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Case Report

A RARE CASE OF SUBTENTORIAL ARTERIO-VENOUS MALFORMATION: CASE REPORT

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Summary

The majority – over 80%, of arteriovenous malformations (AVMs) are supratentorial. The infratentorial AVM are uncommon and different from other intracranial AVMs in terms of history, diagnosis, treatment, prognosis, and follow up. The authors present a case of an intracranial hemorrhage, caused by rupture of an AVM in the posterior cranial fossa. Native (unenhanced) CT and computed tomography angiography (CTA) images of this rare location of AVM are presented.

Key words: subtentorial arteriovenous malformations, CTA

Introduction

There are four types of cerebral vascular malformations: parenchymal arteriovenous malformations (AVM), venous angiomata, cavernous angiomata and capillary teleangiectasias[1].

Of all types, AVMs are the most common symptomatic vascular malformations. They are congenital vascular pathologies due to underdeveloped intracranial capillary vessels in some local region [2]. AVMs consist of arterial feeders, tortuous ("serpentine") vascular connections with direct communication, the nidus and enlarged venous outflow channels [3]. There are no interposed capillaries in between the arterial and venous vessels. This results in direct arteriovenous shunting with increased blood flow through the feeding arteries and delivery of increased blood volume under high pressure to the draining veins [4].

Usually AVMs are located in the supratentorial compartment (more than 80%) and extend from the subpial surface of the brain to the deep white matter. Subtentorial AVMs are relatively rare lesions- they account for only 5-7 % of all intracranial AVMs [5]. Significant efforts have been made to achieve better understanding of the anatomic and haemodynamic complexity of AVMs, aiming to facilitate the diagnosis and decide on an effective treatment strategy. CTA, digital subtraction angiography (DSA) and MRA are the methods used to diagnose and follow up AVMs.

Case report

We present a case of a 55-year-old patient who was admitted to the emergency department with sudden onset of headache on the occipital side and double vision. Her blood pressure was high. She was seen by a neurologist and referred to the CT department for an initial native CT scan. The CT images showed the presence of an intracranial hemorrhage (ICH) in the posterior cranial fossa – at the site of the vermis of the cerebellum and in the pontine cisterns (Figure 1). CTA was performed immediately after that to further clarify the reason for the cerebellar parenchymal and subarachnoid hemorrhage. The CTA images showed there was a conglomerate of tortuous vessels and an enlarged vein vessel that directly communicated with the transversal sinus – an AVM with infratentorial location (Figure 2).

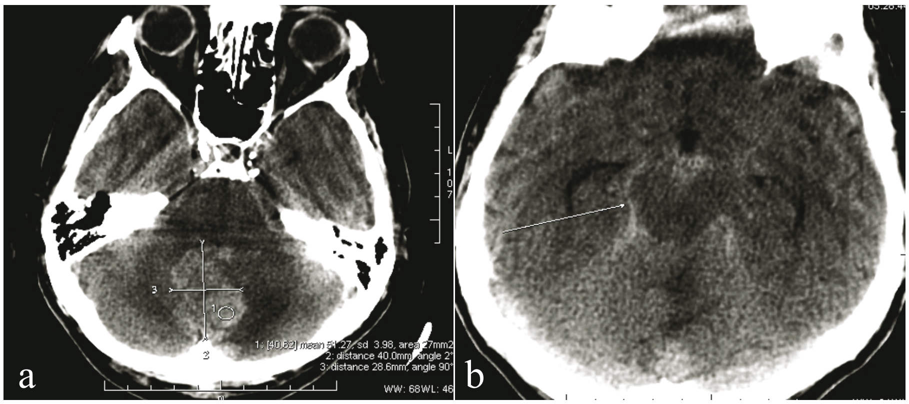

Figure 1. a. The initial native CT scan shows acute intracranial parenchyma hemorrhage in the vermis of the cerebellum. **b.** There is also bleeding into the pontine cisterns - SAH /subarachnoid hemorrhage/ with brain stem edema

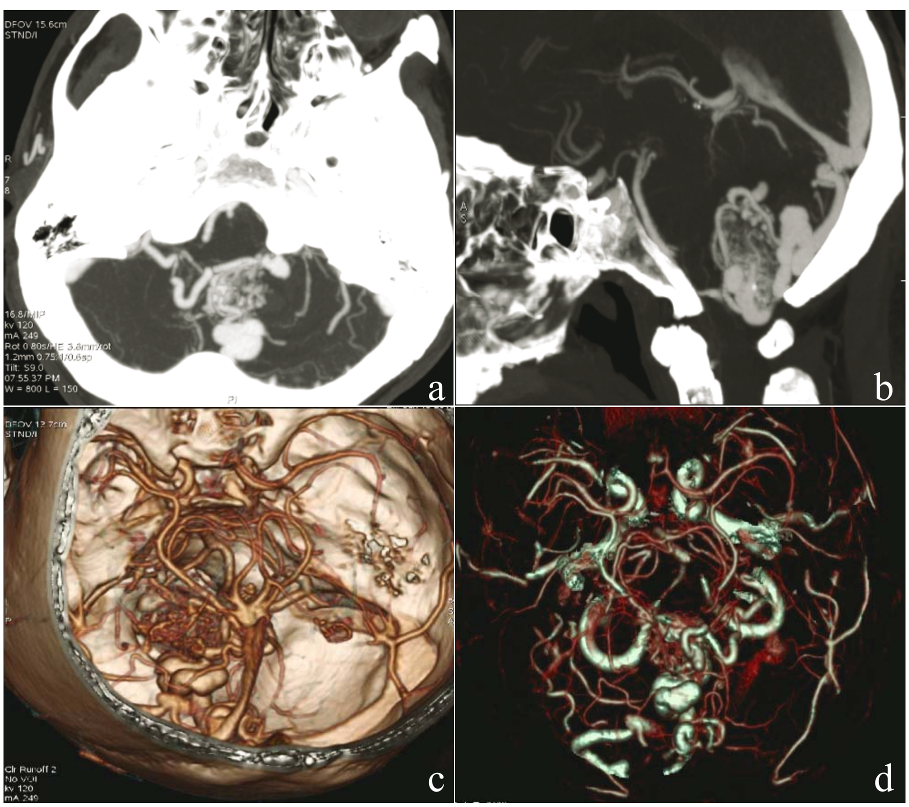

Figure 2. After perfoming CTA, the MIP axial and sagital images and the 3D Volume rendering images are demonstrating the infratentorial AVM with its nidus and enlarged draining vein to the confluence venous sinus.

Because the arterial feeding vessel could not be clearly identified on the CT images, conventional angiography was performed that proved the persistence of the subtentorial AVM with the feeding vessels from the left posterior inferior cerebellar artery /PICA/ and drainage with a massive venous varix to the confluent sinus.

According to the Spetzler-Martin score, the infratentorial AVM in the presented case report was IV-th grade and there was a major risk of permanent neurological deficits in this case (Table 1).

Table 1. According to the Spetzler-Martin score, the infratentorial AVM in the presented case report was IV-th grade (3 points: 2 for medium size and 1 for eloquence)

Size	Small (<3 cm)	1
	Medium (3-6 cm)	2
	Large (>6 cm)	3
Eloquence of adjacent brain	Non-eloquent	0
	Eloquent	1
Venous drainage	Superficial only	0
	Deep	1

According to the characteristics of the AVM, two therapeutic options were suggested: microsurgery or endovascular treatment. The latter was chosen (family decision) and embolization with onyx was performed after selective catheterization of the left vertebral artery. A total occlusion of the AVM nidus was achieved. In the early postoperative period a complication occurred - high intracranial pressure because of acute occlusive internal hydrocephaly. The neurosurgeons performed ventricle puncture with drainage of the lateral horn. After persistence of the hydrocephaly, a "Codman" ventricle-peritoneal system was attached that improved the condition of the patient.

Discussion

With the advances of modern neuroimaging, the frequency rate of posterior fossa AVMs has risen to 10-15 % [6-9]. The incidence of posterior fossa AVMs at autopsy is higher, reaching 20 % of all intracranial AVMs.

Bleeding is the most common form of presentation of infratentorial AVMs [10-12]. In most clinical series the incidence of subarachnoid and intraparenchimal bleeding has been reported as ranging from 75 to 92%. Progressive neurological deficits (including those to secondary mass effect, ischemia and hydrocephalies) were the second most common mode of presentation [14].

Complete neuroradiological evaluation of AVM includes initial CT, CTA, conventional angiography – digital subtraction angiography (DSA) and MRI (magnetic resonance angiography –MRA). Cross-sectional imaging like CT and MRA are used:

- in the initial workup of patients with hemorrhage for initial detection of AVM;
- for localizing AVMs in relation to the surrounding brain parenchyma;
- for providing volumetric information of AVMs (for nidus definition in radiation therapy);
- for assessment and follow up of treated patients.

Most patients with AVMs present with acute neurological symptoms from intracranial hemorrhage (ICH) and are initially assessed using native CT.

The next step is CTA examination that should evaluate not only the side of bleeding but also the circle of Willis to detect aneurysms and the course of the venous drainage path. Multi-planar volume reformats (MPVR), maximum intensity projection (MIP) and 3D volume rendering (3-D VR) images are used to detect asymmetrically enlarged vessels (the draining veins) and the nidus in relation to the hemorrhage. The arteries feeding the AVM are often not well discriminated within the vessel architecture [15]. DSA is the definitive diagnostic tool for detection and detailed characterization of the angioarchitecture of AVMs with its high spatial and temporal resolution and its ability to selectively evaluate the supplying arteries. However, DSA examination may be false negative if it is performed too early after the hemorrhage, because of compression of the nidus by the parenchymal hematoma [16].

Based on investigations of 206 patients with spontaneous ICH by CT and DSA, Zhu et al. [17] recommended considering DSA in all patients except those over 45 years old with pre-existing hypertension in the basal ganglia or posterior fossa hemorrhage.

A grading system by Spetzler and Martin [18] is used as a therapeutical decision-making tool, which gives information about the risk assessment of postoperative neurological deficit in AVMs.

The goal of management of AVM is complete angiographic obliteration, because the risk for hemorrhage (rehemorrhage) persists until the nidus has been completely obliterated [19-21]. A variety of therapeutic options are available to

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