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Research Article

Subarachnoid Hemorrhage Revealing a Fistula Point Venous Aneurysm in a Dural Arteriovenous Fistula at the Craniocervical Junction: A Case Report

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Abstract

Dural arteriovenous fistulas represent approximately 10% of all intracranial vascular malformations, of which 1 to 2% are located at the craniocervical junction. Their location at the craniocervical junction is uncommon but potentially significant, given its proximity to and relationship with vital anatomical structures in this region. The association of a dural arteriovenous fistula with a venous aneurysm is rare. In this report, we present the case of a 49-year-old patient with no particular pathological history who was admitted for acute headaches. The neurological examination revealed a patient with grade I WFNS, intracranial hypertension syndrome. Radiological investigations including CT, CT angio and MRI revealed a fisher IV subarachnoid hemorrhage due to a ruptured venous aneurysm associated to an arteriovenous fistula at the craniocervical junction. He underwent clipping of the veinous aneurysm and microsurgical obliteration of the fistula. The postoperative course was uneventful. The clinical examination at 1-year post-op was unremarkable, and the follow-up MRI at 1-year post-op showed complete obliteration of the malformation. This report showed ruptured dural arteriovenous fistula should be considered in the etiology workup for posterior fossa subarachnoid hemorrhage. Effective and safe obliteration is possible without angiography.

Keywords

Subarachnoid Hemorrhage, Craniocervical Junction, Dural Arteriovenous Fistula, Venous Aneurysm, Microsurgical Obliteration

1. Introduction

Dural arteriovenous fistulas (DAVFs) are acquired intracranial vascular malformations consisting of pathological connections in the dura mater between the pial arteries and vascular veins, including the walls of the dural sinuses,

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bridging veins or transosseous outflow veins. [1] They are most often located in the transverse and cavernous sinuses and represent approximately 10% of all intracranial vascular malformations. [1, 2] DAVFs located at the craniocervical junction are extremely rare (1 to 2% of intracranial/spinal FAVDs). [3] They are distinguished from arteriovenous malformations by their arterial supply from vessels perfusing the dura and by the absence of parenchymal nidus. [1]

The association of a cerebral DAVFs with cerebral aneurysms is rare. David O Onu et al and Hashiguchi et al have shown that the development of a venous aneurysm over time can be part of the natural history of DAVFs and contributes significantly to bleeding complications. [4, 5]

The craniocervical junction (CCJ) is a complex anatomical structure composed of the brainstem and spinal cord, the mixed nerves, the first spinal nerves, the vertebral artery and its branches, and the ligaments uniting the atlas, axis and occipital bone. [6] DAVFs may be discovered incidentally, and if they are symptomatic, the clinical presentation ranges from mild neurological deficits to severe and fatal intracranial hemorrhage. [7] Angiography is the imaging technique of choice for diagnostic research and management planning. The optimal therapeutic strategy for DAVFs of the craniocervical junction remains controversial due to their deep location and the complexity of the feeding arteries and draining veins. [2, 6]. Endovascular treatment is often the first line of treatment for AVFs. Surgery and, to a lesser extent, stereotactic radiosurgery are used when endovascular approaches are impossible or unsuccessful. In low- and middle-income countries, access to endovascular treatment is not always available, making surgery the only option for cure.

Combined DAVFs and aneurysms are rare. To our knowledge, despite the several reports published to date on CCJ's DAVFs, a concomitant association with rupture of a venous aneurysm has not yet been reported. We describe in this report the case of a young patient received for SAH in whom the radiological investigation suspected (due to the unavailability of cerebral angiography in our context), a DAVF confirmed intraoperatively with the discovery of a venous aneurysm which benefited from clipping with microsurgical obliteration.

2. Case Presentations

A 49-year-old patient with no specific pathological history was admitted to the emergency department of the Fann University Hospital Centre for: Acute headache, intense and persistent for 72 hours regardless of oral analgesics. On clinical examination, the patient was awake, with headache and a GCS score of 15/15 with isochoric photoreactive pupils. On neurological examination, the patient was classified as grade I according to the WFNS scale, with intracranial hypertension.

2.1. Investigations

When the patient was admitted, a brain CT scan without injection of contrast medium revealed a Fisher IV subarachnoid haemorrhage (SAH) (Figure 1). A cerebral angio-CT scan was used to investigate the aetiology, and revealed a subtentorial vascular bundle in contact with the left vertebral artery, invading the left posterolateral part of the bulbous-medullary junction, raising suspicion of an arteriovenous malformation with no clearly identified drainage vein (Figure 1). In order to gain a better understanding of the architecture and vascular relationships of the malformation, we performed a brain MRI centred on the craniocervical junction, which confirmed the presence of vascular bundles, with an aneurysmal-type addition image, but without a clearly identifiable drainage vein (Figure 2). Given the radiological perplexity of these different images, and in the absence of arteriography in our working context, two hypotheses were put forward: arteriovenous malformation (AVM) or dural arteriovenous fistula (DAVF) of the CCJ.

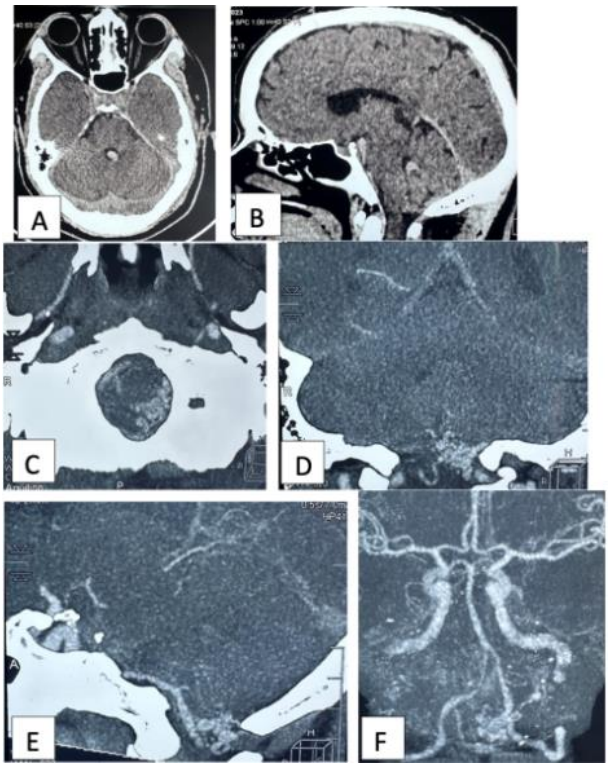

Figure 1. *Brain CT scan, axial (A), sagittal (B) showing SAH fisher.*

IV; CT angiography: axial (C), coronal (D), sagittal (E), MIP reconstruction in frontal section (F): showing a tangle of vessels at the left bulbo-medullary junction without clearly identified drainage vein

2.2. Treatment

The vascular lesions described above were considered more

suitable for microsurgical treatment. The indication for obliteration by left far lateral craniotomy was recommended and carried out after obtaining the patient's informed consent. Intraoperative exploration revealed a tangle of arterialized veins, which we dissected until we identified the fistula point and the venous aneurysm. Once the aneurysm has been occluded, we also occlude the draining vein of the AVF, followed by coagulation and partial removal of the vessels pack (figure 3).

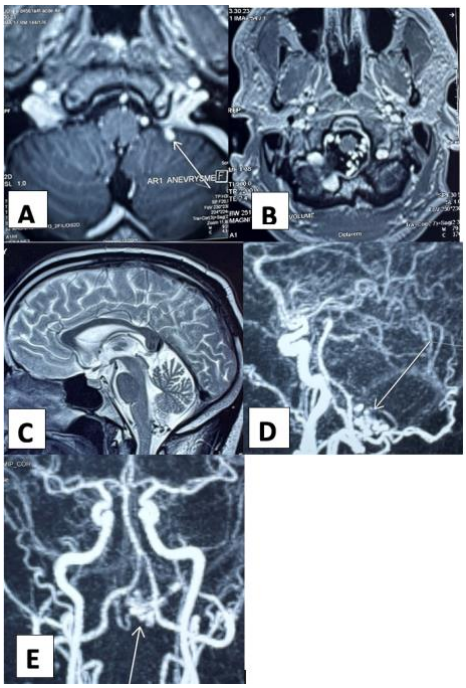

Figure 2. *Brain MRI, axial T2 flair (A), sagittal T2 (B), axial T2 diffusion (C), cerebral MRI with MIP reconstruction in sagittal (D), frontal (E) sections: suspected AVF of the craniocervical junction with a single draining vein oriented superiorly and laterally. Note presence of a venous aneurysm.*

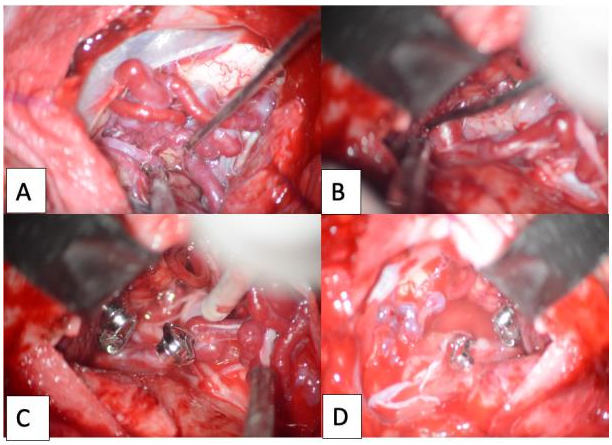

Figure 3. *Intraoperative views. Left far lateral approach showing a tangle of arterialized veins in the lateral aspect of the cisterna magna (A); Identification of fistula point and veinous aneurysm (B); Checking the flow with micro-doppler probe after clip obliteration and veinous aneurysm clipping (C); Veins turning dark at the end of the procedure (D).*

2.3. Outcome and Followof -up

The post-operative course was uneventful and anatomopathological analysis of the surgical part confirmed the existence of a dural arteriovenous fistula. On discharge home, he was well conscious, GCS 15/15, autonomous and had no clinical features. The patient's clinical examination at 1 year post-op was unremarkable. MRI and brain CT scan were satisfactory, with complete obliteration of the fistula. (figure 4).

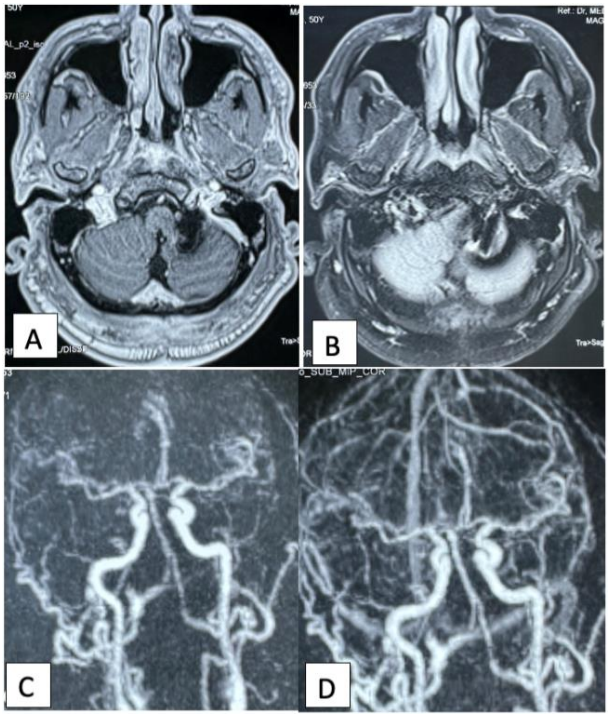

Figure 4. *Brain MRI, axial T2 flair (A), T2* (B): artefact around the vascular clip, Cerebral angiography with MIP reconstruction in frontal section (C): complete DAVFs, absence of any vascular lesion.*

3. Discussion

Intracranial DAVFs are acquired arteriovenous shunts confined to the dura mater, fed mainly by branches of the external carotid artery and generally drained either anterogradely via the dural venous sinuses, or retrograde into other dural or leptomeningeal venous channels [8]. They are often idiopathic but may be associated with venous thrombosis, head trauma, tumours, previous neurological surgery or meningitis [4, 8, 9].

This is the first case of idiopathic DAVFs recorded in our context in a young subject with no reported medical or surgical pathological history. In contrast to the polymorphic clinical presentation (SAH, myelopathy, brainstem dysfunction, radiculopathy, cranial nerve palsy) often reported in CCJ DAVFs [10-13], our patient presented only with signs of intracranial hypertension without any clinical expression in the

spinal cord or radiculo-medullary system. The initial cerebral CT scan revealed a fisher IV SAH. This symptomatology in our patient could be explained by the presence of blood in the subarachnoid space due to a ruptured venous aneurysm located in a pial space. Early diagnosis and management also prevented rebleeding, with all the direct and indirect consequences of this lesion. In their work [14, 15], Aviv RI et al and Kai Y et al explain that the presence of a venous aneurysm or drainage in a cerebral vein significantly increases the risk of SAH.

In this case, the DAVFs was probably fed by branches of the external carotid artery, without a clearly identified drainage vein, but with an associated venous aneurysm. Given the unavailability of cerebral arteriography in our context, the point of drainage and the presence of venous reflux were not clearly defined, but the angiographic sequences suggested drainage into the peri-medullary veins, corresponding to grade III of the Borden classification and grade V of the Cognard-Merland classification.

The most common feature of CCJ AVF is hemorrhagic complications, in particular subarachnoid hemorrhage. Whatever the type of hemorrhage, it results from the rupture of fragile parenchymal veins which have become arterialized as a result of cortical venous reflux and hypertension. This phenomenon is more frequent in the presence of a venous aneurysm and drainage of DAVFs into the deep venous sinuses. [5, 16].

The optimal therapeutic strategy for DAVFs of the craniocervical junction remains controversial due to their deep location and the complexity of the feeding arteries and draining veins. They may be treated by endovascular embolization or microsurgical obliteration. In the coexistence of an DAVFs and a flow-related aneurysm, some authors have strongly recommended simultaneous surgical treatment to avoid the catastrophic consequences of subsequent rupture of either vascular anomaly. [17] Microsurgery was performed in our case because endovascular techniques were not available at the time of surgery.

Diagnosis of DAVFs at the craniocervical junction remains a challenge due to the complex vascular anatomy with tiny and tortuous feeder vessels. The availability of arteriography in our centres will facilitate diagnosis and optimal management.

4. Conclusion

DAVFs of craniocervical junction are rare lesions that often reveal themselves as intracranial hemorrhage. Intracranial or peri-medullary venous drainage and the presence of a venous aneurysm are associated with an increased risk of SAH. Microsurgical obliteration of the feeding arteries or draining veins is an effective and reliable method of treating DAVFs at the craniocervical junction. Embolization is a possible alternative to surgery in the treatment of selective DAVFs.

What is known about this topic and what this case adds.

DAVF of the CCJ is a rare vascular pathology and its disclosure by an SAH secondary to a concomitant venous aneurysm rupture is still exceptional. The presence of a venous varix or upward intracranial venous drainage is significantly associated with the occurrence of SAH in patients with cervical DAVF. A critical analysis of the scientific data available on the disease and the observation of this clinical case suggest that peri-medullary and/or intracranial venous drainage in the case of a vascular anomaly in the CCJ leads to relatively rapid venous flow and increased haemodynamic stress. This can lead to the formation of a venous aneurysm (‘varix’), which, if ruptured, will inevitably result in SAH.

We believe that prior knowledge of this detail will enable clinicians to better plan the surgical procedure in order to avoid early intraoperative rebleeding, which would further complicate the surgery.

Abbreviations

AVM	Arteriovenous Malformation
AVFs	Arteriovenous Fistulas
CCJ	Craniocervical Junction
CP	Contrast Product
CT	Computed Tomography
DAVFs	Dural Arteriovenous Fistulas
GCS	Glasgow Coma Scale
MRI	Magnetic Resonance Imaging
MIP	Maximal Intensity Projection
SAH	Subarachnoid Hemorrhage
WFNS	World Federation of Neurosurgery

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Author’s Contributions

All authors contributed to the study conception and design. Data collection and analysis were performed by Mbaye THIOUB, the first draft of the manuscript was written by Thioub Mbaye and Hugues Ghislain ATAKLA, and all authors commented on previous versions of the manuscript. All authors read and approved the final manuscript.

Declarations

Ethics Approval

The study was approved by the institutional review board of National teaching hospital Fann of Dakar. The patient provided informed consent for the inclusion of his clinical data in this case report.

Consent to Participate

Informed consent was obtained from the patient presented in this case report.

Consent for Publication

Informed consent was obtained from the patient.

Data Availability

The authors confirm that the data supporting the fndings of this study are available within the article and the rest of the data is available on request from the authors.

Conflicts of Interest

The authors declare no conflicts of interest.

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