Clinical case

Coincidental vascular anomalies at the foramen magnum: Dural arteriovenous fistula and high flow aneurysm on perimedullary fistula

Coexistence de deux anomalies vasculaires de la jonction cranio-cervicale : fistule durale artério-veineuse et anévrisme de l’artère spinale antérieure

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ABSTRACT

We report the case of a 59-year-old woman admitted for a sudden headache due to a subarachnoid haemorrhage. On CT scan, the clots predominated into the posterior fossa without high-density in the sylvian or interhemispheric fissures. The vertebral angiography revealed a dural arteriovenous fistula at the foramen magnum associated to an aneurysm of the cervical anterior spinal artery. Due to the high rebleeding risk of a dural shunt, we proposed curative treatment using microsurgical interruption of the intradural draining vein. On the postoperative angiography at 15-day follow-up, the 2 malformations were corrected and the outcome at 6 months was excellent. Based on the literature, we assess this exceptional association and suggest its possible management.

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1. Introduction

Dural arteriovenous fistulas (DAVFs) at the foramen magnum (FM) are located in the dura mater of the foramen magnum supplied by meningeal branches from the vertebral artery, the occipital artery or the ascending pharyngeal artery [1,2]. In contrast to spinal DAVFs essentially responsible for myelopathy, the common characteristics of this location at the cranio cervical junction were the subarachnoid haemorrhage (SAH) as a diagnostic circumstance and a high flow status [3–11]. The association with other vascular anomalies and the management at this location have rarely been reported in the literature [9,12]. In one patient, the reported coexistence of a DAFV at the FM with an aneurysm on the anterior spinal artery (ASA) prompted us to investigate the relationship between both malformations.

2. Case report

A 59-year-old woman, with no previous medical history, was admitted to the emergency department for a sudden headache. On clinical examination, she presented with an isolated meningismus without focal signs. On CT scan without contrast (Fig. 1) performed in emergency, the blood predominated in the peribulbar cistern of the posterior fossa without reflux into the fourth ventricle. No high-density area was observed in the sylvian or interhemispheric fissures. The SAH was classified as Fisher scale type III. The diagnosis of SAH grade I according to the World Federation of Neurosurgery Scale (WFNS) was confirmed. The CT angiogram of the head and neck showed extensive SAH in the upper cervical region as well as the basal cisterns and dilated vessel anterior to the medulla oblongata with an oblique course. No vascular malformation was observed. Comprehensive cerebral and spinal cord angiography was performed. Injection of the left vertebral artery revealed a DAVF at the FM fed by meningeal branches from the vertebral artery at the dural penetration level and drained intracranially to the superior petrous sinus via a tortuous enlarged vein (Fig. 2B). Careful injection of the right vertebral artery showed a fusiform aneurysm measuring 2 mm in diameter located on the ASA at the C1 level (Fig. 2A), which depicted the DAVF at the FM. Thus, the ASA fed the DAVF via the left radiculopial artery of C1. These findings were consistent with a category V of a DAVF at the FM fed by the left vertebral artery and the high cervical ASA via the left radiculopial artery of C1.

The surgical treatment of the malformation was performed 17 days after the bleeding. The patient was placed in semirecumbent position. A lateral suboccipital craniectomy with haemilaminectomy of C1 allowed a limited longitudinal incision of the dura mater in a curve leading to an adequate exposition of the left vertebral artery at its dural penetration. Via this dorsal view, a distended arterialized vein was exposed just distal to the dural penetration of the vertebral artery. The dural arteriovenous fistula was treated via the right vertebral artery. The ASA was occluded by a detachable balloon catheter. The vertebral artery was then ligated and cut. The patient was discharged 10 days later in good condition.

![Fig. 1. Admission CT scan without contrast showing a subarachnoid hemorrhage classified as Fisher scale type III in the peribulbar cistern of the posterior fossa without reflux into the fourth ventricle nor blood in the sylvian or interhemispheric fissures. Scanner sans injection réalisé à l’admission montrant une hémorragie sous-arachnoïdienne grade III de la classificaton de Fisher prédominant dans la citerne péribulbaire sans contamination ventriculaire ou de la vallée sylvienne.](image1)

![Fig. 2. Preoperative angiography. A. Showing 1: a 5 mm aneurysm of the anterior spinal artery. B. Showing 1: the draining vein; 2: the nidus of the arteriovenous fistula of the craniovertebral junction next to the V3-V4 segments. Angiographie préopératoire. A. Montrant 1 : un anévrisme de 5 mm de l’artère spinale antérieure B. Montrant 1 : la veine de drainage de la fistule duree artério-veineuse ; 2 : le nidus de la fistule duree artério-veineuse de la jonction craniovertebrale en regard des segments V3-V4.](image2)
penetration of the left vertebral artery. This vein coursed obliquely upwards to the anterior surface of the medulla oblongata and the protuberance. Using an indocyanine green video-angiography, the disappearance of the DAVF by temporary clipping of the draining vein was confirmed. A 5 mm straight clip was applied interrupting the intracranial venous part of the malformation just distal to the left vertebral artery (Fig. 3). We also confirmed a change in the colour of the arterialized vein from red to blue, but the entire course of the vein could not be observed due to its anterior location to the brainstem. The patient subsequently improved recovering a normal physiological status. The postoperative angiography confirmed the complete exclusion of the DAVF and of the ASA aneurysm (Fig. 4A and B). At discharge, there was no complication. At 6 months clinical follow-up, the patient’s quality of life was excellent and the patient returned to a normal life without restriction. No recurrence of the fistulas was confirmed on magnetic resonance imaging at 6 months after the surgery.

3. Discussion

The coexistence of vascular anomalies associated to DAVFs is a presumed assertion. The incidence of coexisting vascular anomalies was estimated for cerebral DAVFs to be approximately 20% [13], and for the spinal DAVFs 2% essentially in the thoracolumbar region [14]. Nevertheless, for the cervical spinal DAVF, a rate of 41.7% of an associated complex vascular anomaly has been reported [12]. Similarly at the craniocervical junction, a rate of 37.5% with an associated vascular anomaly was recently published [9]. At the craniocervical junction, the incidence of vascular anomalies coexistence is probably underestimated because of the difficulties to perform complete angiographic explorations. As in our case, the associated vascular lesion on the ASA was revealed by a careful slow injection of a contrast agent into the contralateral vertebral artery.

Initially, we had interpreted the lesion as a fusiform aneurysm on the ASA. This interpretation is subject to discussion. In reality, this lesion may be included in a perimedullary arteriovenous fistula (PAVF) located on the anterior surface of the upper cervical spine. Moreover, the spinal aneurysm could be located on the lateral spinal artery as previously described by Lasjaunias et al. [15]. Arterial aneurysms have been observed in 80% of these associated PAVFs [9]. The development of the aneurysm may result from the combination of haemodynamic and genetic factors. The PAVF may cause arterial change due to the high flow related to the arterial feeders supplying the shunt [16]. Moreover, the arterial susceptibility could be related to the embryologic homology of this PAVF to the presence of the DAVF at the same metameric level. Indeed, this combination of both vascular malformations confirmed the hypothesis of spinal arteriovenous metameric syndrome as previously reported by Rodesch et al. [17].

The incidence of SAH as a diagnostic circumstance was estimated to be approximately 45% for these DAVFs of the cranio-
cervical junction [18]. The mechanism underlying the occurrence of SAH has not been completely understood [5,10,18,19], although several characteristics have been associated with this clinical event:

- the spine location on cervical or craniocervical junction in contrast to the thoracolumbar exceptionally responsible for this condition;
- the variecal enlargement of perimedullary veins;
- the rostral direction of the intracranial venous drainage.

The venous hypertension, determining the arteriaealization of the medullary veins and the valveless pial coronal venous plexus with an increase in venous pressure [20], is probably involved in the occurrence of bleeding. However, this angioarchitectural factor, insufficient for the thoracolumbar region, becomes effective for the DAVFs located at the junction. In some case series [1,5,9,12], the drainage pathways appeared as a small size despite the feeding arteries with a high flow as in the case of the vertebral artery and a slight opacification illustrating the low flow into the intracranial sinus in the posterior fossa. Nevertheless, the risk of rebleeding for the cerebral DAVFs has been estimated at approximately 35% for the DAVFs with retrograde cortical venous drainage within 2 weeks after the initial bleeding [21]. This risk could be applied for these DAVFs with retrograde rostral venous drainage into the posterior fossa and we recommend treatment at an early stage of these malformations in order to avoid the possible occurrence of major complications that could be caused by new bleeding.

The treatment of DAVFs at FM remains controversial between endovascular or microsurgical exclusion [4,5,12,14,22,23]. Moreover, the microsurgical interrupion of venous drainage has currently been fortunate because endovascular access is not easy for microcatheterization into these small tortuous arteries. Furthermore, the high risk of possible emboli related to the complex microanastomotic network, as well as the transvenous embolization is not suitable as this includes the high possibility of recanalization after incomplete occlusion. Moreover, the lateral suboccipital approach by a linear incision permitted a reliable exposure of the vertebral artery and the venous draining to achieve blockage. As in our case, the treatment of the venous aspect provided the treatment of both vascular anomalies under partial intraoperative control by indocyanine green video-angiography.

4. Conclusion

The coexistence of vascular anomalies at the FM should be carefully investigated by complete craniocervical angiography. The metameric syndrome of PAVF and DAVFs suggests a possible common genetic origin. The successful interruption of the venous side of the DAVF provided the complete treatment of both malformations.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

References