General Data

- Gender: Female
- Birthday and age: 1932/11/03, 73 y/o
- Occupation: house keeper
- Date of Admission: 2005/03/30

Chief Complain

- Dizziness and light headache for recent 1 year.

Present illness

- Hypertension with regular medication for 1~2 years.
- The patient took drugs for hypertension and dizziness from LMD for 1 year.
- The patient had fainting spells.
- Recently, complaining of light-headache and dizziness.
- No trauma history.

Past Hx and Personal Hx

- Previous Admission and Operation history : denied
- Hypertension: under control for 1~2 years
- Smoking and Alcohol: denied

Physical examination

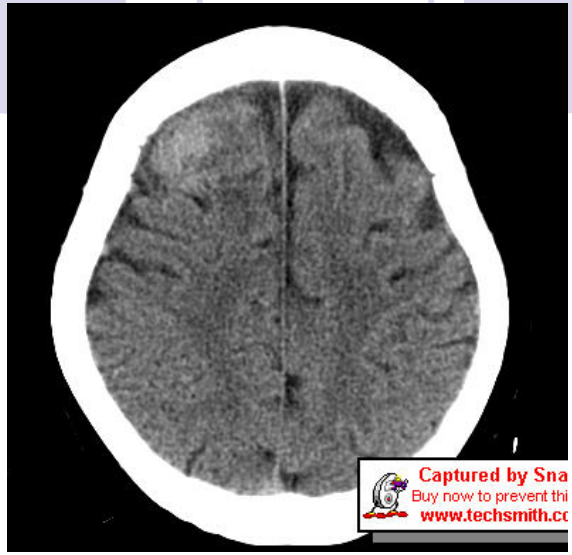
- Consciousness : clear
- Vital signs : TPR : 36.3/72/20 ,
  BP : 154/80 mmHg
  G.A : weakness，chronic ill-looking
- Chest : breathing sound clear
- Heart : RHB without murmur
- Abdomen :soft and flat
- Neurological Examination: intact

Lab Data

- RBC [4.2-6.1 x10.e6/uL] : 4.12
- MCH [27-31 pg] : 31.6
- Others: no positive findings

Image-Brain CT focus on Routine (3/26)

- There is precontrast subtle hydensity noted at Rt frontal-lobe.

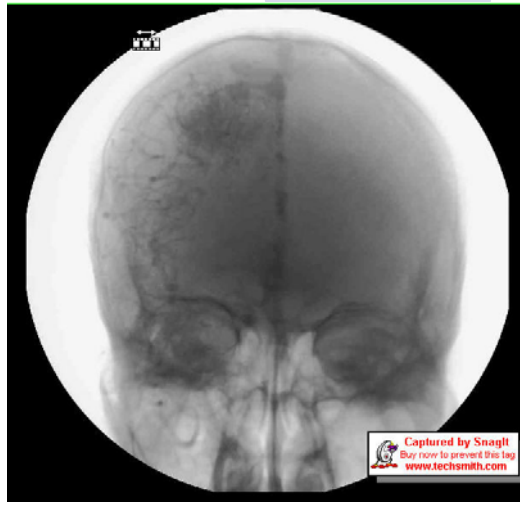


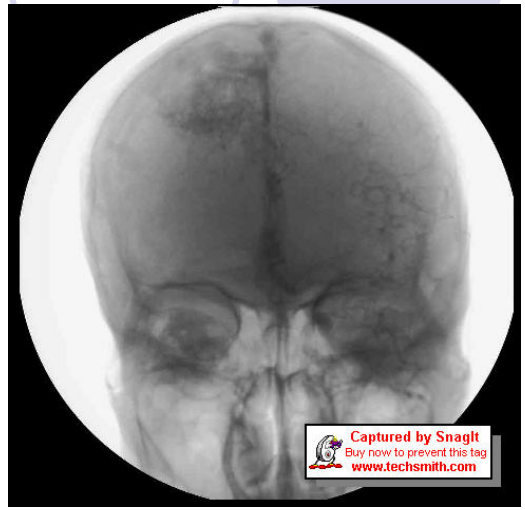
- It is strong enhancement with small and engorge vascularity, measuring about 6x3.4x2.8cm in largest dimension.

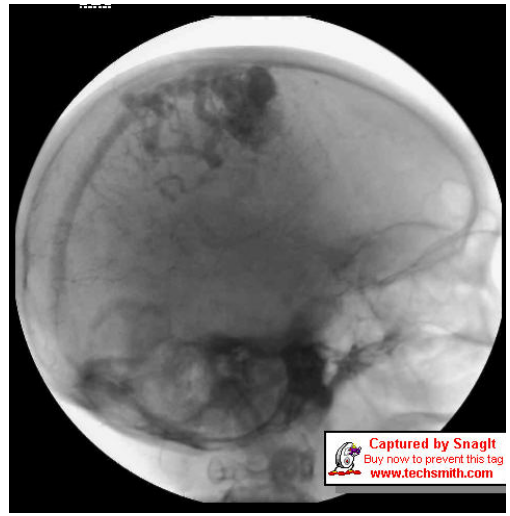
- Impression：
  Mass with hypervascular networks at Rt frontal-lobe.
  R/O Parenchymal AVM.

R+L Carotid angiography and Verebral angiography (1)

- A hypervascular lesion is noted at the right frontal region, the supplied artery is right ACA.
- Evident engorged and enlarged right anterior cerebral artery with early venous return into the superior sagital sinus is noted.
- No mid-line structure deviation.





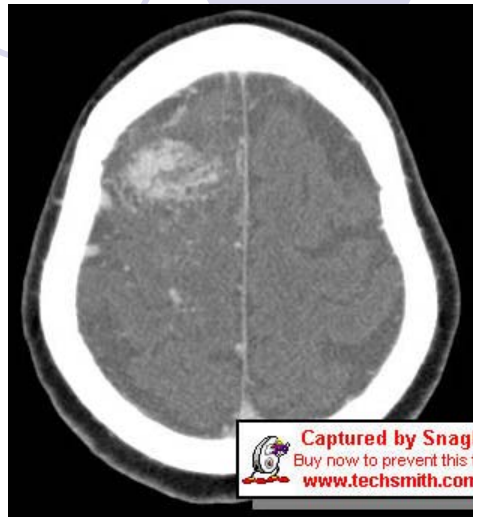


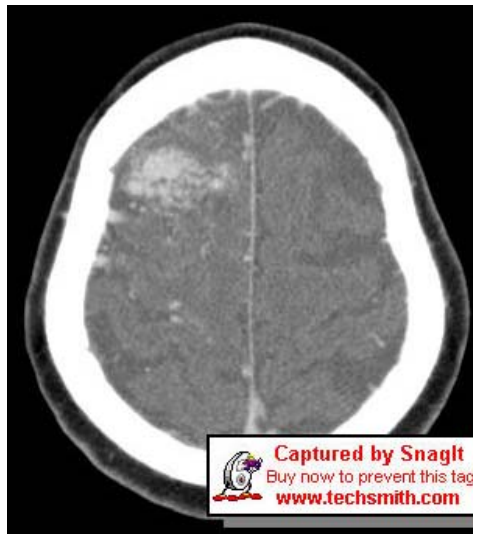
R+L Carotid angiography and Verebral angiography (2)

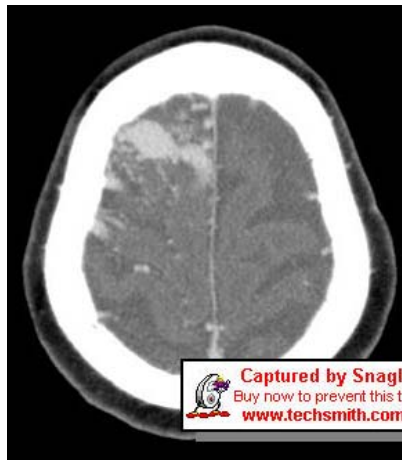
- Impression：Right frontal AVM is supplied by the enlarged right anterior cerebral artery.

Brain CT focus on Routine (4/06)

- A large irregular enhancing mass with engorged venous structure at the right frontal lobe measuring 3 cm in largest diameter.
- The cerebral ventricles are of normal size and symmetrical arranged.
- Impression：
  Right frontal AVM CT localization.







Differential Diagnosis

- Brain, Arteriovenous Malformation
- Brain, Aneurysm
- Brain, Capillary Telangiectasia
- Brain, Cavernous Angiomas

Arteriovenous Malformation(1)

- Tangled cluster of vessels in which arteries connect directly to veins with no intervening capillary bed.
- The lesion may be compact, containing a core of tightly packed venous loops.
- It may be diffuse, with anomalous vessels dispersed among normal brain parenchyma.

Arteriovenous Malformation(2)

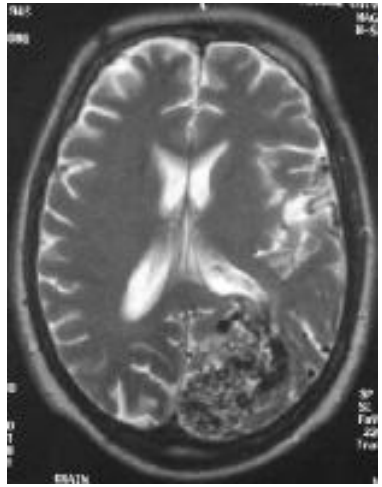
- CT imaging of a brain AVM can show an isoattenuating-to-hyperattenuating hemispheric mass.
- Absence of hemorrhage:
  nonenhanced CT can demonstrate small foci of calcification in as many as 30% of patients.

- Other finding: cystic cavities representing previous hemorrhage and hypoattenuating of surrounding parenchyma representing encephalomalacia, cerebral atrophy, or gliosis.
- **Contrast CT** can demonstrate serpiginous vascular enhancement uniquely typical of an AVM.
- CT can demonstrate edema, mass effect, or ischemic changes that can be associated with an AVM.
- An AVM in the chronic stage of intracerebral hemorrhage appears hypoattenuating relative to normal brain tissue.

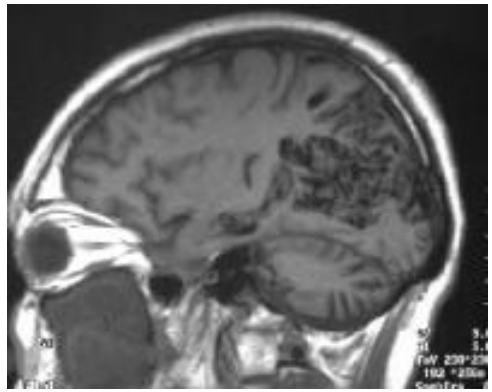
Arteriovenous Malformation(3)

- MRI:

  Typical unruptured AVM appears as a tightly packed or loose tangle of vessels.
- Rapid blood flow through enlarged arteries causes a signal or flow void on routine spin-echo T1- and T2-weighted images. This finding is uniquely characteristic of an AVM.



Axial T2-weighted MRI shows numerous flow voids. (Note the mass effect on the lateral ventricle despite the lack of a mass or hemorrhage.)



Sagittal T1-weighted MRI demonstrates a large occipital AVM with parasagittal flow voids.

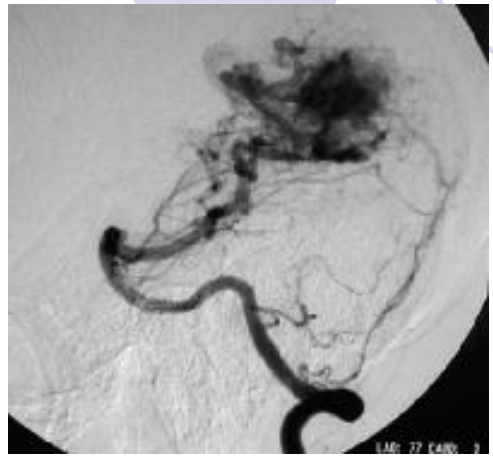
Arteriovenous Malformation(4)

- Conventional cerebral angiography is the criterion standard for the evaluation of AVMs.
- Goal:

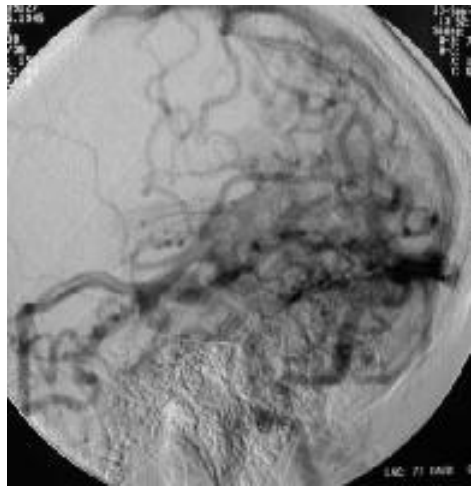
1.Identify the number and location of feeding arteries.

2.The angiographic location and size of the nidus.

3.The shunt type of the lesion, and the pattern of venous drainage.



Lateral left vertebral angiogram demonstrates a huge, left posterior cerebral artery feeder to the nidus



Venous phase of a vertebral angiogram demonstrates numerous superficial and deep draining veins.

Brain, Aneurysm(1)

- An abnormal dilatation of an artery.
- Intracranial aneurysms are classified into saccular and nonsaccular types:
  - Nonsaccular aneurysms- include atherosclerotic, fusiform, traumatic, and mycotic types.
  - Saccular(berry) aneurysms- have several anatomic characteristics that distinguish them from other types of intracranial aneurysms.

Brain, Aneurysm(2)

- Finding often supported by the demonstration of an aneurysm in the area of maximum clot localization or maximum amount of subarachnoid blood.
- ***Fisher grading system*** is used to classify SAH:

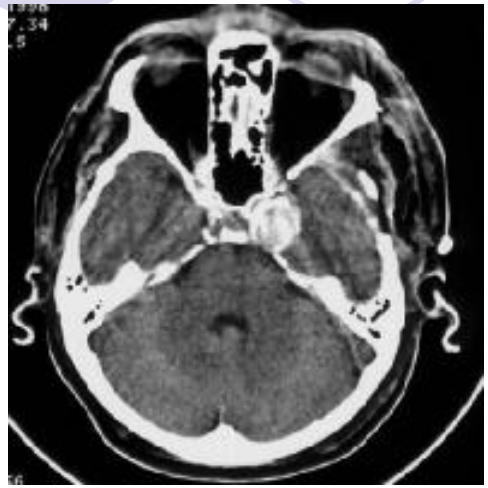
Grade/Signs

1/No subarachnoid blood detected

2/Diffuse vertical layers thicker than 1 mm

3/Localized clot and/or vertical layer thicker than 1 mm

4/Intracerebral or intraventricular clot with diffuse or no subarachnoid blood



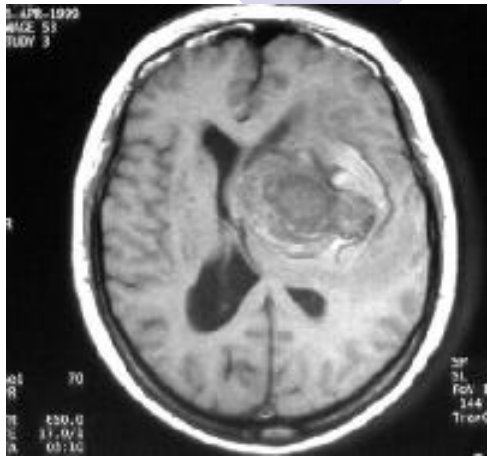
- Nonenhanced CT scan:

a giant aneurysm of the left internal carotid artery in its intracavernous segment. This aneurysm is densely calcified.

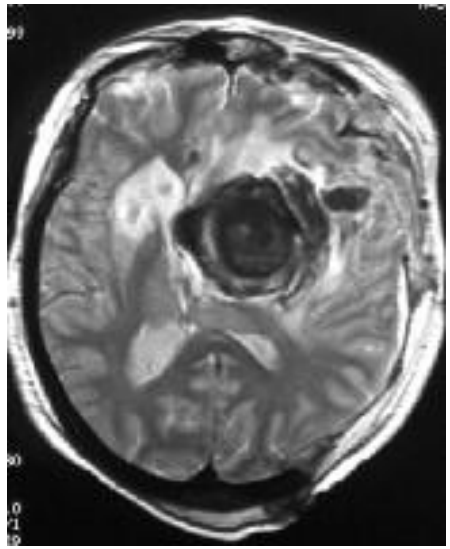
Brain, Aneurysm(3)

MRI:

- Intracranial aneurysms: an area of flow void larger than the healthy vessels in that region.
- Giant aneurysms: calcifications and an intraluminal clot.



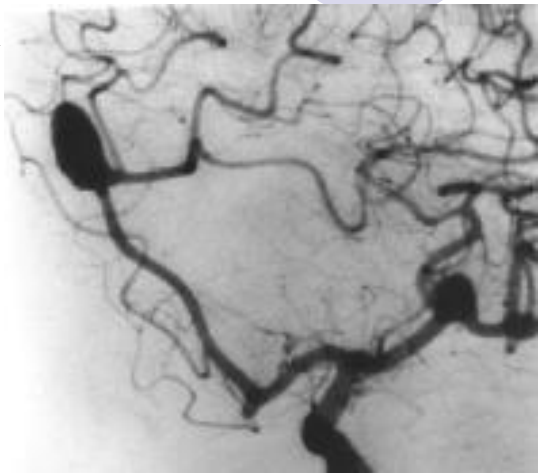
T1-weighted MRI : A large intracerebral mass with a significant amount of surrounding edema is depicted. (giant internal carotid artery aneurysm)



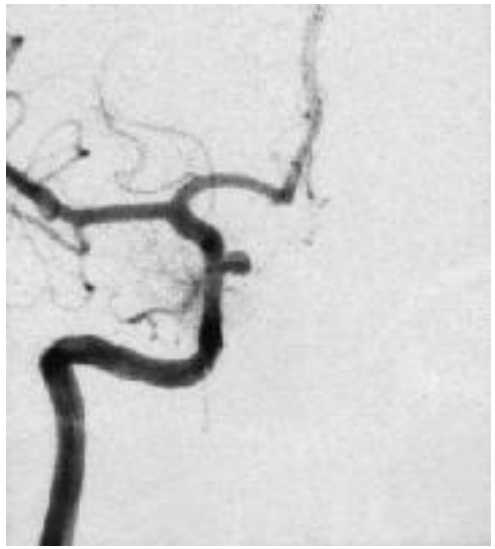
T2-weighted MRI: Note the flow void, the blood breakdown products within the layers of mural thrombus, and calcification within the aneurysm that produces a marked hypointense signal.

Brain, Aneurysm(4)

- Cerebral angiography remains the definitive preoperative diagnostic tool in patients with intracranial aneurysms.



Left oblique cerebral angiogram: an ACOM aneurysm and a middle cerebral artery aneurysm.



Left oblique cerebral angiogram: a proximal intracranial internal carotid artery aneurysm

Capillary telangiectasias(1)

- Capillary telangiectasias (CTSs) are small areas of abnormally dilated capillaries within otherwise normal brain tissue.
- CTSs have been associated with minor symptoms such as vertigo, headache, and dizziness, as well as weakness and seizures.

Capillary telangiectasias(2)

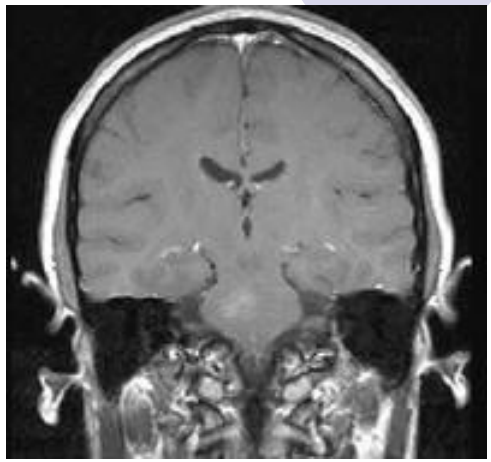
CT

- Nonenhanced CT:

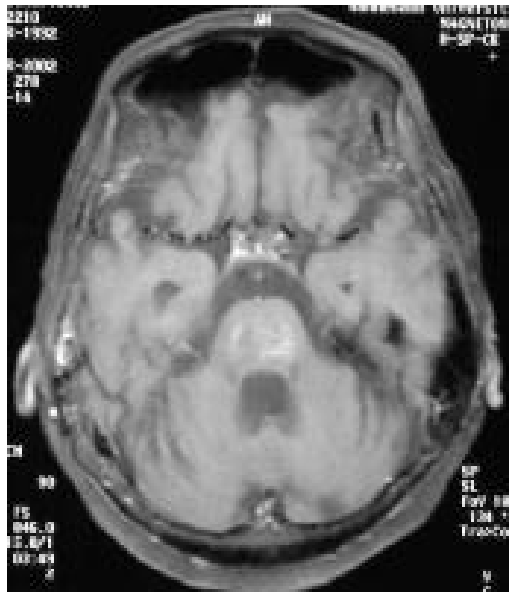
1. do not depict CTS
2. lesions are not visible even after the administration of contrast material.

Capillary telangiectasias(3)

- MRI findings in CTS are variable.
- The enhancement pattern is described as lacelike and usually subtle.



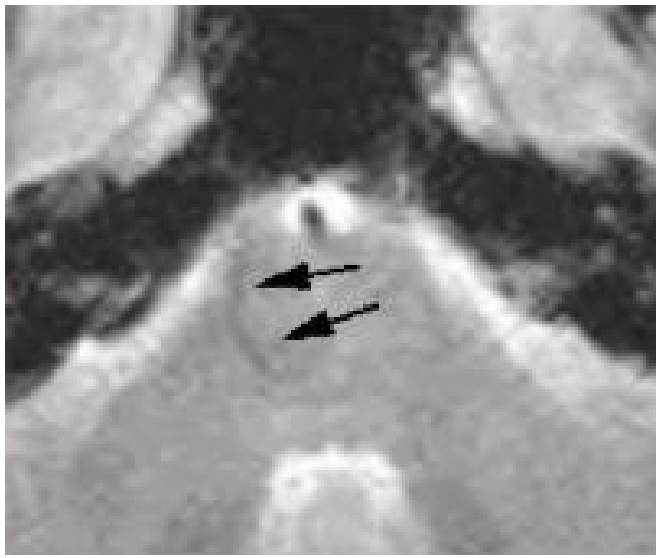
Pontine capillary telangiectasia. Note the lacy enhancement characteristic of this lesion.



Axial enhanced T1-weighted MRI :The typical lacy enhancement pattern of a capillary telangiectasia.

Capillary telangiectasias(4)

- Associated prominent draining vein is present.



- Axial fast low-angle shot gradient-recalled echo MRI obtained through the pons shows **a linear area of decreased signal extending from the inferior edge of the malformation** (arrows). This finding indicates that the lesion may be a combined capillary telangiectasia and developmental venous anomaly because it has characteristics of both.

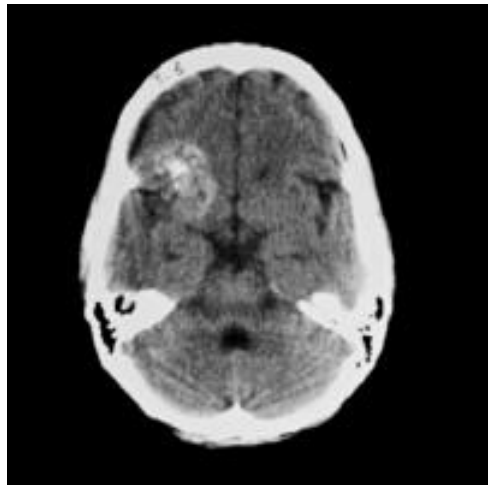
Cavernous angiomas(1)

- Cavernous angiomas belong to a group of intracranial vascular malformations that are developmental malformations of the vascular bed.
- Patients may be asymptomatic, although they often present with headaches, seizures, or small parenchymal hemorrhages.

Cavernous angiomas(2)

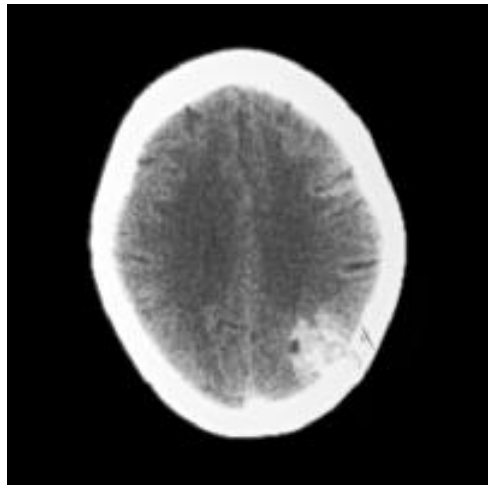
- Nonenhanced CT scans : focal oval or nodular-appearing lesions that demonstrate mild-to-moderate increased attenuation, **without** mass effect on the surrounding brain parenchyma.

Large, right frontal and left occipital cavernous angiomas



- Axial NECT:

a large heterogeneous-appearing lesion in the right frontal region. The lesion is primarily hyperattenuating in its central region, with a more diffuse, peripheral area of increased density resulting from calcification and small areas of hemorrhage.



- Axial NECT:

A large primarily hyperattenuating mass in the left occipital region. Note the relative lack of mass effect on the surrounding parenchyma.

Cavernous angiomas(3)

MRI

- Tyical shows:

1. popcornlike
2. smoothly circumscribed
3. well-delineated complex lesions
4. not associated with mass effect or edema
5. do not demonstrate a feeding artery or draining vein

Cavernous angiomas(4)

- Angiography:

  Nonspecific (20-27%)

  Avascular masses (37-48%) :

  extremely slow flow of blood through these lesions, cerebral arteriographic findings are often normal.

  Capillary blush (12-20%):

  can be seen in a variety of other processes and entities.

Impression

- AVM was highly suspected due to image finding and personal history

Discussion - Arteriovenous Malformation

Introduction:

- Arteries connect directly to veins with no intervening capillary bed.
- AVMs account for approximately 11% of cerebrovascular malformations; the more common venous angiomas account for 64%.
- AVMs are more likely than other types of vascular malformations to be clinically symptomatic.

Categorization of AVMs

- By their blood supply

Pial or parenchymal AVMs :

Internal carotid or vertebral circulation

Most Common and congenital

Dural AVMs:

External carotid circulation

Relatively uncommon and secondary to trauma, surgery, thrombosis of an adjacent venous sinus, or veno-occlusive disease

Mixed AVMs:

Both

the lesion is large enough to recruit blood vessels from both the internal and external carotid arteries

- Vein-of-Galen aneurysm(Pediatric variant of AVM):

an AVM drains to and dilates the great vein of Galen.

Clinical Symptoms

Pial AVMs

- asymptomatic until the second, third, or fourth decade of life.
- most commonly manifest as spontaneous hemorrhage or seizure.
- headache and transient or progressive neurologic deficit.

Dural AVMs

- typically feature pulsatile tinnitus, cranial bruit, headache, or hemifacial spasm.

Infants with a vein-of-Galen malformation

- with hydrocephalus or severe congestive heart failure.

Imaging of AVMs(1)

CT:

- Evaluating acute headache or other acute mental-status changes suggestive of acute cerebral hemorrhage.
- An underlying mass or AVM.
- To identify areas of acute hemorrhage.
- Vascular calcifications associated with AVMs.

Imaging of AVMs(2)

MRI

- dilated feeding arteries + enlarged draining veins
- uniquely show these lesions as a tangle of vascular channels that appear as flow voids.(imaging with GRASS gradient echo+long TR sequences)

Imaging of AVMs(3)

Angiography

- Dynamic real-time study - vascular transit time.
- Dilated efferent + afferent vessels - tangled cluster of vessels (bag of worm).
- AV shunting into early draining vein.
- To evaluate the venous drainage pattern
- Associated risk factors for hemorrhage - aneurysms and venous stenosis.

Spetzler and Martin grading system(1)

Features	Score
Size of nidus	
Small (<3 cm)	1
Moderate (3 to 6 cm)	2
Large (>6 cm)	3
Located in eloquent region†	
No	0
Yes	1
Venous drainage	
Superficial	0
Deep	1

Spetzler RF, Martin NA: A proposed grading system for arteriovenous malformations. J Neurosurg 1986 Oct; 65(4): 476-83

Spetzler and Martin grading system(2)

- Grade = sum of points.
- Grade I: small, located in a non-eloquent region, and has only superficial drainage.
- Grade V: larger than 6 cm, located within or immediately adjacent to an eloquent region, and has at least partial drainage into the deep venous system.
- A ‘Grade VI’ category refers to an inoperable lesion.

Factors influencing treatment

- patient and family preferences
- Spetzler-Martin grade
- lesion site
- angioarchitecture
- clinical presentation,
- neurologic status
- age
- past medical history
- pregnancy

Pathophysiology

- The pathogenesis of AVMs is not well understood.
- Molecular differences between arteries and veins
- Capillary-bed morphogenesis
- Inherited disorders of vasculogenesis.

Frequency

- Internationally:

  incidence of AVMs - 0.04-0.52%

Mortality/Morbidity

- Spontaneous intracranial hemorrhage is 2-3%/year: 10-15% rate of mortality and a 20-30% rate of permanent neurologic deficit.
- After the first hemorrhage, rebleeding rate:

  First year: 6% and than 2-4% per year
- Hemorrhage:implicated in 29% of patient deaths.

Race

- No clear correlation exists between race and the prevalence of AVMs

Sex

- slightly increased preponderance of pial AVMs in men
- Dural AVMs occur more commonly in women.
- Dural AVMs of the anterior cranial fossa occur more frequently in men than in women.

Age

- Pial AVMs are present from birth - More than 95% of patients develop symptoms before age 70 years.
- Dural AVMs, are believed to be acquired and to develop during adulthood.

Anatomy-Anatomic location

- Location:

- 90% are supratentorial and tend to occur at watershed areas: parietal > frontal >temporal lobe> paraventricular > intraventricular region > occipital lobe
- 10% are infratentorial

Anatomy-Anatomic location

- Pial AVMs:

  Brain parenchyma

  Derive blood from the cerebral arteries (ACA/MCA/PCA).

- Dural AVMs:

  Almost infratentorial.

  Feeding arteries: Occipital artery and meningeal branches of the external carotid artery are the vessels.

  Most frequently drain into the transverse and sigmoid sinuses in the posterior fossa

Feeding arteries and vessels

- ***Circumferential feeding artery*** - extends around the nidus and sends branches both to small arterioles connected to the nidus and to normal brain capillaries.
- ***Penetrating feeding arteries*** - bisect the AVM core and send branches to it.
- ***Final feeding arteries*** - either connect directly to an AVM loop or branch to shunting arterioles.

Feeding arteries and vessels

- The larger veins → major draining vein → sulcus → numerous venules → neighboring cortical veins → large hemispheric veins → venous sinuses.

Treatment

- Embolization +/-

1. Direct surgical
2. Microsurgical resection
3. Radiosurgery.

Reference

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