Treatment of Type III Dural Arteriovenous Malformation: Correlation with Neuropsychological Disturbances

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Introduction

The drainage pattern of dural arteriovenous malformations (dAVM) determines clinical symptomatology and the risk of hemorrhage¹⁻³. The retrograde transmission of the arterial pressure in the venous network in combination with impaired venous outflow through the sinuses can lead to extreme venous hypertension. These angiographic features are strongly related with global cognitive dysfunction⁴.

We present a patient with a slow onset of cognitive disturbances followed by a rapid deterioration in whom neuropsychological examination was performed before and after treatment of the dAVM.

Case Report

A 47-year-old male was known to have a history of deep venous thrombosis related to a protein C deficiency (1996) for which he was treated with anticoagulants.

From 1996 till 2000, six short periods of word-finding difficulties and a transient right-sided hemiparesis occurred, accompanied by headache localized in the right occipital region. These episodes were diagnosed as transient ischemic attacks. Loss of initiative, incapacity to

perform multiple tasks and disturbance of spatial orientation were noticed. Because of persisting aphasia he was hospitalized August 2000. Neurological examination revealed a mixed aphasia, right-sided hemianopia, and right-sided apraxia. Ten days later, he developed a postural and intention tremor followed by apathy and bradyphrenia.

The intensity of these deficits fluctuated during the day. Neuropsychological examination of general cognitive functioning (Wechsler Adult Intelligence Scale-Revised (WAIS-R)⁵), memory (Wechsler Memory Scale (WMS)⁵) and constructional ability (Drawings to Command Test and the Reproduction Drawings Test⁵) showed a severe global cognitive deterioration with pronounced amnesia, aphasia and impaired executive functioning. The corresponding WAIS-R full-scale IQ was 72 (verbal IQ = 93, performance IQ = 52), whereas the WMS full-scale memory quotient was 62. Moreover, constructional ability was severely impaired (figure 1).

A CT scan performed at admission showed focal areas of increased density in both occipital lobes. The peri-ventricular white matter showed increased signal intensity on T2 weighted images (FLAIR) due to edema (figure 2A). Throughout the brain parenchyma, dilated ves-

Figure 1 Examples of the Drawings to Command Test (clock 11:10 hour, tree) and the Reproduction Drawings Test (three-dimensional block and two-dimensional cross) before (left) and after closure of the dAVM. Notice the moderate to severe distortions with nonrecognizable drawings, gross distortion of the basic gestalt, perseverations and loss of three-dimensionality before treatment and the almost full recovery of constructional ability after treatment.

sels were present (figure 2B). Phase contrast MR angiography showed hypervascularity in the occipital region. Cerebral angiography with

selective injection of the occipital arteries revealed a dAVM in the right transverse sinus with extension in the torcular (figure 3A). Venous drainage was retrograde into the straight and superior sagittal sinus (figure 3B). From both sinuses, the entire venous network of the brain, up to the frontal areas, was filled retrogradely (figure 3C).

Because both sigmoid sinuses were occluded, the only venous exit was through the cavernous sinus towards the pterygoid plexuses. Selective injections of both internal carotid arteries showed delayed venous drainage. Again, the only venous exit was through the cavernous sinus (figure 3D).

Endovascular treatment was performed, with the aim to close the transverse sinus at the level of the fistulous point of the dAVM. Because both sigmoid sinuses were occluded, a direct catheterization of the superior sagittal sinus was performed through a borehole in the vault, just above the superior sagittal sinus (figure 4A). A microcatheter (Turbo Tracker 18, Boston Scientific) was positioned in the transverse sinus.

Figure 2 A) T2 weighted (FLAIR) image shows increased signal intensity in the centrum semiovale. B) Gadolinium enhanced T1 weighted image shows multiple dilated vessels throughout the whole brain parenchyma. Areas of flow void are present in the occipital region.

Figure 3 A) Selective injection of the left occipital artery (PA projection) shows dural AVM in the region of the transverse sinus and torcular. Reflux is noticed in the superior sagittal and straight sinus as well as in a leptomeningeal vein. B) Selective injection of left occipital artery (lateral projection) shows massive reflux in the entire venous system. C) Late venous phase of left occipital injection (lateral projection) shows final drainage through the cavernous sinus towards the extracranial venous system. D) Late venous phase of left internal carotid artery injection (lateral projection) shows delayed transit of contrast (27 seconds after starting of injection of contrast). The only venous exit is the cavernous sinus.

This segment of the sinus was completely occluded by packing with coils. Control angiography showed complete closure of the dAVM and normalization of the pace of venous drainage (figure 4B).

A control neuropsychological investigation

was performed in January 2001, with significant improvement of cognitive functions. The WAIS-R full-scale IQ and the WMS full-scale memory quotient were now 116 (verbal IQ = 119, performance IQ = 110) and 114, respectively. These indices were almost compatible

Figure 4 A) Injection of right occipital artery with 4F sheath positioned in the superior sagittal sinus. The microcatheter tip is positioned in the transverse sinus. B) Left internal carotid artery injection after obliteration with coils of the right-sided transverse sinus shows significant improvement of venous drainage. The venous phase is now reached in 11 seconds.

with the estimated pre-morbid situation based on patient's education and occupation (estimated pre-morbid full-scale IQ and WMS memory quotient both over 110).

MR imaging performed in the same period showed reduction of venous ectasias in the occipital region (figure 5A). The periventricular white matter showed unchanged high signal intensity on the FLAIR images (figure 5B). A control angiography performed four months later, confirmed complete closure of the dAVM.

Discussion

Dural AVMs are acquired lesions, probably caused by enlargement of pre-existing micro-shunts in the dura, in the presence of increased arterial or venous pressure. Two classifications of dAVMs have been proposed^{1,3}. The common concept of both classifications is to differentiate between lesions with antegrade drainage in the dural sinus (benign type dAVM), and dAVMs causing retrograde drainage in leptomeningeal veins.

The importance of the drainage pattern lies in the correlation with clinical symptoms and

the risks of hemorrhage². The retrograde transmission of the arterial pressure in the venous network leads to venous hypertension and congestion and impairs parenchymal venous drainage.

This leads to chronic (venous) ischemia⁴. In those cases where extensive reflux is seen in combination with impaired venous outflow through the sinuses, venous pressures can rise to very high levels. Both features were present in our patient and, as was reported previously⁴, this can be strongly related with global cognitive dysfunction.

Prior to treatment, venous reflux was seen angiographically in the straight and superior sagittal sinus, which led to impaired venous drainage of the brain parenchyma. Areas of high signal intensity on T2 weighted images are either compatible with edema⁶ or are due to gliosis caused by prolonged venous hypertension.

The impaired venous drainage is reflected in the delayed venous phase on the internal carotid injection. This impairment of venous drainage correlated well with the global cognitive deterioration. Four months after treatment, neuropsychological findings returned to

normal. MR imaging findings correlated only partially with the reversal of clinical symptoms. Ectasia of cortical veins diminished, however T2 weighted images (FLAIR) showed unchanged high signal intensity in the centrum semiovale, and are probably the result of gliosis. Although normalization of the pace of venous drainage on the internal carotid angiogram was seen, this does not imply that neuropsychological symptoms will resolve to the same extent.

The longstanding presence of reflux caused by the dAVM can cause irreversible damage to the brain.

Dural AVMs with retrograde drainage in leptomeningeal veins require aggressive treatment¹. Closure of the dAVM either surgically or through an endovascular approach is obligatory. Partial treatment will lead to recurrence of symptoms and will sometimes not prevent death^{4,7}. When partial treatment can be achieved through an arterial route this is preferable, but if this can not be accomplished, occlusion of the sinus at the fistulous point is necessary.

This can only be performed when the normal venous drainage of the brain is respected. If the sinus is no longer used for the drainage of the brain parenchyma, and posterior fossa veins do not enter the diseased segment the sinus can be sacrificed. In patients with occlusion of the affected sinus at both proximal and distal sites (isolated sinus), an alternative route to the diseased sinus has to be chosen⁸.

We choose the least invasive surgical approach to the sinus. Through a small bore-hole in the midline of the vault, the superior sagittal sinus was punctured then a 4F sheath was positioned. This allowed direct access to the transverse sinus and unrestricted closure of the sinus was achieved.

Conclusions

In patients with severe cognitive disturbances due to a dural AVM with leptomeningeal reflux and impaired venous drainage, complete elimination of the reflux has to be obtained to reduce the venous hypertension.

Even in cases of severe cognitive disturbances, closure of the dAVM can result in significant improvement.

Figure 5 A) Gadolinium enhanced T1 weighted image 5 months after closure of the dAVM shows marked reduction of the venous ectasias. B) T2 weighted (FLAIR) image 5 months after closure of the fistula shows unchanged high signal intensity in the centrum semiovale.

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