Reversible tonsillar prolapse and syringomyelia after embolization of a tectal arteriovenous malformation

Case report and review of the literature

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The authors report the case of a 21-year-old woman who presented with headaches, frequent sensations of loss of equilibrium, and intermittent strabismus. A tectal arteriovenous malformation (AVM) was diagnosed based on magnetic resonance (MR) imaging findings. The AVM drained toward the straight sinus and was associated with a tonsillar prolapse (Chiari malformation Type I [CM-I]) and cervical syringomyelia. The tectal AVM was embolized with N-butyl cyanoacrylate, and disconnection of about 80% of the lesion was obtained. All clinical symptoms resolved after embolization, and radiosurgery was proposed to treat the malformation remnant. A control MR image confirmed the regression of the tonsillar prolapse and the disappearance of the syrinx. This report emphasizes that CM-I and syringomyelia may be acquired and related to hydrovenous disorders. (DOI: 10.3171/JNS-07/08/0412)

Key Words • arteriovenous malformation • Chiari malformation Type I • syringomyelia • venous hypertension

Chiari malformation Type I and syringomyelia are often linked, and their association has been explained as the consequence of long-lasting obstruction of the subarachnoid space at the foramen magnum, creating differences of flow of CSF between the spinal and the intracranial subarachnoid compartments. Chiari malformation Type I is often considered a congenital condition that exists at birth and gives rise to secondary complications; however, several acquired cases of various origins have been recently described.2–4,8,15,18,21 We report the case of an adult patient with CM-I and syringomyelia associated with a tectal AVM, both of which resolved spontaneously after endovascular treatment of the vascular malformation.

Case Report

Presentation and Examination. This 21-year-old woman presented to the Department of Neurosurgery because of headaches and intermittent sensations of loss of equilibrium. She had been seen by an ophthalmologist for episodes of strabismus unrelated to the headaches. Her medical history was remarkable for premature birth (at 6 months’ gestation), although she did not suffer any further complications. Her neurological examination revealed normal findings.

An unenhanced computed tomography scan of the brain showed supratentorial ventricular dilation without any sign of raised intracranial pressure, and a vascular lesion at the posterior part of the third ventricle was suspected. A large tectal AVM was diagnosed based on MR images. The AVM drained toward the straight sinus and was associated with tonsillar prolapse, diagnosed as a CM-I, and cervical syringomyelia extending from C-2 to C-7 (Fig. 1A–C).

Treatment. The patient’s complaints were considered to be related to the AVM and not to the syringomyelia or the CM-I. It was thus decided to treat the tectal lesion first, and embolization was chosen as the therapeutic option. Angiography showed that the quadrigeminal AVM was vascularized by tectal and circumferential arteries arising from the vertebrobasilar system and draining into the straight sinus via dilated tectal and superior cerebellar veins (Fig. 1D and E). Secondary congestion of the posterior fossa caused mainly by the left hemispheric cerebellar veins was noted. Between May 2004 and February 2006 the patient underwent four sessions of endovascular therapy in which N-butyl cyanoacrylate glue (Braun Melsungen AB) was used as the embolic agent. The final postembolization angiogram showed a reduction of about 80% of the AVM size and flow (Fig. 2A and B).

Posttreatment Course. The patient’s headaches improved after the first session of treatment, and after the second treatment they fully disappeared. She did not experience
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another episode of strabismus, and findings from her neurological examination remained normal. A control MR image confirmed the reduction of the AVM as well as regression of the supratentorial ventricular dilation (Fig. 2C), tonsillar prolapse (Fig. 2D), and syringomyelia (Fig. 2E).

As no further endovascular treatment could be used to treat the AVM remnant, we decided that the patient should undergo radiosurgery to obtain a cure of the lesion.

Discussion

Brain AVMs have been thought to be congenital vascular lesions. This belief has recently been revised, however, and AVMs are recognized as being acquired lesions caused by endothelial cell disorders. Secondary triggers set off the cellular disease, and angiogenesis and postnatal matura-

mation of the intracranial venous system play a role in the onset of these lesions. The consequences of the malformation are often related to hydrovenous disorders that will vary according to the age of the patient at the time of the diagnosis (that is, “melting brain” is often found in neonates, and seizures, deficits, or hemorrhages occur in adults). Posterior fossa hydrovenous disorders responsible for tonsillar prolapse have been reported in vein of Galen AVMs in infants as the result of inadequate venous drainage. Chiari malformation Type I of vascular origin is reversible if adequate venous drainage can be reconstituted following proper embolization of the AVM.

Chiari malformation Type I is a condition defined by caudal descent of the cerebellar tonsils through the foramen magnum. It is often considered to be congenital, but several cases of acquired CM-I have been reported. This malformation is frequently associated with syringomyelia. The pathophysiology of the latter is still a matter of debate. If both CM-I and syringomyelia are associated, it is important to diagnose which condition is responsible for the patient’s symptoms and manage it first. Our patient did not present with any neurological sign that could be caused by the tonsillar prolapse or the syrinx. We considered that the hydrovenous disorder associated with the acquired AVM created the CM-I that secondarily caused the syringomyelia. Intracranial mass effect caused by an AVM has been considered to be responsible for ventricular dilation by compression of the aqueduct, and to result in CM-I be-
cause it was believed to force the tonsils to protrude into the cervical canal.\textsuperscript{16} We do not feel that this can be considered a proper explanation in our patient as MR imaging failed to show any clear imprint of the tectal AVM on the surrounding structures. In the same way, this mechanical phenomenon has not been considered the main cause of the ventriculomegaly detected in children with vein of Galen AVMs reported by Girard et al.\textsuperscript{4} The experience of other groups\textsuperscript{4,9,22} confirms these data.

The headaches and gaze palsy in our patient were thought to be related to the tectal AVM as well as the venous congestion of the quadrigeminal plaque, leading to dysfunction of the tectal and mesencephalic structures involved in eye movements. Thus we decided to first manage the AVM. We thought that the endovascular approach was more appropriate for safe reduction of the malformation and rapid resolution of the venous congestion and of its consequences. The clinical improvement already noticed after the first session of embolization confirmed that the pathophysiological theory that had been considered was correct.

To our knowledge, this is the first case of reversible tonsillar prolapse and syringomyelia related to an intracranial AVM occurring in an adult. Our findings confirm that the features seen in our patient may be transient in adults as well as children, if managed properly. In a previous paper, O’Shaughnessy et al.\textsuperscript{18} reported on a patient in whom an acquired form of CM-I (without any syringomyelia) was diagnosed after the development of an AVM and severe restriction of the sinus venous outflow. Our case confirms that both CM-I and syringomyelia can be exclusively related to venous hypertension in the posterior fossa, even without any sinus involvement.

Conclusions

Chiari malformation Type I and syringomyelia may be acquired (and reversible) lesions when associated with intracranial AVMs. In our patient, the tectal AVM is certainly the first lesion that appeared. The venous congestion of the posterior fossa had created the tonsillar prolapse that was at the origin of the syrinx because of the disturbances of the CSF circulation. Proper understanding of the pathophysiological cascade giving rise to both symptoms and secondary lesions allows proper treatment of the patient with low therapeutic risks.

References

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