REPETITIVE SUBARACHNOID HEMORRHAGE FROM A SPINAL CORD ARTERIOVENOUS MALFORMATION

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ABSTRACT

We present the case of a 32-year-old female who was admitted to the Neurology Clinic for occipital headache and severe cervical pain. Based on the first investigations (cerebral CT scan, cerebral MRI and cerebral angiography) the diagnose was intraventricular hemorrhage. One year later the patient was hospitalizad again for left cervical and left arm pain followed by motor déficit in the left arm and sensitive troubles in the left lower limb. Cervical MRI and spinal angiography revealed cervical intramedullary arteriovenous malformation (C7 – T1). The particular case points out on the importance of checking the spinal cord in cryptogenic subarachnoid hemorrhage.
Key words: cervical pain, subarachnoid hemorrhage, arteriovenous malformation

ANATOMICAL CONSIDERATIONS ON THE SPINAL VASCULATURE

Spinal arteries

The blood supply of the spinal cord is derived from a series of segmental vessels arising from the aorta and from branches of the subclavians and internal iliac arteries as follows: the cervical cord is vascularized by branches of the vertebral arteries, the thoracic and lumbar cord is nourished by segmental arteries from the aorta and internal iliac arteries while the sacral cord vascularization is supplied by segmental branches of the lateral sacral arteries. A segmental artery divides into an anterior ramus (most of them are small and never reach the spinal cord) and a posterior ramus which gives rise to a spinal artery.

The spinal arteries are radicular arteries that travel along the nerve root, join the cord forming an anterior and posterior axis and split into a cranial and caudal division.

The anterior axis supplies the anterior 2/3rds of the spinal cord and presents two radicular arteries of origin (for the cervical and thoracolumbar enlargement).

There are few anastomosis mentioned mainly outside the cord (conus medullaris - the arterial basket of Lazorthes) that are not able to supply the ischemia in one arterial territory.

a. Anterior spinal artery

- At the cervical level the spinal artery could have different patterns (bilateral, unilateral or intertransversal) and several origin points such as:
  - the intracranial segment of the vertebral artery
  - the external carotid artery (ascending pharingeal, occipital)
  - the ascending cervical artery
  - the deep cervical artery
- At the thoracolumbar level the origin of the spinal artery is arising from the aorta
- At the sacrococcigeal level the origin could derive from:
  - the internal iliac
  - the middle sacral arteries

b. The paired posterior spinal arteries which supplies the dorsal third of the cord by direct penetrating vessels and a plexus of pial vessels.

Spinal veins

The spinal venous system is divided into extradural and intradural venous system. The intrinsic veins originate into the deep gray matter, pass radially and are collected into extrinsic veins. There are two ventral spinal veins and one dorsal spinal vein with rich anastomotic system (including epidural system).

All spinal veins drain into radicular veins, traveling along the nerve root, with a final drainage

into the vertebral vein, vein azygos, ascending lumbosacral veins, vena innominata, superior cava vein, left renal vein and iliac veins.

Particularities of the spinal vasculature

The spinal vasculature presents few important particularities such as:

a. single small vessel (anterior spinal artery) feeds a long segment of the CNS
b. the pressure of the tissue drained by the veins varies tremendously
c. gray/white matter distribution is lined up in an inside/outside distribution, opposite to the brain.

VASCULAR MALFORMATIONS OF THE SPINAL CORD

- arterial aneurysms are usually rare and localized in the anterior spinal artery (could appear in association with the arteriovenous malformation or with the coarctation of the aorta). The subarachnoid hemorrhage is the most frequent presenting symptom and the angiography is confirming the diagnosis.
- cavernous angiomas are rare as well and could have the subarachnoid hemorrhage as the first symptom.
- arteriovenous malformations and arteriovenous fistulas are more frequent and could be classified by the size of the shunt (high flow or low flow) or by the location of the shunt (dural or intradural) [1].

Classification of the arteriovenous malformation and arteriovenous fistulas:

1. Dural AVF (Type I)

This type of fistula is frequently in men (40-70 years of life) and in 50% of cases is caused by trauma. From the clinical point of view the patients are developing slowly progressive paraparesis; the diagnose is confirmed by cervical MRI.

2. Intramedullary AVM (Type II, juvenile)

The malformation is frequent in young male patients and it is usually found in the upper cervical region. The clinical debut could consist into acute pain followed by quadriplegia suggestive for subarachnoid hemorrhage.

3. Perimedullary AVF (Type IV, III?, glomus ?)

This type of fistula is more large and complex and is consisting in multiple arterial feeders from more than one vertebral level. It is diagnosed more frequent in young male patients with slowly progressive paraparesis or acute onset and quadriplegia.

** Metameric AVM (Cobb's syndrome)*

Cobb's syndrome consists in an extramedullary malformation that tends to occur in the thoracolumbar area and cutaneous and osseous changes.

CASE PRESENTATION

We chose to report the case of a 32-year-old female with no personal medical history except for meningitis during childhood and frequent headache since the age of ten that was admitted in the Neurology Clinics for severe occipital headache and cervical pain started one week before. The neurological exam revealed mild nuchal rigidity without other neurological signs. The differential diagnose imposed the paraclinical evaluation was needed.

Lumbar puncture (normal eye funduscopy) revealed a CSF with hemorrhagic aspect, 291mg/dl proteins, 200/mmc leucocytes, 46 mg/dl glucose, erhytrocytes-countless *Cerebral CTscan* showed presence of the blood density in 3rd and 4th ventricle raising the suspicion of AVM in the choroid plexus.

Cerebral MRI showed the same aspect as in the CTscan.

Cerebral angiographic exam was normal.

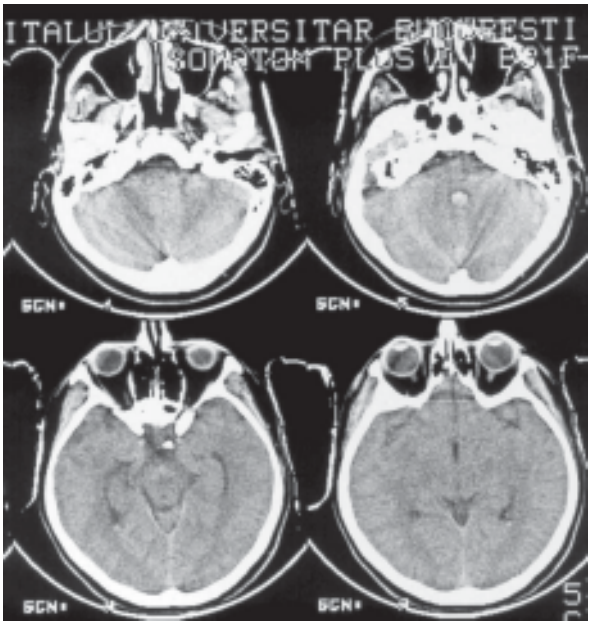
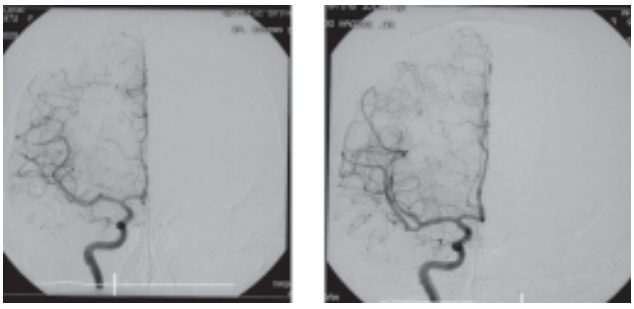
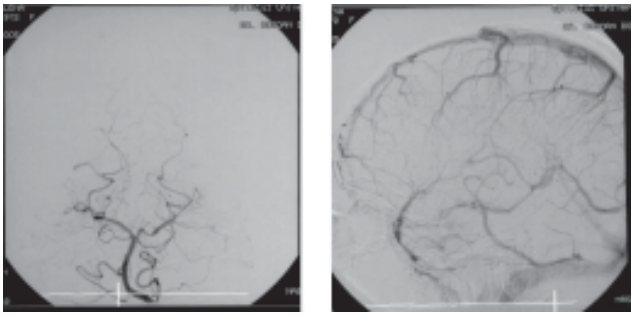


Figure 1. Cerebral CTscan. Presence of the blood density in the 3rd and 4th ventricle – AVM choroid plexus?



Right and left carotid axis



Vertebro-basilar system and venous system

Figure 2. *"Four vessels" cerebral angiography*

The patient was released from the hospital with the final diagnosis intraventricular hemorrhage, in good clinical condition.

One year later the patient is hospitalised again for few days for severe headache, with no neurological signs at the clinical exam, normal cerebral CT scan, difficult lumbar puncture with non-interpretable result and good clinical outcome after treatment with non-steroidal antiinflamatory drugs.

Six month later the patient returns into the emergency room for cervical and left arm pain and two days later she develops motor deficit on the left arm. The neurological exam revealed motor deficit, sensitive troubles and pyramidal signs on the left side of the body.

Cervical CTscan was performed which revealed a spinal cord left paramedian hemorrhage of 0,9/ 0,7 cm on the C4 – C7 level.

The cervical MRI diagnosed a cervical intramedullary AVM on the C7 - T1 level.

The spinal angiography confirmed the presence of the arteriovenous malformation corresponding to C7 – T1 with feeding artery from a cervical artery originating in right vertebral artery and rapid venous drainage by a vein which has an posterior aneurysmal dilation at C5 [2].

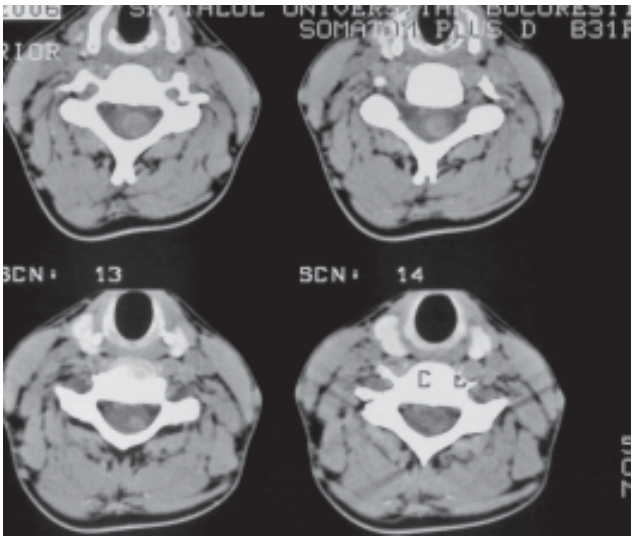
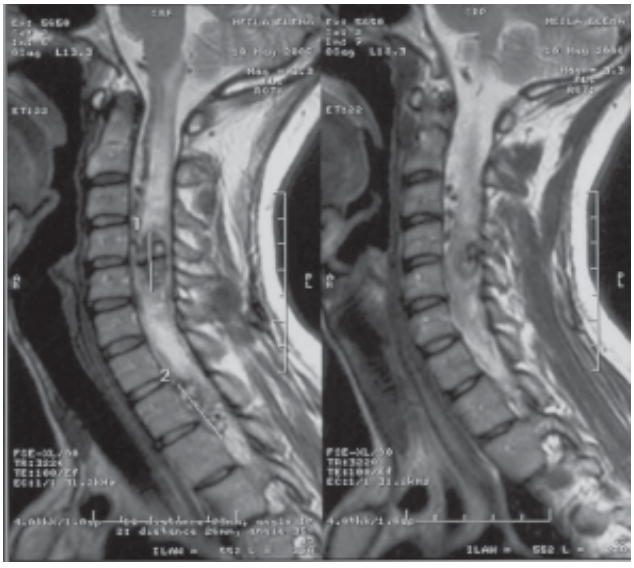


Figure 3. *Cervical CTscan. Spinal cord left paramedian hemorrhage C4 – C7*



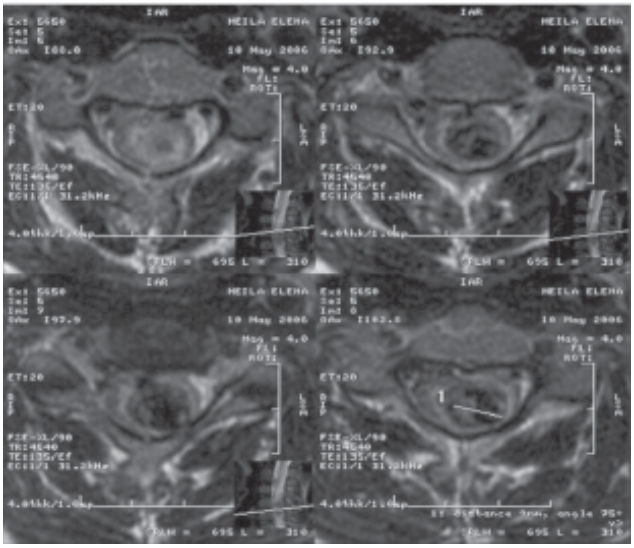


Figure 4. *Cervical MRI. Cervical intramedullary AVM (C7 - T1)*

Our final diagnosis was: Cervical spinal cord hemorrhage. Spinal arteriovenous malformation type II.

THE TREATMENT OF THE VASCULAR MALFORMATIONS OF THE SPINAL CORD

The treatment of the vascular malformations of the spinal cord consists in endovascular obliteration of the fistula or nidus or in surgical approach.

The endovascular obliteration of the fistula or nidus has two main objectives since the obliteration of the anterior spinal artery can lead to spinal cord infarction:

1. To prevent devastating bleeding or rebleeding of perimedullary AVM into the subarachnoid space or of intramedullary AVF into the parenchyma; dural AVF do not bleed.
2. To treat venous hypertension. It has been observed a good improvement after decreasing the venous pressure in weeks to months. Since progressive venous thrombosis after embolization is reported to occure, anti-coagulation is indicated.

Method:

The frequently used agent is acrylate and injection with amital and lidocaine through a catheter and control by SEP and MEP can be useful to assess the consequences of embolization.

Large AVMs usually do not benefit from embolization.

The surgical treatment has indications that need to be respected:

1. AVM under 2 vertebral bodies, located posteriorly and medially, away from the anterior spinal artery
2. in cases of recurrence after embolization

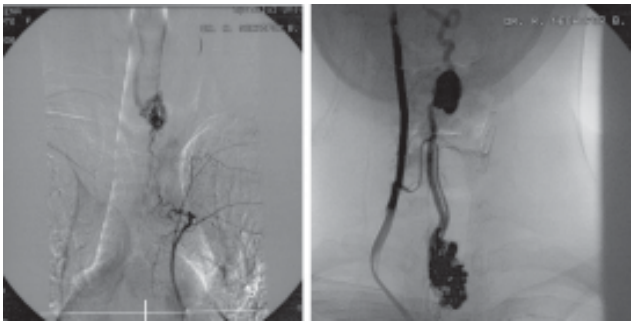


Figure 5. *Spinal angiography. AVM corresponding to C7-T1*

CONCLUSION

The patient's AVM is too large to benefit from embolization or surgery and therefore at any moment exist the danger of quadriplegia. At the release from the hospital the patient recovered completely the leg function and partially the arm function.

The presented case points out the importance of checking for spinal arterio-venous malformation when there is no cerebral explanation for a subarachnoid hemorrhage.

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