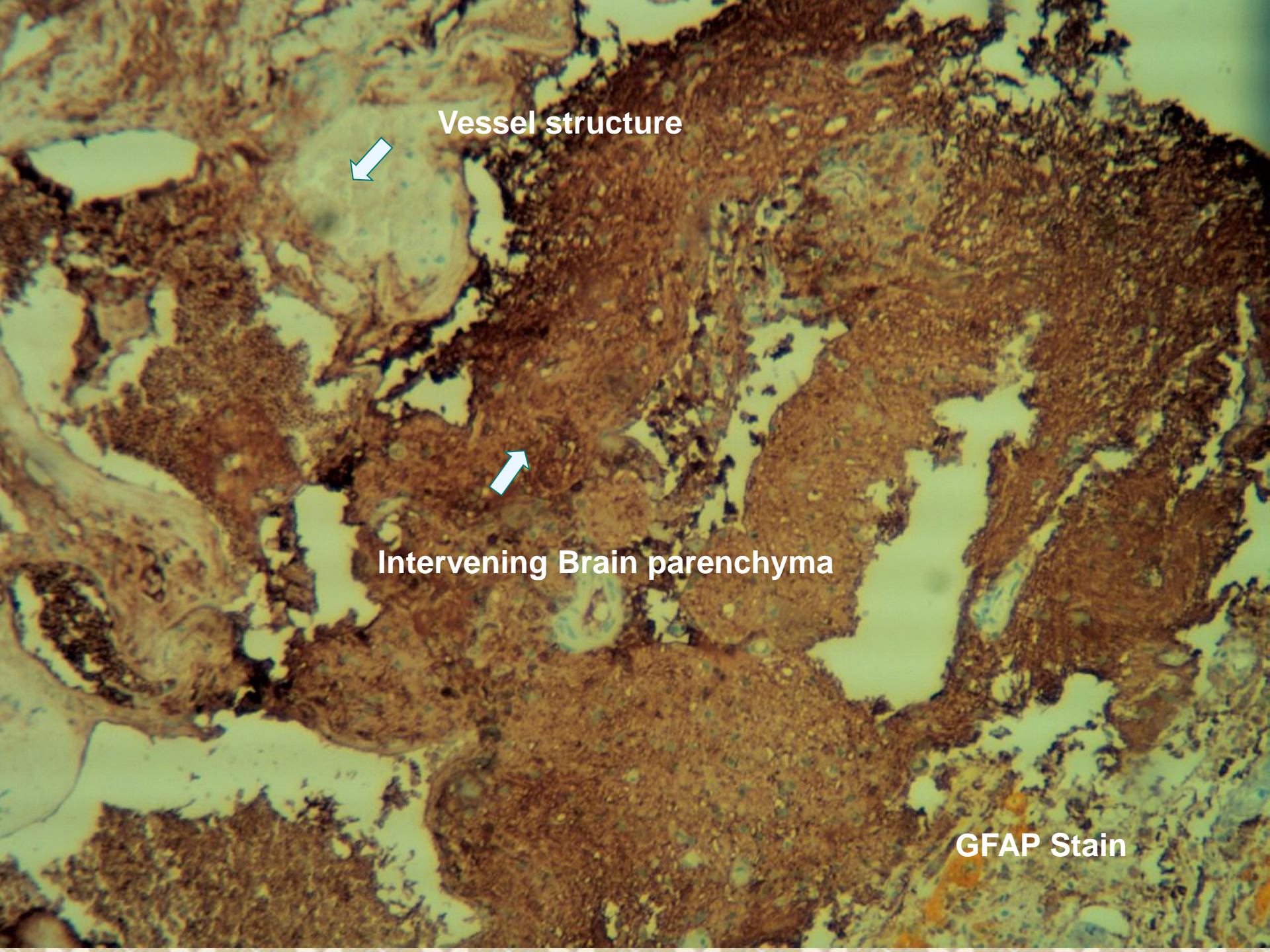
Thrombus



Intervening Brain parenchyma



Intervening Brain parenchyma



Vessel structure



Intervening Brain parenchyma



GFAP Stain



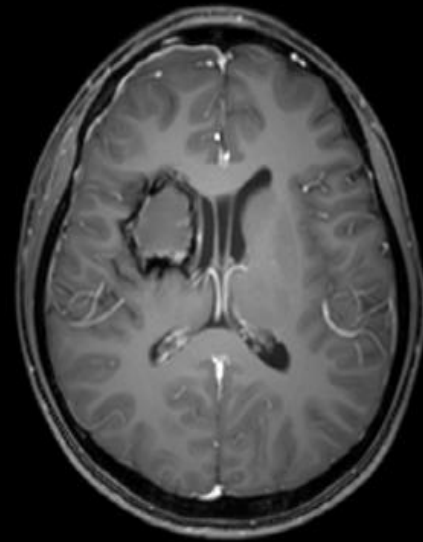
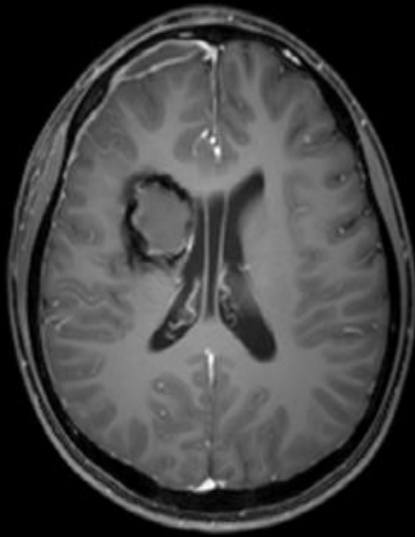
Post-op Image



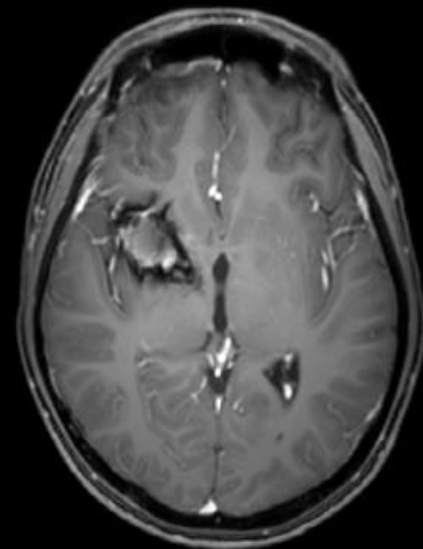
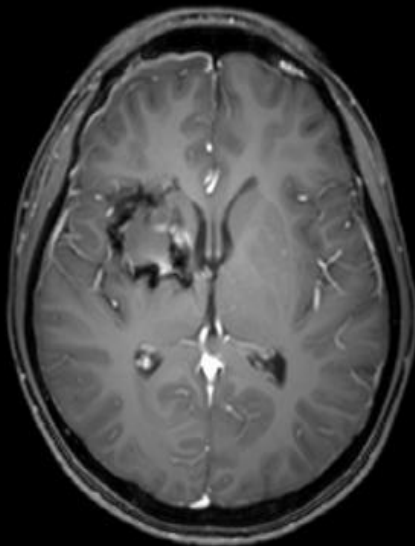


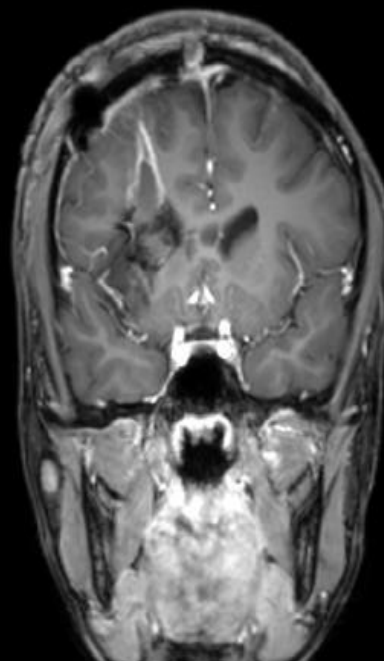
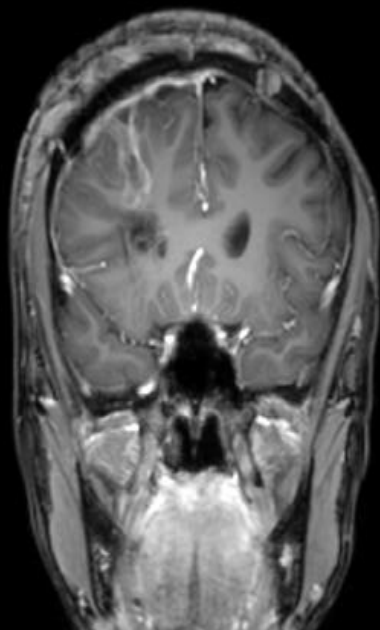
Post-op CT



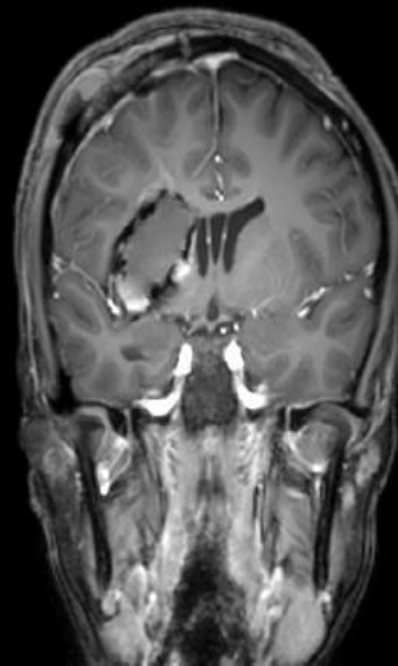


Post-op MRI





Post-op MRI





Discussion

Terminology of CNS vascular malformation

Vascular malformation with A-V shunting

1. Arteriovenous Malformation
 - Thrombosed AVM/Occult AVM
2. Arteriovenous Fistula
 - (1) Dural AVM/Dural AVF (dAVF)
 - (2) Vein of Galen Malformation (VOG)
 - (3) Carotid-Cavernous Fistula (CCF)

Vascular malformation without A-V shunting

1. Developmental venous anomaly (DVA)/Venous angioma
/venous malformation
2. Cavernous malformation/Cavernous angioma/Cavernous hemangioma
/Cavernoma/Capillary hemangioma
3. Capillary Telangiectasia
4. Sinus Pericranii

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Angiographically Occult
Vascular Malformation
(AOVM)

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graph TD; A["Thrombosed AVM/Occult AVM"] --> B["Angiographically Occult Vascular Malformation (AOVM)"]; C["Dural AVM/Dural AVF (dAVF)"] --> B; D["Vein of Galen Malformation (VOG)"] --> B;
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Vascular malformation without A-V shunting

1. Developmental venous anomaly (DVA)/Venous angioma /venous malformation

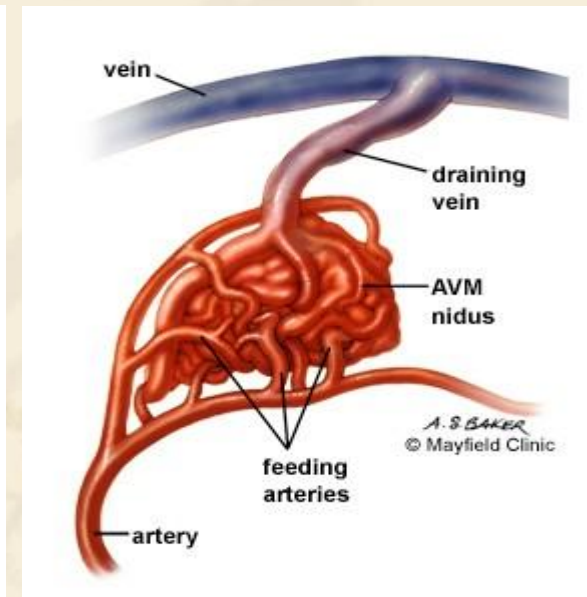
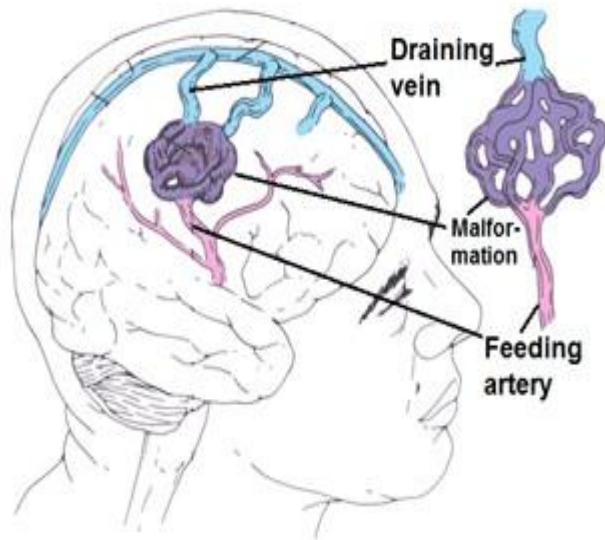
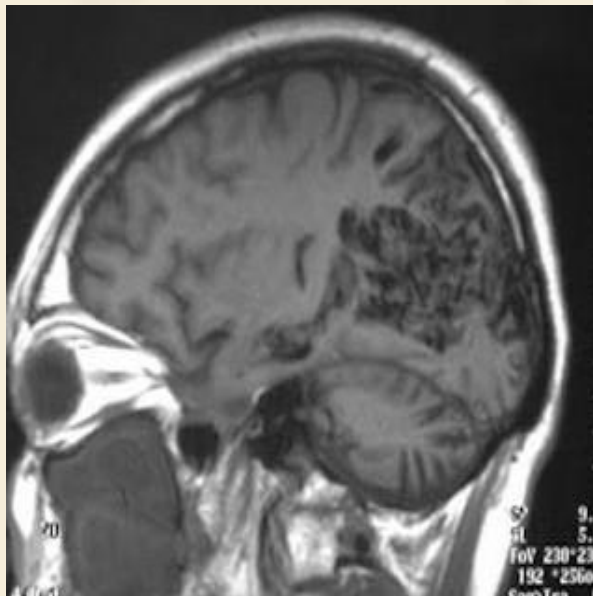
2. Cavernous malformation/Cavernous angioma/Cavernous hemangioma
/Cavernoma/Capillary hemangioma

3. Capillary Telangiectasia

4. Sinus Pericranii

Arteriovenous Malformation

- Definitions: Vascular malformation with arteriovenous shunting & no intervening capillary bed
- Best Diagnostic clue: Bag of black worm (flow voids)



Arteriovenous Malformation

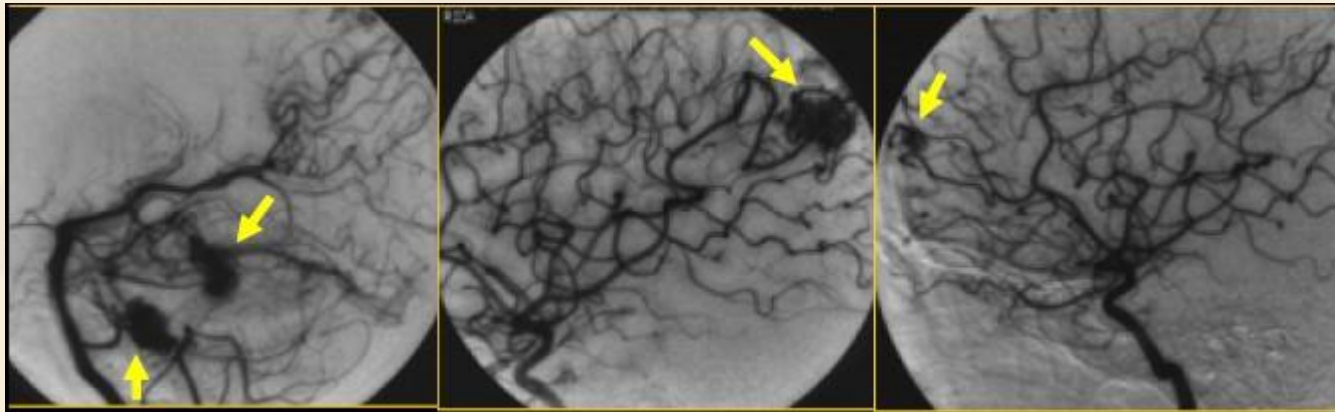
- 98% solitary

Multiple AVMS usually syndromic:

=> Hereditary hemorrhagic telangiectasia (HHT)

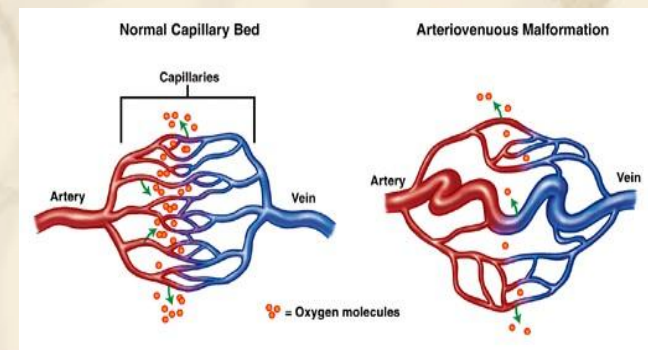
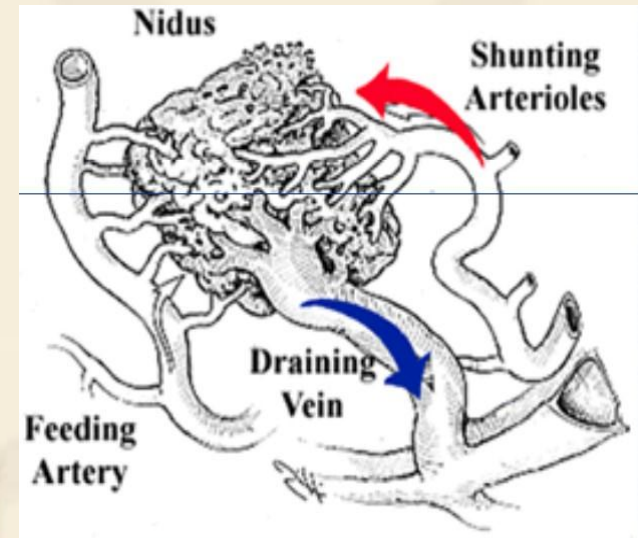
(遺傳性出血性毛細血管擴張症)

=/Osler-Weber-Rendu syndrome



Arteriovenous Malformation

- Occur anywhere in brain or spinal cord
85% supratentorial, 15% posterior fossa
- Peak age: 20-40 year old
Risk of hemorrhage: 2-4%/year
Spontaneous obliteration rare: < 1%
- Flow-related aneurysm on feeding artery: 10-15%
- Vascular “steal”: Ischemia in adjacent brain



Arteriovenous Malformation

- CT

- Variable Hemorrhage

- Calcification: 25-30%

- Enhance post-contrast

- CTA: Enlarged arteries & draining veins

- MRI

- Flow Voids: “Bag of worms”

- Variable hemorrhage

- T2: Increased signal - gliosis

- Contrast: Strong enhancement

- Minimal edema and mass effect



Arteriovenous Malformation

- Conventional Angiography

Best method of imaging

Must image ICA, ECA & vertebral circulations

27-32% of AVMs have dual arterial supply

Dural arterial supply via leptomeningeal anastomoses
or transdural anastomoses

Transdural anastomoses affects treatment decisions

Arteriovenous Malformation

- Hemorrhage annual rate

First bleed: 2 to 4% / year

Recurrent bleed: 6% Graf (JNS 1983; 58: 331) to
18% 1st year (Neurosurgery 1984; 15: 658)

Constant: 4% / yr Ondra (JNS 1966; 25: 467)

Calculating risk of bleed

Life time risk = $1 - (\text{risk of no hemorrhage})^{\text{expected years of remaining life}}$

Arteriovenous Malformation

➤ Spetzler Martin Grading system (1986)

Journal of neurosurgery 65:476,1986

Character		Points
Nidus (size in cm)	Small (<3 cm)	1
	Medium (3-6 cm)	2
	Large (> 6 cm)	3
Eloquent cortex	Yes	1
	No	0
Deep venous drainage	Yes	1
	No	0

AVM grade = sum (size + eloquence + deep component)

Arteriovenous Malformation

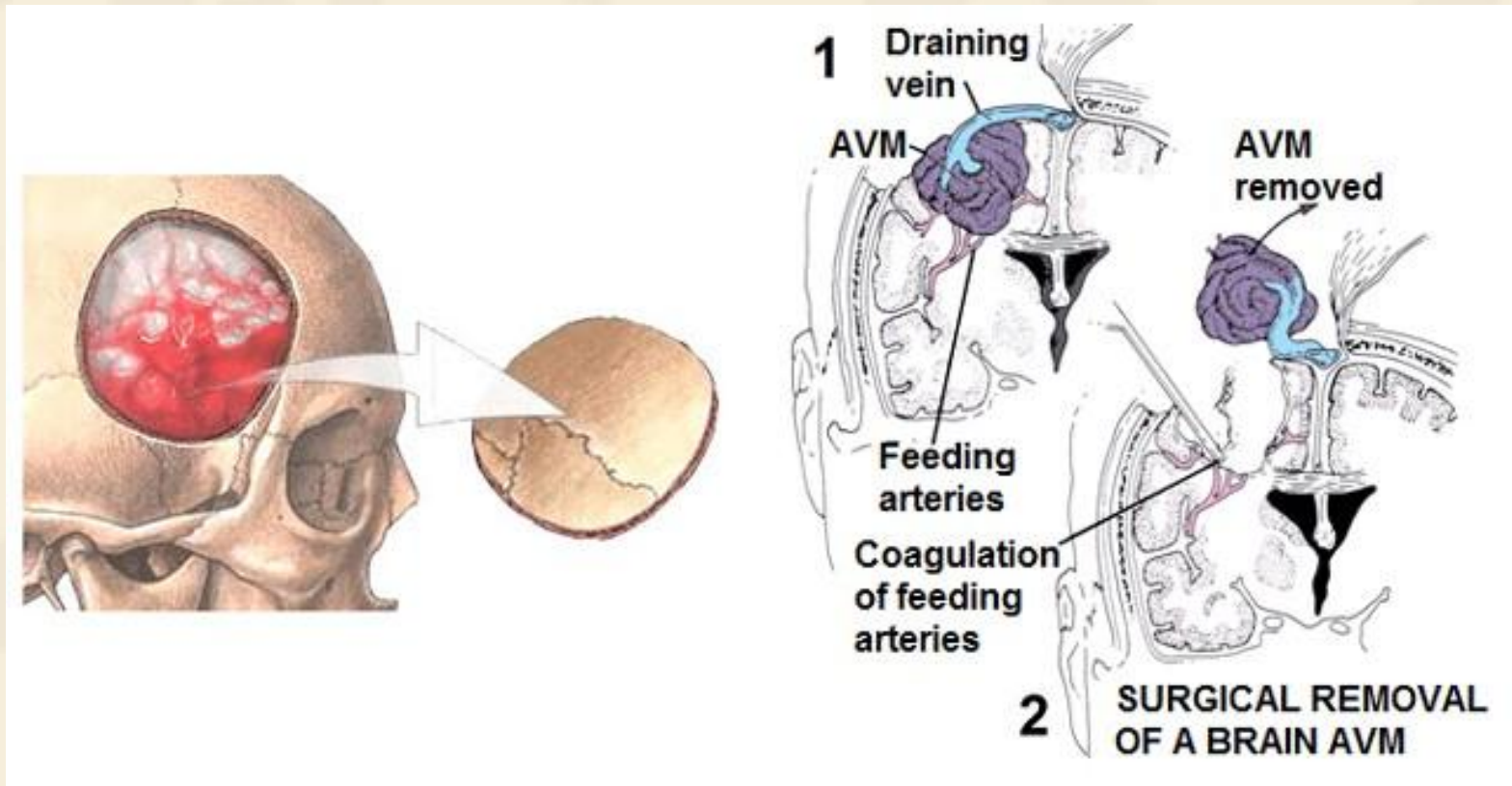
➤ Spetzler Martin Grading system

Risk of surgery *Spetzler and Martin 1986; Heros et al 1990*

Grade	Minor Deficits	Major Deficits	Favorable outcome
I	0	0	92- 100%
II	5%	0	95%
III	12%	4%	88%
IV	20%	7%	73%
V	19%	12%	57%

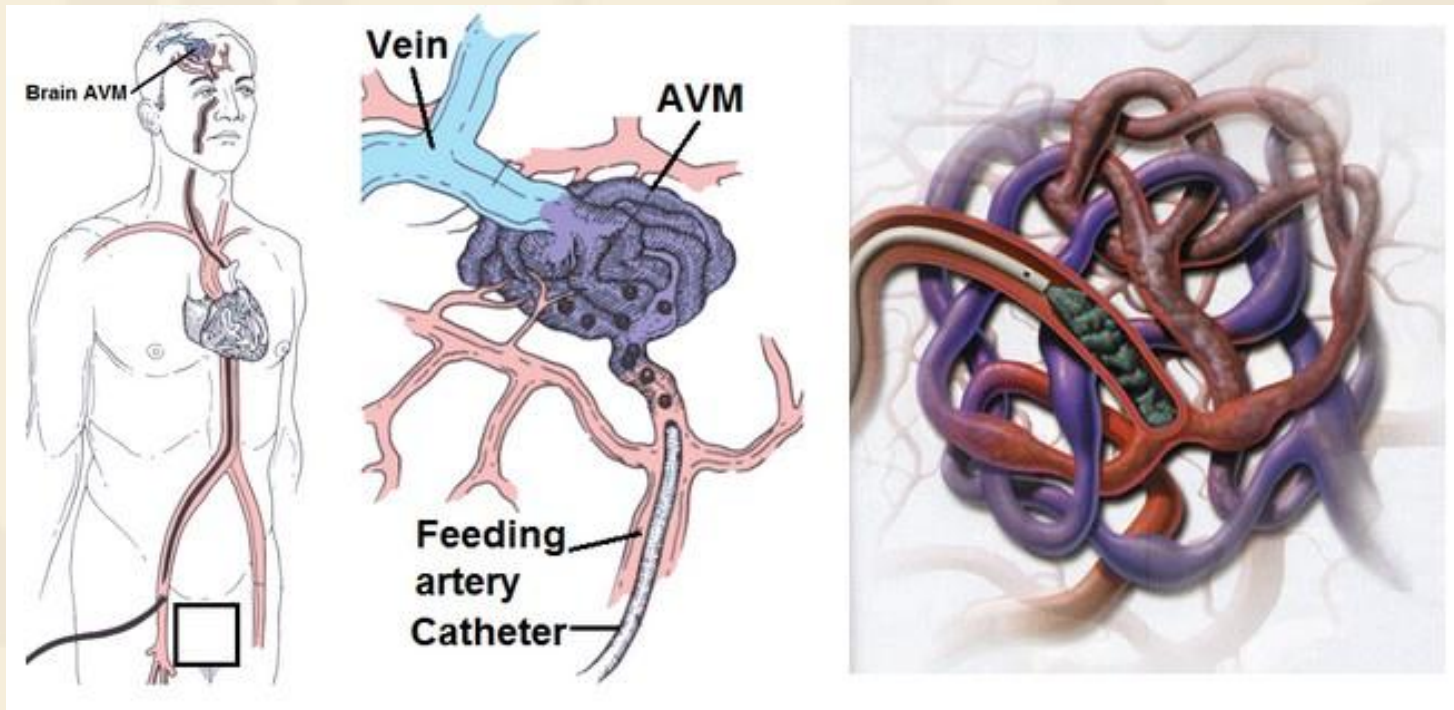
Arteriovenous Malformation

- Treatment



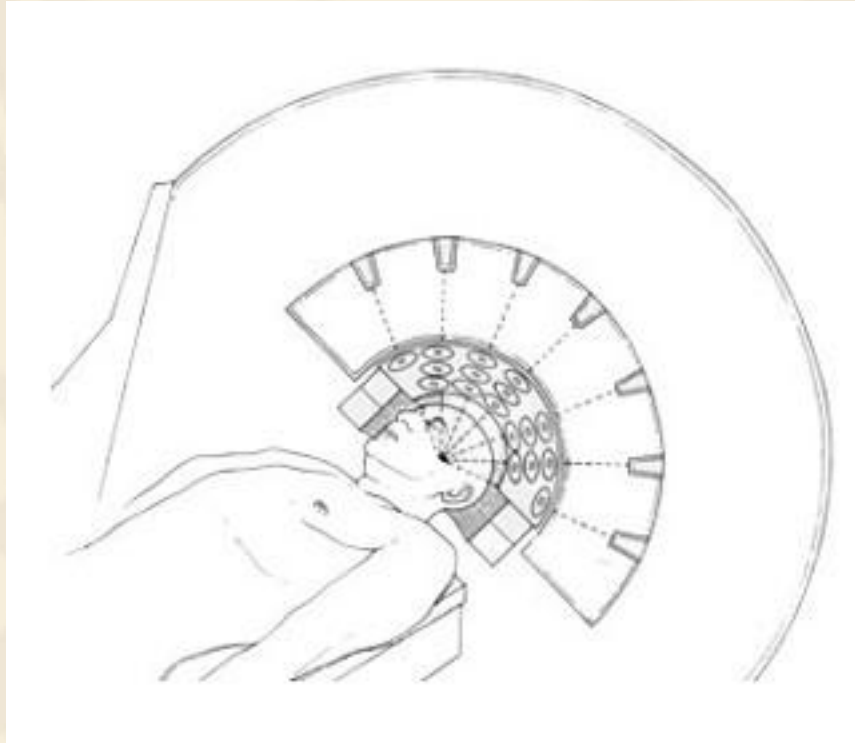
Arteriovenous Malformation

- Treatment



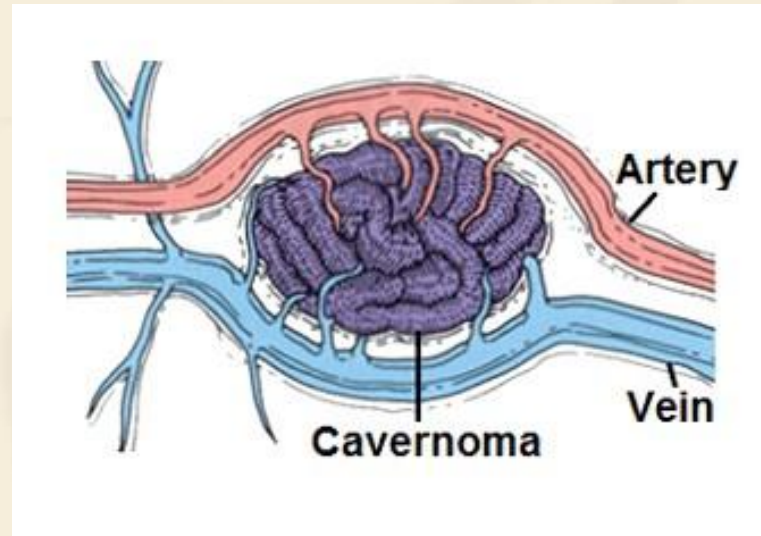
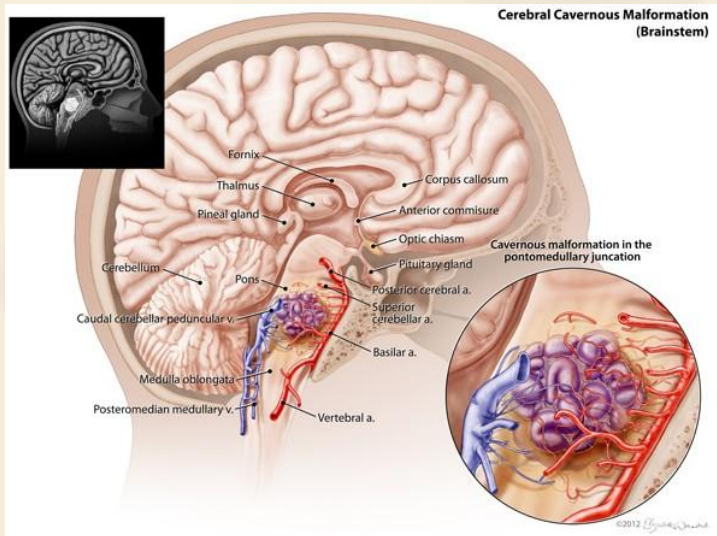
Arteriovenous Malformation

- Treatment



Cavernous malformation/Cavernous

- **Definitions:** Benign vascular hamartoma with masses of closely apposed immature endothelial-lined, hemorrhage-filled vessels (caverns) without intervening normal brain



Cavernous malformation/Cavernous

- Best Diagnostic clue: Popcorn ball appearance with complete hypointense hemosiderin rim on T2W MR

- 10-30% multiple

Multiple (familial) cavernous malformation is autosomal dominant, variable penetrance

Three separate loci implicated (CCM1, CCM2, CCM3 genes)

Cavernous malformation/Cavernous

- Nature History

Broad range: may progress, enlarge, regress

Propensity for growth via repeated intralesional hemorrhage

Rehemorrhage rate high initially, decrease after 2-3 years

Familial CMs had high risk for hemorrhage

1% per lesion per year

- Treatment

Total removal via microsurgical resection

If mixed DVA, venous drainage must be preserved

SRS limited effectiveness

Capillary telangiectasia

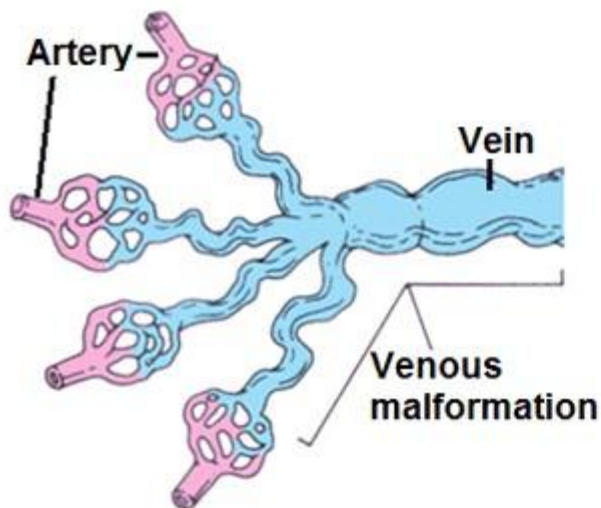
Capillary telangiectasia



Capillary telangiectasia consists of groups of dilated capillaries similar to those seen in the legs of many people. They rarely cause damage to neighbouring tissue and haemorrhages are exceptional.

Brain telangiectasia

Venous malformations



Venous malformations involve hypertrophied veins draining blood from neighbouring nervous tissue without interfering with its function. They rarely bleed and the vast majority of them are asymptomatic and benign. They cannot be removed because doing so usually damages the nervous tissue on which they sit.

The image features a traditional Chinese ink wash painting of a plum blossom branch. The branch is dark and gnarled, with small, delicate flowers in various stages of bloom. The background is a light, textured paper. A large, faint watermark of the Chinese characters '福山' (Fúshān) is visible in the background. The text 'Thanks for your Attention' is overlaid in the center in a bold, blue font. The image is framed by a decorative border at the top and bottom, consisting of a repeating geometric pattern of triangles and circles in a gold color.

Thanks for your Attention