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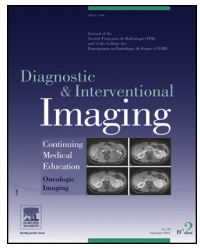

CONTINUING EDUCATION PROGRAM: FOCUS...

Intracranial arteriovenous malformations

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Dural arteriovenous malformation;
MRA (magnetic resonance angiography);
CT angiography

Abstract Intracranial arteriovenous malformations (AVM) are rare lesions that are often discovered fortuitously. They should be identified on CT scan and MRI before resorting to angiography; the latter is used to prepare the treatment. This article describes the various types of subpial or dural AVM and the specific characteristics that enable their differentiation with non-invasive imaging. The factors that determine the severity of these lesions, whether discovered before or after a haemorrhage, are described, as well as prognostic indicators.

General information

Definition

An intracranial arteriovenous malformation (AVM) is defined as the existence of a congenital or acquired arteriovenous shunt. The imbalance between arterial input and venous output (dysregulation) can result in an aneurysmal dilation of the draining venous sector due to excessive flow, but also within the nidus and the afferent arteries. Unless there is a haemorrhagic or thrombotic complication, there is no space-occupying effect on the adjacent parenchyma.

Macroscopically, between a few millimeters to several centimeters in diameter, this haemodynamic ''short-circuit'' occurs within a complex network known as a nidus, where arterioles and veins are directly connected, without an intermediate capillary bed. The nidus is surrounded by a dilated capillary network, sometimes connecting it to the normal peri-nidal arterial and venous network.

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Microscopically, the components of the vascular tissue present varied alterations, with zones of nonspecific fibrosis, excess collagen I and III, interruptions of the media, or even interruptions of the internal elastic membrane. There is no cerebral tissue within the nidus, but a peripheral gliosis may be observed.

Epidemiology

Intracranial AVM are a rare pathology. The prevalence and incidence are difficult to evaluate, estimations put the prevalence (all forms of discovery) at between 10 and 15/100,000 inhabitants, and the incidence at around 1.3 new cases/100,000/year [1,2], with a marked predominance of pial AVM compared with dural AVM. In France, in theory, we should discover around 800 AVM/year, of which 300 are haemorrhagic.

The different types of AVM

Several types of AVM can be distinguished, notably thanks to the use of magnetic resonance angiography (MRA), with differing angio-architecture, symptoms, prognosis, and treatment.

Subpial AVM

These are the most common. The nidus is located in the subpial space, within the cortex and/or the white matter. Variable in shape, pseudo-spheric, sometimes flat, often triangular with an external base, and may be several centimeters in diameter, it is sometimes diffuse, with a very tiny network, and sometimes extensive.

The afferent arteries are principally branches from the intradural network of the carotid and/or vertebro-basilar circulation. With cortical and therefore peripheral AVM one can detect arterial supply from the dural network (branches of the external carotid as middle meningeal artery or posterior meningeal artery issued from the vertebral artery) or subcutaneous anastomoses issued from the occipital artery or superficial temporal network. Certain cortical AVM are very small lesions (1 to 5 mm), located on the surface of the cortex. They can be very difficult to detect, sometimes requiring several arteriographic examinations.

Superficial pial AVM

The most commonly observed in the paediatric population, they are composed of an almost-direct shunt (true fistula). The flow is very high and a major venous dilation is evident. These are rare lesions with a high haemorrhagic potential.

Dural AVM

Incorrectly termed ''dural fistulas'', these are also nidus with a specific location since they are located within the dural wall of venous sinuses. They represent around 10 to 15% of intracranial AVM; unlike cerebral AVM, these are vascular malformations that are acquired following a triggering event such as cerebral phlebitis that may have gone unnoticed [3]. It is important to look for a previous history of cranial surgery, infection (mastoiditis), trauma, or a predisposition for venous thrombosis. The primary cause of these dural AVM is venous hypertension. The most common location is the sigmoid sinus. The nidus is often very small (a few millimeters) and the major part of the pathological network is represented in imaging by the dural and/or cortical venous drainage network.

The supply is almost exclusively from the dural branches of the external carotid network (middle meningeal artery, occipital artery, ascending pharyngeal artery, etc.), vertebral (posterior meningeal artery) or more rarely dural branches of the intracranial network (artery of the free border of the tentorium, dural branch of the posterior choroid network or cerebellar network, dural branches of the ophthalmic artery).

Clinical signs

Intracerebral haemorrhage

This is the most serious and common manifestation. The bleeding is intra-parenchymal and/or intraventricular (Fig. 1), linked to a rupture within the nidus or on a

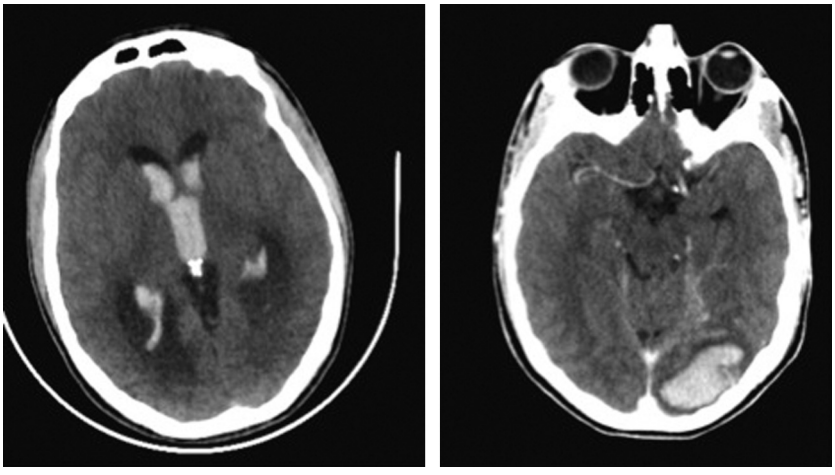

Figure 1. Two types of AVM ruptures in CT: on the left, intraventricular haemorrhage. One the right, left occipital cerebral haematoma (CT angiography).

sometimes aneurysmal draining vein. It is generally estimated at around 2—4% per year [4,5] for a lesion that has never bled before, but this rate does not take into account complex forms of AVM that have a much higher risk [6,7] (deep venous drainage, aneurysms). The risk of an acute rebleed is much lower than with intracranial aneurysms, but it can be as high as 30% in the first year in cases of deep AVM, with a single deep venous drainage (internal cerebral veins) and for which bleeding is the first indicative sign [7]. The neurological deficit is sudden, linked to the location of the bleed. Tolerance, as with the prognosis, is often better than in haemorrhages related to arterial hypertension, except for AVM of the posterior fossa or the mesencephalon. The triggering factor for bleeding is often unknown, very rarely linked to exertion, other than significant episodes of blockpnea that increase the venous pressure suddenly and massively. The venous side of the drainage is sometimes responsible via venous hypertension upstream from a stenosis or a thrombophlebitis. However, the haemorrhagic risk is closely linked to the cortical veins for the dural AVM where it is linked to the fragility of these veins put under pressure by the shunt. Aneurysms on the afferent network may rupture (classic aneurysmal subarachnoid haemorrhage [SAH]), as with intra- or paranidal aneurysms (intracranial hypertension (ICH) on a AVM rupture).

Seizures

This is the second symptom in terms of frequency, they are most often seen in junctional AVM, with cortical drainage, in the temporal zone, and in middle-aged subjects. Partial, simple, complex, or even generalized, they may also be observed following a haemorrhage or in the event of thrombophlebitis on the drainage network of the AVM with an associated cortical oedema.

Transient neurological deficit

Not to be confused with a deficit following haemorrhage, its symptoms, which are always transient, could correspond to deficit epileptic episodes or local vascular stealing phenomena.

Headaches

These are very common, atypical, and often impossible to link to the presence of the AVM. Their very localized, sometimes pulsatile character, or differing from a migraine known to the patient, should raise the alarm and may be linked to an alteration in the drainage of the anomaly, such as a fissured venous aneurysm (look for an intra-aneurysmal thrombus) or an early thrombosis within the AVM or on the drainage network.

Clinical signs of venous hypertension

These are almost exclusively seen with dural AVM and very rarely with cerebral AVM. Other than haemodynamic turbulence phenomena, the high flow rate within the venous system results in a local increase in venous pressure. This may be exacerbated in the event of venous stenosis or thrombosis leading to congestive signs upstream, which are linked anatomically to the drainage veins in question. On auscultation, the systolo-diastolic murmur can be heard fairly easily towards the mastoid or over the eye. The signs are very variable, as a function of the location of the shunt:

- typical pulsatile tinnitus linked to turbulences in the sigmoid and/or petrous sinus (AVM of the sigmoid sinus, the most common) (Fig. 2);
- Congestive orbital signs (turgescent red eye) by hypertension in the superior ophthalmic vein draining an AVM of the cavernous sinus (Fig. 3);
- medullary oedema with progressive tetraparesis for a dural condylar AVM or of the occipital foramen blocking the cervical medullary venous return (Fig. 4);
- confusion and dementia that may progress to coma via generalized intracranial venous hypertension on fistula with a very high flow rate of the torcular or longitudinal sinus (Fig. 5).

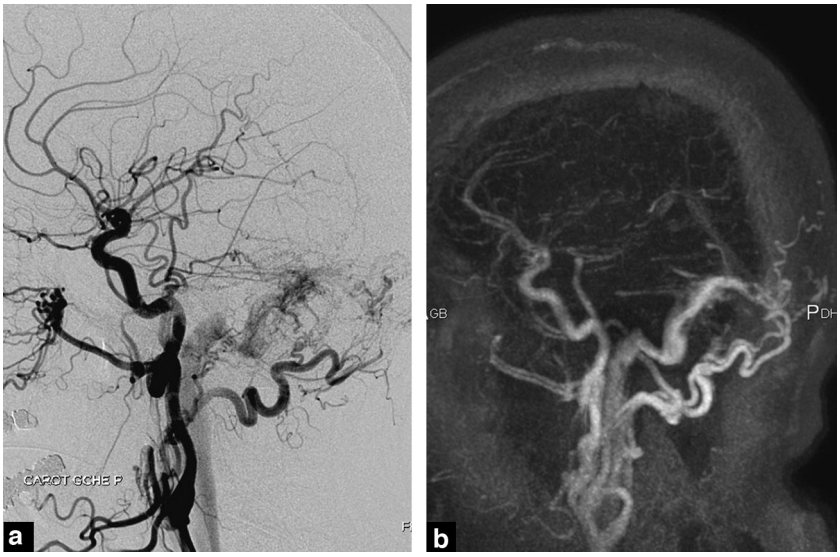

Figure 2. Pulsatile left tinnitus on a dural AVM of the left sigmoid sinus seen from the side in angiography (a) and dynamic 4D MRA (b). Opacification of the arterial phase of the sigmoid sinus by the external carotid dural network.

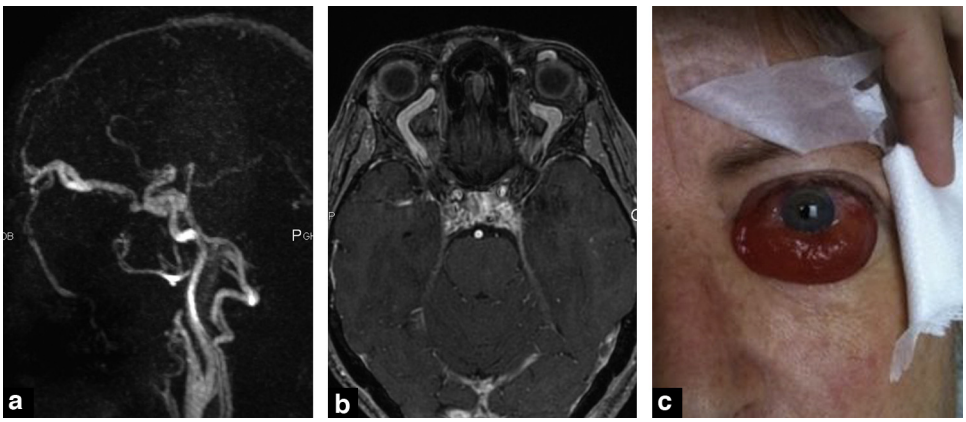

Figure 3. (a) Dynamic 4D MRA showing the venous drainage of a dural AVM of the cavernous sinus towards the superior ophthalmic veins (SOV). (b) Same patient: raw slices in axial T1 with contrast; note the dilation of the 2 SOV. (c) Congestive ophthalmological signs typical of a dural AVM of the cavernous sinus.

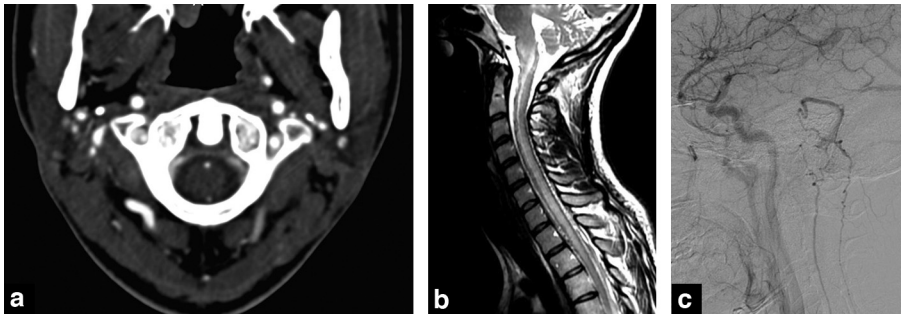

Figure 4. Dural AVM of the occipital foramen with inaugural tetraparesis. (a) Dilation of the anterior spinal vein in CT angiography. (b) T2 hyperintensity of the cervical spinal cord. (c) Pathological ''slowed'' opacification of the cervical perimedullary venous network after left carotid injection.

Modes of discovery

Subpial AVM: the mean age at diagnosis is around 40 years. Around 25% of AVM are discovered fortuitously following a CT scan or MRI requested for a symptom that is unrelated to the AVM. Fifty-five percent of AVM discoveries are made outside of a haemorrhage context, following a seizure or neurological deficit. Ruptured AVM (intracranial bleeding) represent 45% of detected AVM.

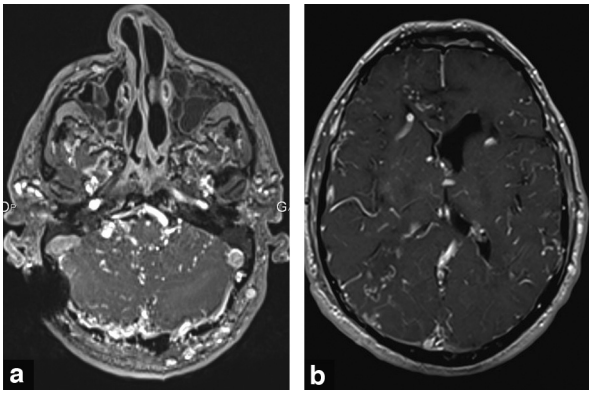

Figure 5. Dural AVM of the sigmoid sinus with confusion then rapidly progressive coma (a, b): massive opacification of the superficial and deep cortical cerebral veins.

Radiological signs

Bleeding (or imminent bleeding) linked to an AVM

CT scan

The bleeding is intracerebral, of variable amplitude not related to the size of the lesion (Fig. 1). An intraventricular haemorrhage is very common and sometimes isolated, it signals rupture of the para-ependymal contingent of the AVM (nidus and/or drainage vein) and can then cause an acute or delayed hydrocephalus (iterative CT controls). Rarely a subarachnoid haemorrhage can be associated if the AVM is superficial (dural AVM) or if the rupture occurs on an aneurysm located on the arteries that are afferent to the lesion. Thrombophlebitis may be observed on the cortical drainage veins, which should be treated as an emergency before a haemorrhage occurs.

MRI

In the acute phase of the bleed, MRI is of no use, except for detecting a low volume bleed, which may be visible as a hypointensity on T2* weighted sequence or a hyperintensity with the FLAIR technique (subarachnoid bleed). Other than a recent haemorrhagic episode, MRI can detect signs of undetected old bleeds in the form of cortical siderosis

or sub-cortical deposits of haemosiderin, often arciform or sinusoidal in shape, not to be confused with venous vascular trajectories (Fig. 6). This notion is very important to distinguish an unruptured AVM from a ruptured AVM (even old), as the therapeutic implications are very different. Any trace of recent or old bleeding should in principle prompt treatment of the lesion.

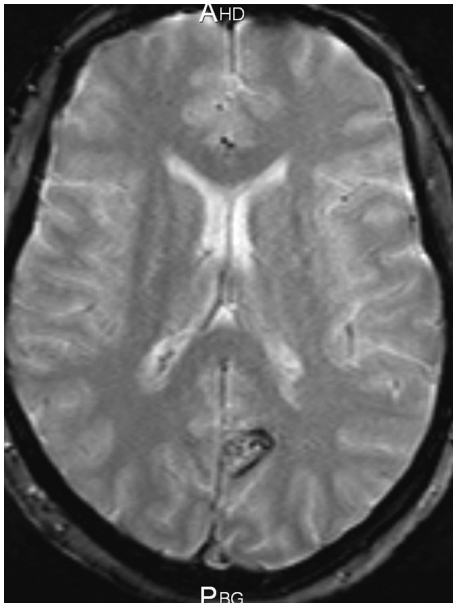

Figure 6. Old asymptomatic bleed discovered on T2* weighted MRI (work-up for asymptomatic AVM).

CT angiography or MRA

This should form part of the systematic work-up of any non-traumatic SAH (subarachnoid haemorrhage) or ICH (intracranial haemorrhage), especially before 65 years of age. The analysis of raw slices of these examinations is not useful to diagnose a haemorrhage, but can be used to detect signs of thrombophlebitis that constitute a potential pre-haemorrhagic state by an increase in the venous and nidal pressure, which could require anticoagulant treatment to prevent bleeding. One can then observe an intraluminal hypointensity on CT in a drainage vein or within a venous aneurysm (fissuration). In MRI, the thrombus can be detected in the form of an intraluminal hypointensity in T2* or on the raw slices of a time of flight sequence of after gadolinium injection. Similarly, other than their benefit for the demonstration of an arteriovenous shunt, dynamic 4D MRA can detect these stenoses and/or venous occlusions (Fig. 7). When the haemorrhage has already occurred, these various signs give the etiology of the bleeding. Very often these phlebitic signs are associated with a vasogenic oedema of the adjacent parenchyma, causing headaches and/or seizures. Diffusion-weighted MRI then shows an increase in the diffusion coefficient on the ADC cartography (Fig. 7c and d). Any space-occupying syndrome and any oedema around

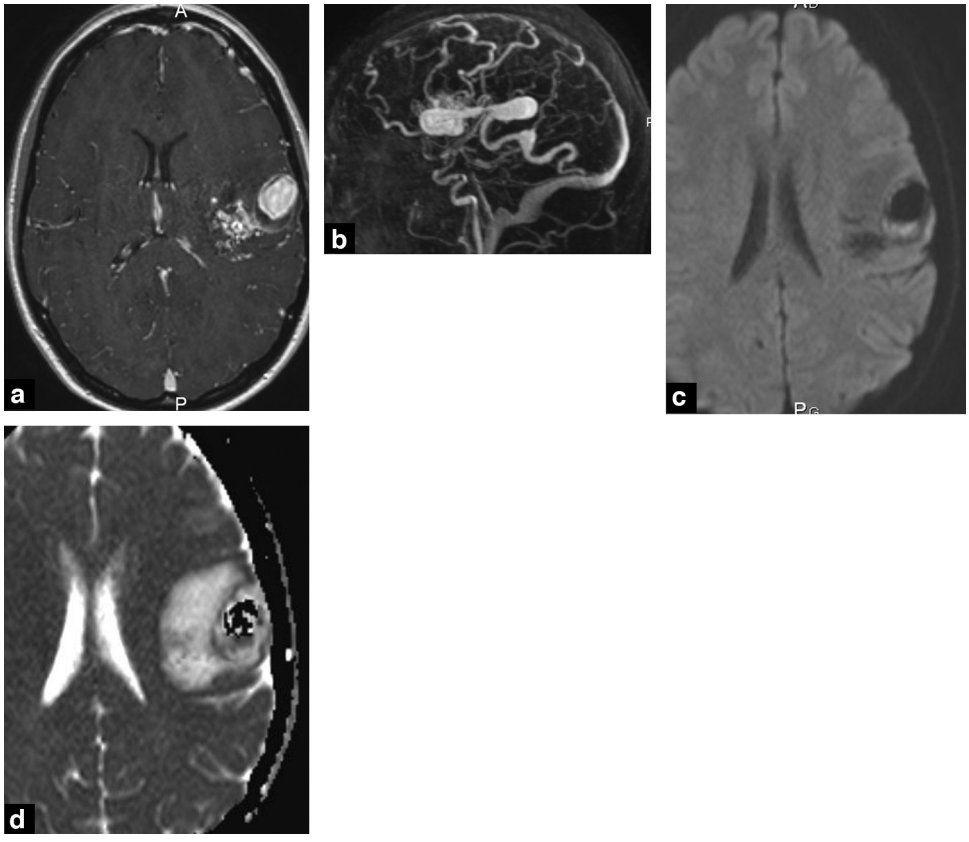

Figure 7. Seizure on AVM undergoing thrombosis. (a) Partially thrombosed (laterally) cortical venous aneurysm with an arteriovenous nidus. (b) 4D MRA (MIP) showing the aneurysmal venous network. (c, d) Diffusion image and ADC showing the acceleration of the diffusion coefficient around the aneurysm indicative of venous oedema on the drainage territory.

an AVM without haemorrhage is an emergency due to the imminent haemorrhagic potential.

Anomalies associated with bleeding or not

The nidus

This is the ''heart'' of the AVM. It may be visible on unenhanced CT, if it is big enough, in the form of a discrete hyperintensity, sometimes incorrectly interpreted as a bleed. Calcifications may be observed, punctiform or like bunches of grapes. After contrast injection, parenchymal AVM (subpial), have the form of a clump of dense vessels with arterial uptake, from a few millimeters to several centimeters in size. The nidus may have a compact or more diffuse appearance. Its often conical shape with peripheral base (Fig. 8), pointing towards the ventricles, may actually be highly variable, such as flat, or linear. Peripherally, one can observe recruitment of the surrounding vessels, slightly dilated, often helical or sinuous in appearance, corresponding to normal vessels that have been ''recruited'' by the nidus. They correspond to a stimulation of angiogenesis peripheral to the AVM, but do not constitute the nidus itself. The imaging profile is completed with CT angiography or MRA. The use of a TOF sequence performed after injection of gadolinium is very useful to provide a clear definition of the nidus in comparison with a simple TOF sequence where the intranidal turbulences and the speed of flow may generate signal losses and hide certain areas of the nidus. Care should be taken when calculating the size and volume of the nidus, not to include the reactional peripheral angiogenesis, or the primary drainage veins of the AVM, which may lead to an overestimation of the size of the lesion. With the exception of evolutive complications, the nidus never causes a space-occupying syndrome.

In the event of dural or pial AVM, the nidus is not visible as it is millimetric; only the efferent arteries and veins are visible. These AVM are therefore essentially detected via indirect signs.

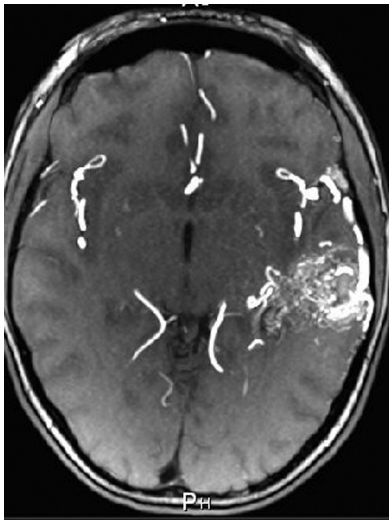

Figure 8. MIP projection TOF sequence after gadolinium. Left temporal cortico-sub-cortical arteriovenous nidus. Note the internal and cortical venous dilations.

Vascular dilations

The reduction in vascular resistance caused by the shunt and absence of capillary bed lead to an increased flow rate, as well as dilation of the afferent arteries. MRA or CT angiography reveals dilation of the vessels on the same side as the lesion. Their tortuosity is also increased (Fig. 9). Treatment of the lesion and of the shunt leads to rapid normalization of the arterial caliber.

The increased venous flow also leads to visible venous dilation of the cortical and deep veins; the dural sinuses are not affected. The veins are tortuous, enlarged, and sometimes aneurysmal (Figs. 7 and 9).

In the event of micro-AVM, only an isolated image of moderate and asymmetrical arterial or venous dilation must catch the eye. The analysis should therefore be very attentive so as not to qualify the haematoma as idiopathic and omit an AVM that could bleed at a later date (Fig. 10). If there is any doubt, an arteriography should be performed to confirm the lesion. This is especially important in cases where haemorrhage has obscured the micronidus; the arteriography should be delayed by a few days to a few weeks to

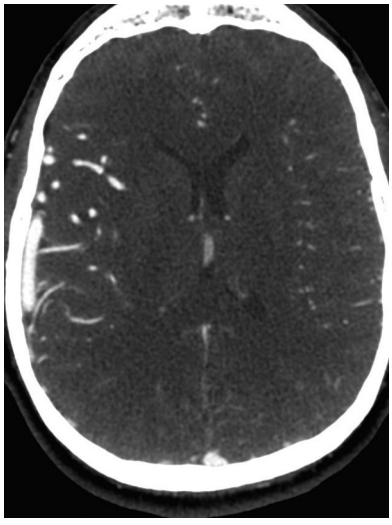

Figure 9. Right parietal AVM (not visible on this image). Note the dilation of the afferent vessels (right sylvian artery) and of the cortical drainage veins.

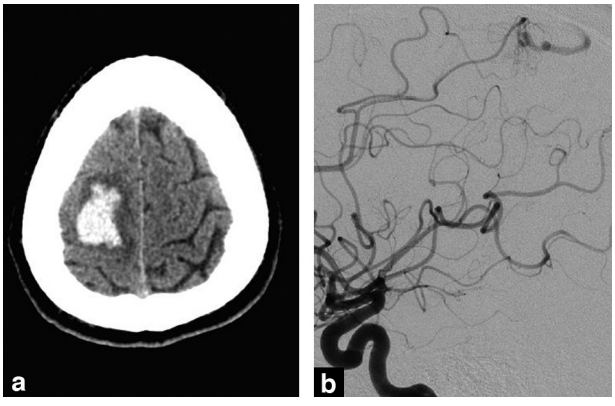

Figure 10. (a) Cortical haematoma, no AVM visualized on CT angiography. (b) Detection of a micronidus at the 3rd arteriographic follow-up at 8 months post-haemorrhage.

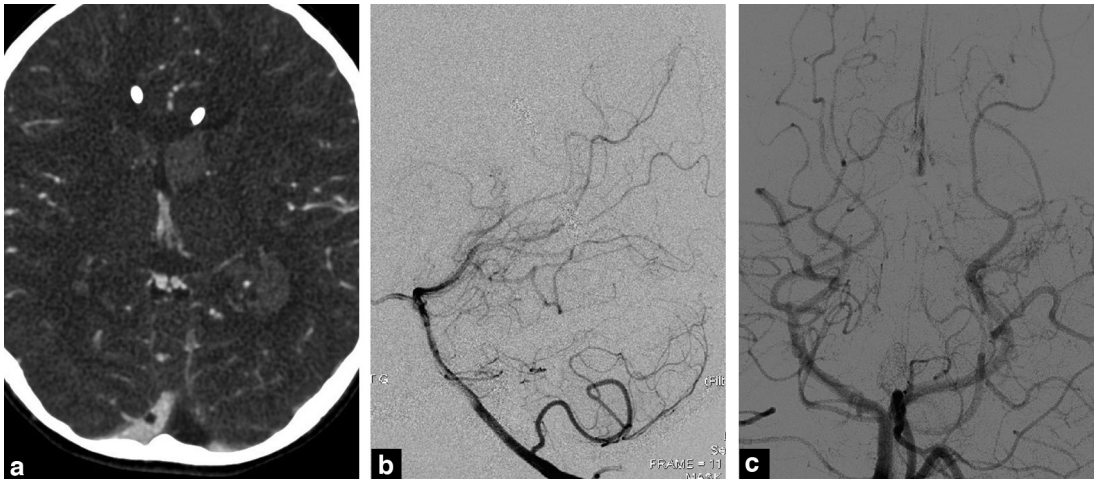

Figure 11. Internal parietal haematoma, 8-year-old child. The CT angiography (a) revealed an isolated vascular micro-ectasia in the acute phase. (b) Initial arteriography normal. (c) Check-up at one month: detection of a micronidus on a posterior-medial choroid branch of the left posterior cerebral network; note the early venous drainage of the right sinus.

allow the nidus to appear after resorption of the haematoma (Fig. 11).

Arterial aneurysms

These are linked to two phenomena: intraluminal hyperflow (mechanical factor) and anomalies of the parietal connective tissue (congenital factor) of the cerebral vessels participating in the same phenomenon as the genesis of the AVM itself [8]. Their presence has been demonstrated in certain studies as an independent risk factor for intracranial bleeds [6]. They are detected on MRA or TOF sequences, preferably on the raw slices for the detection of intra- or paranidal lesions. They are almost exclusively seen in cerebral AVM and not in dural AVM. There are three types of arterial aneurysms linked to cerebral AVM (Fig. 12):

- aneurysms of the circle of Willis: these are no different from classic intracranial aneurysms, other than that they are associated with the presence of the AVM. They may be single or multiple. They confer an added risk of haemorrhage as their own risk is added to the risk of the AVM. Thus their rupture (meningeal haemorrhage) may be seen independently of a rupture of the nidus (intracerebral haemorrhage). They may regress after treatment of the AVM (Fig. 12a and b). They can be treated at the same time as the AVM itself;
- paranidal aneurysms: located on the afferent arteries close to the nidus (Fig. 12c). They are of variable size and constitute a factor for potential fragility. They are more common on the cerebellar arterial network [9];
- intranidal aneurysms (Fig. 12d). Small in size, they are commonly seen in the acute post-bleed phase and can then form the point of rupture of the lesion (false aneurysm). They can be the object of targeted emergency treatment (partial embolisation) to secure the nidus before treating the lesion in a more global fashion. They are clearly identified as a factor that increases the risk of haemorrhage of AVM, unlike other aneurysms.

Venous aneurysms

These are linked to the mechanical factors of the local hyperflow. Located close to the nidus, or sometimes very far from it in the draining network, these venous ectasias are potential weak points and may justify the treatment of a lesion that was asymptomatic before diagnosis. One can observe signs of partial stress, such as an early thrombosis, signaling their potential danger. The use of 4D sequences is particularly useful as these make it possible to perfectly define the location of these lesions on the venous side of the shunt and thus to differentiate them from arterial aneurysms (Fig. 13).

Specific signs of dural AVM

These AVM, as we have seen, are essentially detected by a dural form of arterial recruitment and a dural and/or cortical venous drainage. The nidus is almost invisible as not very extensive. The radiological signs are therefore principally represented by the demonstration of the specific afferent meningeal and drainage network.

The afferent dural network

It is principally the dilation of the subcutaneous retromastoidian occipital network or of the posterior meningeal network (branch of the vertebral artery) at the internal base of the occipital bone that is observed, as sigmoid dural AVM are more common (classically resulting in pulsatile tinnitus) (Fig. 14). The appearance is that of an excessive local arterial vasculature in the subcutis. This sign is valid in both raw slices of MRA/TOF sequences and CT angiography. We also often see a trans-osseous arterial network at the base of the skull, evidence of the recruitment of the extra-cranial arteries towards the dural AVM. The middle meningeal artery is a very common contributor of dural AVM, but is difficult to demonstrate on imaging other than with arteriography. This artery, dilated in comparison with the healthy side, is detected on the medial side of the squama temporalis. Specific to rare dural AVM of the anterior aspect of the skull base, there is dilation of the transethmoidal meningeal arteries, originating from the facial artery (external carotid) and ophthalmic artery, fairly visible in the inferomedial corner of the orbit (angular artery). For dural

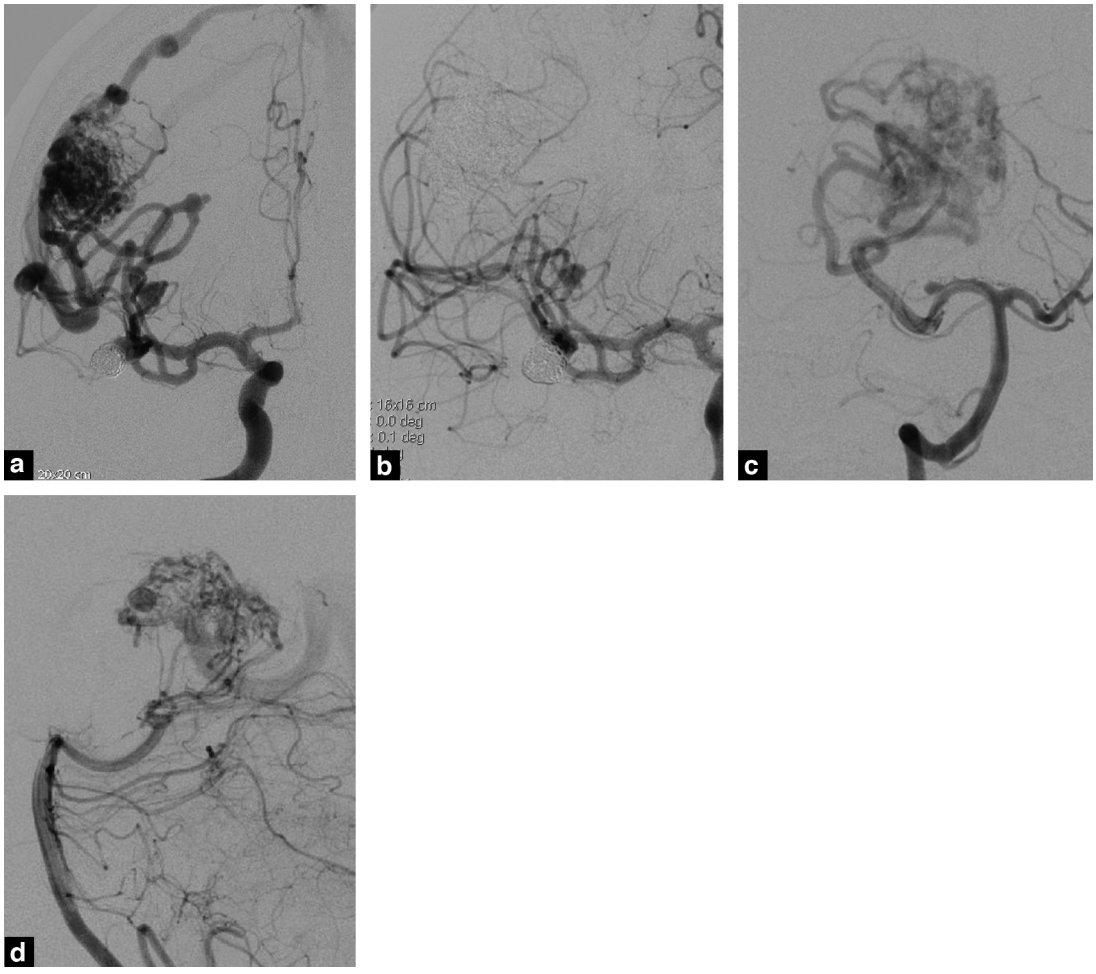

Figure 12. Arterial aneurysms. (a) Multiple sylvian aneurysms on the circle of Willis (one embolised) on a parietal AVM. (b) Regression of the aneurysm and of the arterial diameter after embolisation of the AVM. (c) Right posterior cerebral aneurysm on an occipital AVM. (d) Intranidal aneurysm on a choroid AVM.

AVM of the cavernous sinus, the afferent arterial network is generally poorly visible because of the shortness of its arterial feeders to the cavernous sinus. Cervical Doppler scan is sometimes used to detect signs of arteriovenous shunt in the cervical afferents (external carotid, occipital). One must look for alterations in the spectral profile, with the appearance of a diastolic flow on the external carotid network (giving an appearance of low vascular resistance). However, Doppler is all too often not sensitive enough and should ~~be~~ systematically be coupled with CT angiography or MRA.

The venous drainage system

Nosologically, the main characteristic of a dural AVM is that it usually primarily drains into a dural sinus, as the shunt is located in its own wall.

''Simple'' dural AVM

They are characterized in standard imaging by arterial signs (Fig. 14) as the sinus does not dilate and remains relatively normal looking. However, dynamic imaging with 4D MRA or 4D CT, is particularly helpful for diagnosis [10]. The use of these sequences should be systematic if there is a clinical suspicion of dural AVM [11]. The analysis is then made on the MIP projections of the acquisition volume where one looks for two signs; the abnormally early appearance of a dural sinus (radiological sign of a shunt) (Fig. 15) and a possible stenosis and/or venous thrombosis of a dural sinus.

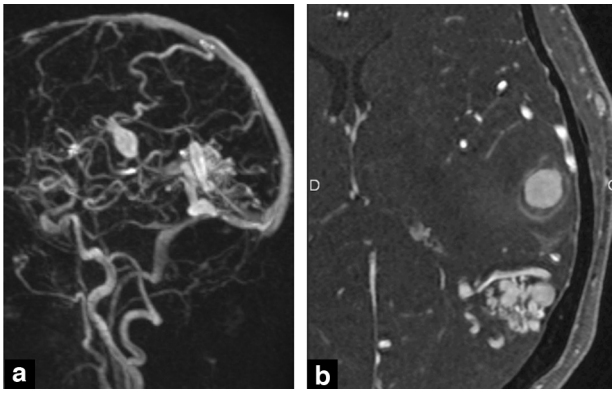

Figure 13. Inaugural temporal attack with no bleeding. (a) 4D MRI. Detection of a parietal AVM and distal cortical venous aneurysm at the temporal level. (b) Note the partial venous thrombosis of the aneurysm (TOF after gadolinium).

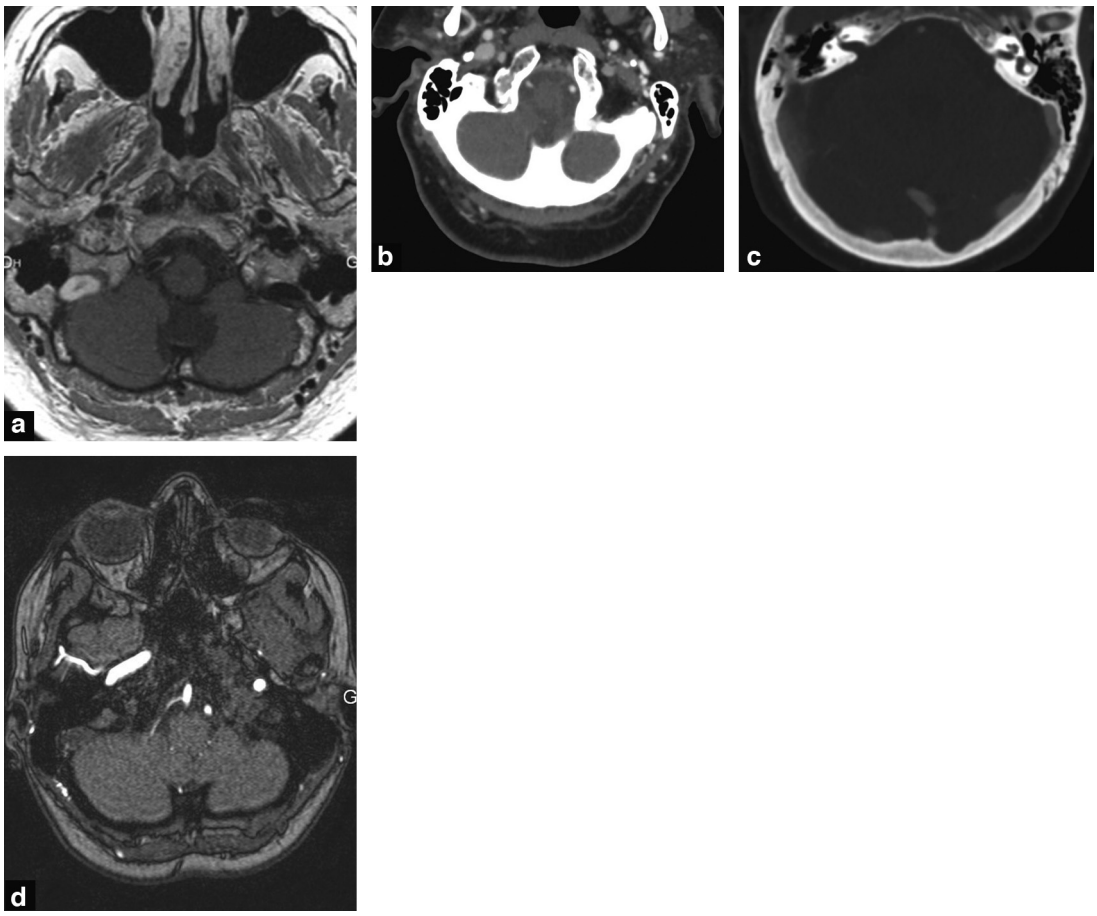

Figure 14. (a and b) Occipital subcutaneous dilation of the external carotid network (left occipital artery) on a dural AVM of the left sigmoid sinus. (c) Same patient: note the trans-osseous canals visible on the left occipital squama. (d) Dilation of the middle intracranial meningeal artery due to a right sigmoid dural AVM.

The existence of an old or recent dural thrombophlebitis is often associated with dural AVM as the genesis of the shunt is linked to the thrombophlebitis itself.

These dynamic MRA sequences should cover the entire skull if it is not known which side the AVM is on, or can be centered if a zone is already suspected, the latter allows increased spatial and temporal resolution. The sensitivity/specificity are markedly dependent on the temporal resolution of the acquisition to supply the maximum number of images, thus enabling clear differentiation of the arterial, parenchymal, and finally venous phases. However, the sensitivity of these 4D MRA may be low and their negative predictive value remains poor for small dural AVM. The use of arteriography should then be envisaged to avoid false negatives.

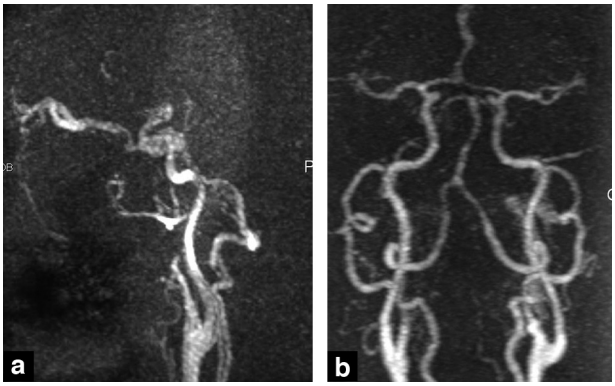

Figure 15. (a) Chemosis of the right eye. 4D MRA sequence (lateral MIP): early opacification of the cavernous sinus and of the drainage towards the superior ophthalmic vein (dural AVM of the cavernous sinus). (b) Pulsatile tinnitus on the left. Same sequence: early opacification of the right sigmoid sinus on a dural AVM of this sinus (frontal MIP).

Dangerous dural AVMs

The prognosis is, as with imaging, dependent on the involvement of the cortical veins and how abnormal they are. The flow within the shunt is then too important, surpassing the absorptive capacities of the dural sinus, the blood refluxes into the sinus and back into the cortical veins that are supposed to be emptying into the sinus. These fragile structures are consequently enduring arterial blood pressures, which results in venous oedema (neurological deficit, seizure, ocular congestion, headaches), or even haemorrhage. There again, MRA and CT angiography are essential for the diagnosis but the sensitivity on cortical venous drainage can be low and arteriography is generally required to determine the severity of the dural shunt. The most important sign to detect is obviously the opacification of a cortical

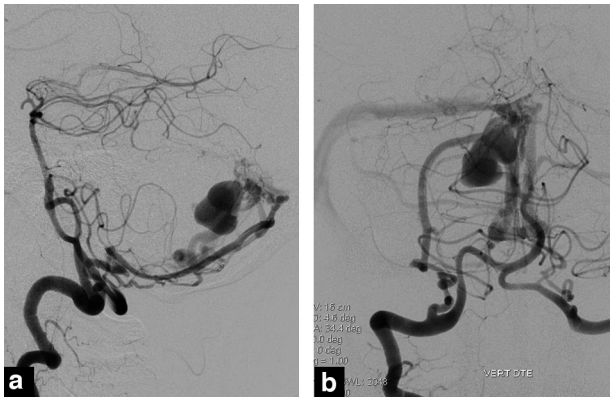

Figure 16. Angiography. Dural AVM of the torcular. Dural shunt developed on the sub-tentorial network of the right posterior-inferior cerebellar artery. Note the retrograde and aneurysmal drainage towards the cerebellar cortical veins (a) and the drainage towards the right lateral sinus (b).

vein in the arterial phase (Fig. 16). With dural AVM of the cavernous sinus, the opacification of one or more superior orbital veins does not represent a risk of bleeding, but carries a risk of glaucoma and opthalmoplegia if the fistula is not treated rapidly. With very high flow rate fistulas, the duro-cortical overload is such that one observes pathognomonic images of these congestive dural AVMs, with the abnormal visibility of masses of deep cortical veins with punctiform or ''vermicelli'' vascular patterns (Fig. 5). They are typical of congestive dural AVM responsible for sudden onset dementia, progressing to coma, if treatment is not instigated rapidly. Dural AVM of the base of the skull that overload the medullary drainage network (anterior spinal vein) are responsible for a similar congestive presentation, misleading at the medullary level, of progressive tetraparesis (Fig. 4).

Key elements of the report

The diagnosis of intracranial AVM is essentially based on cross-sectional imaging and dynamic 4D sequences, both in the haemorrhagic phase or unruptured AVM, with or without symptoms. The imaging criteria are central to therapeutic decision making in parallel with clinical criteria. The radiological report should therefore provide the following key elements:

- size: dimensions (height × width × depth in mm) and if possible an estimation of the volume in mm^3. Exclude the venous elements of the AVM as far as possible; the measurement should only include the nidus;
- location:
  - lobar (specify the topography),
  - cortical and/or deep,
  - infra- or supratentorial,
  - site of the shunt (dural sinus) for the dural AVM;
- type of venous drainage:
  - single or multiple,
  - deep and/or cortical,
  - for the dural AVM: look for an extra-cerebral venous drainage (orbital, medullary),
  - anomaly associated with a dural sinus for the dural AVM (thrombosis, stenosis, agenesis);
- afferent arterial system:
  - anterior, middle, or posterior cerebral artery,
  - branches of the external carotid;
- Spetzler-Martin grading system (cerebral AVM):
  - this enables a certain degree of appreciation of the therapeutic risk; (see Appendix 1);
- Cognard classification (dural AVM):
  - this angiographic classification is used to evaluate the prognostic risk of a dural AVM; (see Appendix 2);
- search for signs of severity:
  - recent or old haemorrhage,
  - mass syndrome, peri-lesional oedema that is hyper signal on T2 weight sequence, diffusion or FLAIR,
  - intranidal or distant thrombophlebitis,
  - venous aneurysms and possible signs of endoluminal thrombus,
  - arterial aneurysms,
  - presence of a venous stenosis on MRA,
  - NB: the association of a deep AVM, with single deep venous drainage in a young patient with a history of bleeding is a risk factor for more severe bleeding (33% risk in the first year).

Imaging report of an intracranial AVM

The imaging report is as follows:

- CT and CT angiography: emergency exam (bleeding, seizure). This is then completed with an MRI and a MRA. The latter is delayed with respect to the cerebral haematoma (2 to 8 weeks) to allow the time for resorption, so as not to mask all or part of the nidus. Ideally, CT angiography should cover the head and neck;
- MRI and MRA:
  - FLAIR (detection of oedema),
  - T2* gradient echo (detection of haematoma, cortical siderosis, venous thrombosis),
  - time of flight after gadolinium (focused on the suspected zone; improved detection of the nidus than an examination without contrast),
  - diffusion (detection of elevated ADC in the event of venous thrombosis),
  - 3D T1 after gadolinium in millimetric slices possibly in neuronavigation condition (advantage of MPR reconstructions enabling localisation of the AVM in the cerebral anatomy, advantage of neuronavigation with a view to stereotactic radiotherapy for dosimetric planning),
  - 4D sequence: analysis of the venous drainage, detection of a shunt, notably in dural AVM;
- cerebral angiography:
  - this is systematic in intracranial AVM to finalise the analysis of the lesion and notably the fine angio-architecture,
  - it can be delayed over time after a significant haematoma, which may mask the adjacent nidus via mass syndrome,
  - it may be performed in the event of a negative examination on MRA well conducted in the context of a dural

AVM where the clinical signs are patent (pulsatile tinnitus, persistent ocular redness and oedema),
- it is performed systematically after any idiopathic intracranial haemmorhage, in particular in the event of a haematoma with a lobar topography and should be repeated within 3 to 6 months after normal initial angiography.

TAKE-HOME MESSAGES

- The diagnosis of intracranial AVM may be achieved with CT angiography and MRA.
- The use of 4D sequences to detect the shunt, notably for dural AVM.
- Cerebral and dural AVM can be differentiated by their clinical presentation, prognosis, and architecture.
- Cerebral AVM: Pure intracranial arterial afferents towards a visible nidus. Venous dilation and frequent afferent aneurysms.
- Search for risk factors:
  - intranidal aneurysms, deep venous drainage, and deep localisation of the nidus,
  - detection of old haemorrhage, even clinically occult.
- Dural AVM: detection on clinical signs such as pulsatile tinnitus, chemosis, cerebral venous hypertension, cerebral haemorrhage. Indirect CT and MRA signs on venous dilation essentially. Invisible nidus.
- Search for signs of severity:
  - cortical venous drainage,
  - obstruction to the venous return.

Clinical case study

This 23-year old woman presented with a sudden weakness at her workplace. Loss of consciousness, generalized seizures, then coma necessitated intubation.

Unenhanced CT was performed as an emergency.

Questions

1. List the three anomalies detected on this CT scan (Fig. 17).

2. CT angiography was also performed (Fig. 18). What are the CT signs that are highly evocative of the diagnosis, and what is the diagnosis?

3. Angiography (including selective catheterization of the right posterior cerebral artery) was then performed (Fig. 19 a and b). What are the two anomalies?

Answers

1:
- Cerebral haemorrhage (right thalamus, cerebral peduncle).
- Intraventricular haemorrhage.
- Hydrocephalus.

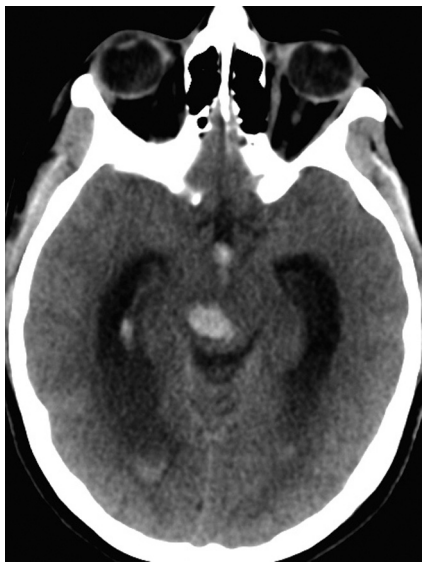

Figure 17. Unenhanced CT on admission.

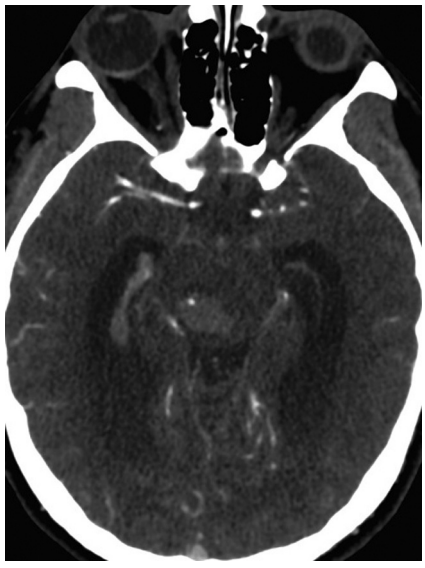

Figure 18. CT angiography at day 1.

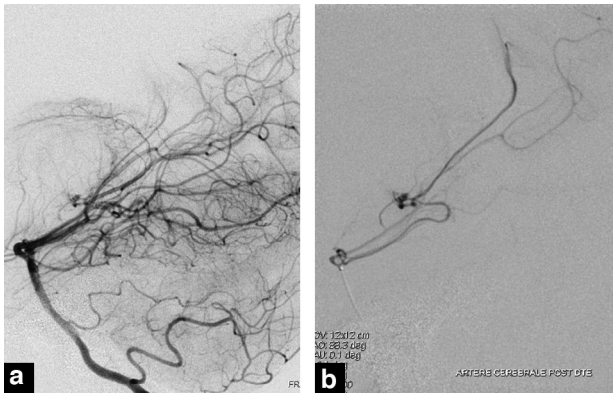

Figure 19. (a) Posterior vertebral angiography. (b) Selective catheterisation of the right posterior cerebral artery.

2:
- Punctiform vascular contrast enhancement, of 1 mm in diameter, located in the intracerebral haematoma.
- Suspected diagnosis: Ruptured cerebral AVM.

3:
- Detection of an arteriovenous shunt on the right posterior cerebral system at the level of the right posterior-medial choroidian artery by early opacification of a choroid vein joining the deep venous system (internal cerebral vein) around the right sinus.
- Detection of a micronidus.

Definitive diagnosis: Intracerebral and intraventricular haematoma consecutive to a ruptured micro-AVM of the right choroid system.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

Appendix 1. Spetzler-Martin grading system

Size:
- < 30 mm: 1 point
- 30—60 mm: 2 points
- < 60 mm: 3 points

Localisation in an eloquent area (functional):
- Localisation of the nidus in an eloquent area (functional): 1 point
  - Pre- or post-rolandic sensorimotor area
  - Language area
  - Visual cortex
  - Thalamus, hypothalamus
  - Internal capsule, cerebral peduncles
  - Cerebral trunk
  - Peduncles and deep cerebellar nuclei
- Any other zone: 0 point

Drainage:
- Deep only 1 point
- Any other drainage: 0 point

The grade is given by the number of points.

Appendix 2. Cognard classification

- Type 1: Shunt drains into a dural sinus (antegrade).
- Type 2a: Same as type 1 but with inversion of the direction of the drainage of the dural sinus (retrograde).
- Type 2b: Same as type 1 but reflex towards the cortical veins with retrograde circulation.
- Type 2a + b: Reflux into the dural sinus and into the cortical veins.
- Type 3: Shunt drains directly into a cortical vein.
- Type 4: Type 3 shunt with cortical venous aneurysms.
- Type 5: Shunt drains into the cervical medullary veins (anterior spinal vein).

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