Vascular malformations of the brain: tips and tricks for young residents at a glance

Poster No.: C-0499
Congress: ECR 2016
Type: Educational Exhibit
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Keywords: Vascular, Neuroradiology brain, CT, MR, Catheter arteriography, Angioscopy, Embolisation, Diagnostic procedure, Aneurysms, Arteriovenous malformations, Fistula
DOI: 10.1594/ecr2016/C-0499
Learning objectives

To review the most important entities concerning vascular malformations of the brain vessels.

To recognize the most useful imaging method for each pathology.

To become familiar with the typical aspects of these malformations on CT, MRI and angiography.

To review the therapeutic possibilities.
Background

Vascular malformations of the brain are an important neurological finding that are encountered frequently in day to day practice. Therefore, we consider that a young radiologist should be acquainted with some basic information on the subject when viewing scans. From our clinical experience, we reviewed five entities that are the most common, the most relevant and will be the focus of this presentation: aneurysms, arterio-venous malformations, arterio-venous fistulas, venous angioma and cavernous angioma. For each one, we'll review the key point regarding pathology, clinical aspects, treatment options and of course imaging aspect on CT, MR and angiography.
Findings and procedure details

To begin with, we'll be discussing separately the five entities mentioned, including therapeutic possibilities. The emphasis will be on the imaging aspect. In the end we'll try to review some quick tips and tricks to help junior doctors in their daily practice.

Aneurysms (Fig. 1-4)

There are several categories: saccular or berry aneurysm (also known as true aneurysm), pseudoaneurysm, fusiform aneurysm and blood blister-like aneurysm. Since the sacular type is the most common and with the most relevance, it will be the main topic of this section.

The berry aneurysm represents a focal outpouching involving part of an arterial wall; it has a particular morphology lacking the elastic layer or the medial one. It is also of importance that almost all of them all acquired, due to abnormal hemodynamics and stress of the vascular wall. There is an increased risk of appearance in case of vascular asymmetries, congenital vascular anomalies (bicuspid aortic valve, aortic coarctation, congenital anomaly of the anterior cerebral arteries) and certain syndromes or diseases (such as Marfan, Ehlers-Danlos, type I neurofibromatosis or autosomal dominant polycystic kidney disease).

They are found mostly in the anterior circulation (anterior and posterior communicant arteries, middle cerebral artery bifurcation), with only a small percentage involving the basilar tip or the cerebellar arteries. Size ranges from below 3mm to above 2.5cm.

Clinically, they are most commonly asymptomatic. Mean age of diagnosis ranges between 40 and 60. They represent up to 90% of the causes of nontraumatic subarachnoid hemorrhages, where the main complaint of the patient is an intense headache ("thunderclap") . The risk of bleeding is usually low if the size is less than 7mm, but once ruptured, regardless the size, there is always a risk of recurrence without intervention.

There are two ways to approach them. The surgical method, although invasive, offers a lower risk of recurrence. The endovascular approach uses coil embolization and is especially beneficial for unruptured aneurysms, with low risks, low procedural costs and faster recovery of the patient.
Imaging findings:

CT:
- Subarachnoid hemorrhage when the aneurysm ruptured
- Hyperdensity if there is any thrombus
- Large lesions may appear as hyperdense masses
- Occasional mural calcifications
- When using contrast medium, the lumen enhances uniformly, however if there is any thrombus, there will be a rim enhancement

MR:
- T1: there is a 50%-50% chance of flow voids or iso/heterogenous signal; with thrombus, the signal depends on its age. Also, there is hypointensity and blooming on the susceptibility sequences (GRE and SWI).
- T2: hypointensity (flow void); possibly laminated with hypointense rim
- FLAIR: possible acute subarachnoid hemorrhage
- DWI: restriction due to vasospastic ischemia and thromboembolic episodes

Angiography - Gold Standard imaging method for aneurysms:
- Typically performed by injecting the contrast medium in the carotid and basilar arteries, bilaterally;
- Occasional extravasation in case of hemorrhage

Differential diagnostic:
- Vessel loop: other angles of viewing should be used to solve this issue
- Arterial infundibulum: symmetric, conic dilatation at the origin of the vessel, less than 3mm diameter
- Fusiform aneurysm: has the shape of a sausage, involving the whole vessel circumference, with separate inflow and outflow ways
- Pseudoaneurysm: further from the circle of Willis, with more of a fusiform or irregular shape
- The flow voids could be mistaken for an aerated anterior clinoid process

Arterio-venous malformations (AVMs) (Fig. 5-7)

They represent a conglomerate of vessels (nidus) with thin walls and with arterio-venous shunting, without any capillaries between the two. AVMs are congenital lesions that gradually progress and are usually diagnosed after the age of 40. The majority are
solitary, but can sometimes be multiple (less than 2%), as in the case of Rendu-Osler-Weber syndrome or Wyburn-Mason syndrome.

Pathologically, 85% are located supratentorial and with size that varies from 2 to 6 cm. Ovoid or pyramid like in shape, the largest surface is adjacent to the cortex and the tip points to the ventricles. The nidus itself does not contain normal brain tissue, but may contain any number of thrombus, calcifications, hemorrhagic residue and sometimes nonfunctioning gliotic brain tissue. The area adjacent to the nidus is often abnormal, with the occasional presence of a perinidal capillary bed.

Clinically, the patient presents with any of the following symptoms: headache accompanied by parenchymal hemorrhage (50%), seizures (25%) or focal neurologic deficits (25%). The hemorrhagic risk increases with age, deep localization, deep venous drainage, intranidal aneurysm, previous hemorrhage and stenosis of the draining vein. To grade the AVMs, the most commonly used scale is the Spetzler-Martin, which takes into account the size of the AVM, its localization and its venous drainage.

Imaging uses CT, MR and angiography, the last two being the best options. Below, the main imaging methods for AVM will be reviewed, followed by a few notes on the differential diagnostic.

CT:

- Small sized AVMs may appear normal
- Usually described as a "bag of worms" with minimal or no mass effect
- Hemorrhage can be visualized as well as occasional calcifications
- With contrast, AVM enhances intensely and uniformly

MR:

- T1: described as a "honeycomb" of "flow voids"; with contrast, there is strong enhancement within the nidus and the veins; however, due to the high flow rate, the enhancement may be absent
- T2: described as the same compact, serpiginous aspect of T1, with or without hemorrhage; some minimum gliotic brain tissue within the nidus, with high signal, may occur
- T2*: frequent foci of "blooming" when hemorrhagic residue is present
- FLAIR: flow voids and surrounding high signal due to gliosis

Angiography:
• - Tortuous and enlarged arteries
• - Associated angiopathy: dilatation, thickened endothelium, stenosis, thrombosis and occlusion
• - Pediculal aneurysm
• - Half of the cases present an intranidal aneurysm, with minimum or no mass effect
• - Opacification of the draining veins during the mid and late arterial phase; they appear tortuous and enlarged; optionally, varices may create a mass effect

Differential diagnostic:

• - Glioblastoma, which shows intense enhancement, a large amount of neoplastic tissue between the vessels and mass effect
• - Thrombosed AVM, which may show only a mass effect with "stagnating vessels"; it may be difficult to distinguish these lesions from hemorrhagic neoplasms or other malformations
• - Cerebral proliferative angiopathy, which contains a large amount of feeding vessels, there is no nidus and normal brain tissue is found between the vessels

Treatment options:

• - Microsurgical resection
• - Stereotactic radiosurgery: success rate is higher with small lesions
• - Embolization: more efficient in smaller lesions and is often performed prior to surgery; in case of a rupture, the target should be the site of the bleeding in order to prevent recurrence

**Arterio-venous fistulas (AVFs) (Fig. 8)**

Also referred to as dural arterio-venous shunts, AVFs represent a network of small vessels, similar to capillaries, that connect meningeal arteries to small venules within the wall of a dural venous sinus. Basically, multiple enlarged feeding vessels target the wall of a thrombosed sinus, where they are connected to draining veins with the help of microfistulas and have the possibility to create a focal mass.

In adults arterio-venous fistulas are acquired, mostly after dural sinus thrombosis or after a traumatic incident; in children this entity is congenital. The most commonly affected sinuses are the transverse, the sigmoid, the cavernous and the superior sagittal ones and are associated with cortical venous drainage, edema, encephalopathy, high risk of
hemorrhage and lateness in a child's development. Also, there are two classifications commonly in use: the Cognard and the Borden.

Clinically, the patient presents bruit and tinnitus in case of transverse/sigmoid sinus involvement. Should the cavernous sinus be implicated, one can encounter pulsatile proptosis, chemosis, retroorbital pain, bruit and ophthalmoplegia. Lastly, there are the "malignant" AVFs, with cortical drainage that can lead to seizures and focal neurologic deficit.

Treatment:

- Conservative: observation; in case of hemorrhage, one of the following should be used:

  - Endovascular embolization of the arterial feeders with liquid agents and of the venous component with coils
  - Surgical resection
  - Stereotaxic radiosurgery

Imaging findings:

CT:

- Enlarged dural sinus or draining vein
- Enlarged vascular channels within the skull (seen on the bone window)
- Enlarged ipsilateral foramen spinosum
- Hemorrhage (in case of cortical venous drainage) and edema (in case of venous hypertension)
- Contrast study shows enlarged tortuous arteries, enlarged veins and sometimes an aneurysm

MR:

- The absence of a nidus and the presence of enlarged cortical veins may suggest the diagnostic
- T1: thrombosed dural venous sinus with flow voids
- T2: same aspect as T1, but also with focal hyperintensity in the adjacent tissue
- T1C: enhancement of the chronically thrombosed sinus
- T2*: "blooming" of the thrombosed sinus and hemorrhage
- FLAIR: isointense thrombosed sinus with adjacent edema
- DWI: normal aspect if there is no ischemia or infarction
Angiography:

- Multiple enlarged dural or transosseous arteries originating in the external carotid artery; there is the possibility of dural or tentorial vessels originating in the internal carotid artery or the vertebral artery
- A branch of the meningo-hypophyseal trunk can be part of a AVF in the transverse/sigmoid junction
- The presence of thrombosis, inverted flow with cortical venous drainage, pial tortuous veins
- Dysplastic venous "pouches" that may cause a mass effect

Differential diagnostic:

- Thrombosed dural sinus with collateral venous drainage: the feeding arteries are not enlarged and there are no microfistulas
- Pseudolesion of the jugular bulb: no thrombus on T2*, no abnormal arteries and no enlarged venous collaterals
- Pial fistula/AVM: they are a direct shunt between parenchymal arteries and enlarged cortical draining veins; they appear within the brain or at its surface

**Venous angioma (Fig. 9)**

Also referred to as developmental venous anomaly, it is the most common vascular malformation and is easily recognized by its umbrella-like aspect. This entity contains mature dilated venous elements with thin walls, located in and separated by normal nervous tissue.

The occurrence of venous angioma is genetically linked to chromosome 9 mutations and may associate with cavernous malformations, sinus pericarini, blue rubber bleb nevus syndrome, giral anomalies.

The usual location is in the white matter, next to the frontal horn of the lateral ventricle or near the 4th ventricle. The size varies, but they are usually smaller than 3cm. The best hint is the "medusa head", which will be shown in the imaging section.

Clinically, 98% are asymptomatic. The remaining 2% may lead to hemorrhage or infarction due to stenosis or thrombosis of the collecting vein. They may be associated with cavernous venous malformations or even a triad of cavern-vein-capilay malformation. When symptomatic, one can experience headaches and seizures.
There is no treatment indication for the solitary venous angiomas. In case of venous angiomas accompanied by other malformations, therapeutic approach should be dictated by the latter entity.

Imaging:

CT:
- Frequently normal on nonenhancement studies
- Possible hyperdense collecting vein
- Calcifications if there is an associated cavernous malformation
- Rarely: acute hemorrhage
- With enhancement, linear or dotted enhancement foci, converging towards a collecting vein

MR:
- T1: may be normal on solitary DVAs; also, possible hemorrhage and flow voids, depending on size and flow
- T1C: strong enhancement, with stellate/tubular vessels converging towards a vein
- T2: possible flow voids and blood
- T2*GRE: possible hypointensity (blooming) if the DVA is large or there is a hemorrhagic cavernoma associated
- FLAIR: frequently normal; hypointensity in case of venous ischemia or hemorrhage
- DWI: frequently normal; rarely, acute venous infarction as a hyperintensity (diffusion restriction)

Angiography:
- Arterial phase: frequently normal, with an occasional capillary stain
- Venous phase: the medusa head, specific to this malformation

Differential diagnostic:
- Mixed vascular malformation, usually cavernomas: frequently present hemorrhage
- Vascular neoplasm: enlarged medullary veins, mass effect, enhancing
- Chronic dural sinus thrombosis: venous stasis, enlarged medullary veins
- Sturge-Weber syndrome: possible very large medullary/subependimal/choroid plexus veins and an associated facial angioma
**Cavernous angioma** (Fig. 10,11)

Also called cavernoma, it is a benign vascular hamartoma, containing hemorrhages of different ages within caverns; they are well defined and contain no brain tissue. May be inherited or acquired (usually after radiotherapy) and represent the most common component of mixed malformations.

Cavernous angiomas may be located anywhere in the central nervous system and have variable size. With berry-like aspect, cavernomas contain well defined hemorrhagic vessels, no brain tissue and have a rim of gliotic hemosiderin filled brain tissue.

Clinically, it is the third most common type of cerebral vascular malformation, with the mean age of occurrence at 40-60 years. The patient usually presents with seizures, headaches or neurological deficit. Lesions may develop in time and have a considerable hemorrhagic risk.

Treatment options include:

- Microsurgical resection, for symptomatic cases with hemorrhage
- Stereotactic radiosurgery for nonsurgical cases
- Venous drainage must be preserved in mixed lesions

**Imaging:**

**CT:**

- Frequently normal on nonenhancement studies
- Possible round hyperdense lesion, well defined
- Normal surrounding brain
- With contrast, little to no enhancement in non-mixed lesions

**MR:**

- **T1:** variable aspect (it depends on the hemorrhage); frequently, "popcorn ball" aspect, representing foci of hyper/hypointense blood; there is also a perilesional hyperintensity, differentiating it from other hemorrhages
- **T1c:** minimum to no enhancement; it can show associated venous malformations
- **T2:** popcorn-like lesion; the core has mixed intensity with a hypointense rim; it can also show blood foci with liquid-liquid levels
- **T2*GRE:** prominent susceptibility effect (hypointense blooming); in case of multiple malformations: many hypointense dot-like foci ("black dots")
• - DWI: normal
• - FLAIR: possible surrounding edema

Angiography:

• - Frequently normal, with the exception of the extradural malformation, which is very vascular
• - Slow intralesional flow, without any arterio-venous (AV) shunt
• - If the lesion is large or any acute hemorrhage is present, there is a mass effect
• - Useful in mixed lesions
• - Rarely: venous pooling

Differential diagnostic:

• - For the popcorn aspect: AVM, hemorrhagic neoplasm and calcified neoplasm
• - For the multiple black dots: contusions, hypertensive microbleeding, amyloid angiopathy, capillary telenangiectasis
Fig. 1: Internal carotid artery aneurysm: Axial T2: hypointense mass in the suprasellar region Axial T1: the lesion shows isointensity to the surrounding tissue Axial T1Gd: marked homogenous enhancement Coronal T1: the lesion is hypointense; the signal intensity varies in different planes of acquisition due to the flow phenomena Coronal T1Gd: there is mixed intensity, with signal loss due to pulsation

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Fig. 2: Middle cerebral artery aneurysm: Nonenhaced CT: round area of discretely high density, anterior to the sylvian fissure. Enhanced CT: the lesion enhances uniformly. CT and standard angiography: demonstrates the presence of the aneurysm.

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**Fig. 3:** Partially thrombosed aneurysm of the middle cerebral artery: Nonenhanced CT: area of mixed density, with a peripheral hyperdense area, located in the sylvian fissure. Enhanced CT: eccentric enhancement of the lumen and the periphery of the aneurysm. T1: the patent lumen of the aneurysm with flow voids; the surrounding mixed signals represent blood clots in different stages; the hypointense rim represents the wall of the lesion. Angiography: confirms the presence of the aneurysm and the thrombus.

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Fig. 4: Ruptured aneurysm of the anterior cerebral artery: A: subarachnoid haemorrhage in the suprasellar and prepontine cistern, sylvian fissure and interhemispheric fissure B: haemorrhage located in the interhemispheric fissure, suprasellar, interpeduncular and ambien cistern; hematoma in the interhemispheric fissure; hydrocephalus C: subarachnoid haemorrhage in the sylvian and interhemispheric fissures; hematoma in the septum pellucidum

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Fig. 5: Talamo-nuclear AVM: A, B: enlarged veins (including the vein of Galen) and anterior/middle cerebral arteries C: heterogenous mass located in the level of the right thalamus and basal ganglia; it is made of numerous serpiginous structures, mostly with low signal due to the flow effect D: TOF-ARM: branches of the middle cerebral artery are enlarged and surround the AVM; also, some enlarged draining veins can be seen

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Fig. 6: Cortico-ventricular AVM: Nonenhanced CT: poorly differentiated area, spontaneously hyperdense Enhanced CT: heterogeneous enhancement of the lesion PD: numerous serpiginous hypointense channels (flow voids) T2: the lesion is hypointense, with hyperintense intranidal areas (gliosis)

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Fig. 7: Standard angiography: clearly shows the AVM, with the nidus and the dilated feeding artery and raining veins

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Fig. 8: AVF with intracerebral and subdural haemorrhage: Enhanced CT: focal enhancement next to a recent occipital hematoma; small subdural hematoma MR (PD): vascular channel with flow void adjacent to a recent intracerebral hematoma; recent hyperintense subdural hematoma Angiography: AVF between branches of the ECA and the vein of Galen (via the infratemporal vein); the venous pseudoaneurysm is the presumed site of haemorrhage

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Fig. 9: Right frontal venous angioma: Axial T1 and T2: right frontal stellate lesion, with the flow void phenomena Coronal T1: shows the star-shape characteristic of the malformation Angiography: shows the typical medusa head shape and the enlarged superficial draining vein

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Fig. 10: Cavernous cavernoma: CT: hyperdense lesion adjacent to the occipital horn of the ventricle; the second scan was taken 3 years later after an acute episode of headache and shows the enlargement of the lesion T2 MR: taken after 5 year and shows the reduced size of the angioma with a hypointense rim (hemosiderin); also note small hypointense lesions in the temporal lobes and a mixed lesion in the midbrain

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**Fig. 11:** Multiple cavernous malformations: T2: mixed signal pontine cavernoma; additional lesions in both cerebral hemispheres that are poorly seen on T2 PD: the additional lesions are better seen as they appear hypointense

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Conclusion

Vascular malformations are neurologic findings all residents should at least suspect on a basic level when interpreting brain imaging. One must know when additional and more advanced methods of investigation are required, how to gather all the information and how to make a proper description of the lesion. Accurate reports are critical in deciding the therapeutic approach.
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Acknowledgement:

I would like to thank Andreea Marinescu, Bogdan Dorobat and Professor Gheorghe lana for the images and supporting me in my work.
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