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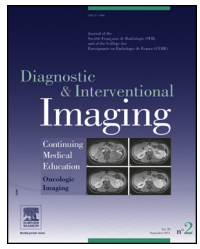

CONTINUING EDUCATION PROGRAM: FOCUS...

Intracranial arteriovenous malformations

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Cerebral arteriovenous malformation;
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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.
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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

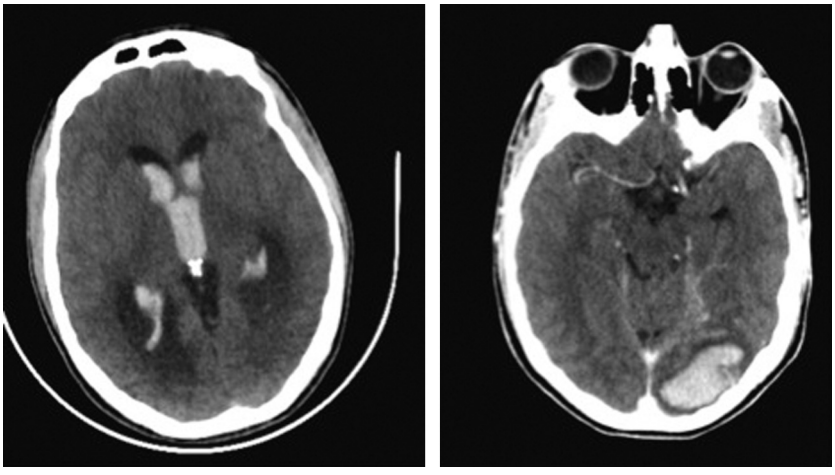

Figure 1. Two types of AVM ruptures in CT: on the left, intraventricular haemorrhage. One the right, left occipital cerebral haematoma (CT angiography).

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