A Patient with a Dural Arteriovenous Fistula at the Craniocervical Junction in Whom Endovascular Treatment was Initially Performed, Followed by Surgical Management

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Objective: Dural arteriovenous fistulae at craniocervical junction (CCJ dAVF) cause subarachnoid hemorrhage (SAH). We present a case of CCJ dAVF with SAH in which endovascular treatment was initially performed, followed by surgical management, leading to radical cure.

Case Presentation: Transarterial embolization was initially performed because of unfavorable clinical condition of the patient, resulted in main feeder occlusion. As this made the shunting point clearer, surgical drainer occlusion consequently facilitated, and radical cure could be achieved.

Conclusion: To treat CCJ dAVF, endovascular treatment and surgical management were performed. The former alone did not result in radical cure, but decreased the shunt volume, facilitating the assessment of vascular anatomy.

Keywords ▶ craniocervical junction, dural arteriovenous fistula, subarachnoid hemorrhage, endovascular treatment, craniotomy

Introduction

Dural arteriovenous fistulae at the craniocervical junction (CCJ dAVF) are rare, but are important as an etiologic factor for subarachnoid hemorrhage (SAH). The source of hemorrhage cannot be identified using CT angiography in many cases, and detailed angiography is necessary. This disease must be differentiated from other shunt diseases at the same site, such as perimedullary AVF and arteriovenous malformation (AVM) because treatment methods are different. In this study, we report a patient with CCJ dAVF in whom endovascular treatment was initially performed, followed by surgical management. When selecting a treatment method, it is the most important to understand vascular anatomy.

Case Presentation

A 73-year-old male patient had sudden onset of consciousness disturbance at home, and he was taken to a previous hospital by ambulance. The Glasgow Coma Scale (GCS) score was 13 (E3V4M6) on arrival. A brain CT revealed Fisher group 3 SAH (Fig. 1). Subsequently, angiography was performed, leading to a diagnosis of CCJ shunt disease. Treatments in acute stage were considered, but the patient developed pneumonia and acute renal failure, and was referred to our hospital on Day 14. In our hospital,
intensive care for medical complications was conducted, and angiography was performed on Day 23, when they had improved. A diagnosis of CCJ dAVF based on its vascular anatomy was made. It had a feeder of meningeal branch of radicular artery branching from left vertebral artery (VA) immediately after penetrating the dura, a shunting point of ventral side of medulla oblongata, and a drainer of pontomesencephalic vein ascending the anterior surface of bulbopontine with abnormal dilatation and torsion (Fig. 2). 3D DSA findings revealed a continued vascular dilatation was observed immediately before shunting. Angiography confirmed the single feeder, and no involvement of the anterior spinal artery as feeding artery. The detailed vascular anatomy is shown in Fig. 3. His consciousness had improved to be alert, but the activities of daily living (ADL) were poor because of disuse syndrome. Although surgical management was required, we should withhold until his condition would turn to tolerate it. Transarterial embolization was scheduled on Day 30, targeting radical endovascular treatment based on the patient’s vascular anatomy. Under general anesthesia, a 6 Fr Roadmaster guiding catheter MPDA 90 cm (Goodman, Nagoya, Aichi, Japan) was inserted to the V2 segment of the left VA. A Marathon microcatheter (Covidien, Minneapolis, MN,
left lateral suboccipital craniotomy was performed on Day 148. The draining vein was identified on the dura adjacent to the hypoglossal nerve, and a tortuous blood vessel ascending the anterior surface of the medulla oblongata connected to the draining vein was confirmed. The draining vein was occluded with a clip at a site adjacent to the dura (Fig. 5), and the cessation of the blood flow of the tortuous blood vessel was confirmed using indocyanine green (ICG) angiography. Postoperative angiography also confirmed the disappearance of the shunt (Fig. 6).

Subsequently, the patient was referred to another hospital for rehabilitation on Day 209. During the 1-year follow-up after onset, dysphagia persisted, but his consciousness remained clear, and he was independent.

**Discussion**

CCJ dAVF are rare. According to a national survey in Japan, the annual incidence of dAVF is 0.29 per 100,000 persons, and CCJ dAVF accounts for 2.4% of all patients with dAVF (annual incidence: 0.7 per 10,000,000 persons). Zhao et al. reviewed 56 patients with CCJ dAVF, and reported that dAVF was more frequent in females, whereas CCJ dAVF was more frequent in males (male-to-female ratio: 3:1). Myelopathy initially occurred in many patients with dAVF of the thoracic spinal cord or lower, whereas SAH initially developed in many patients with...
Fig. 4  (a) Oblique view on left vertebral artery angiography after embolization and (b) MIP image of 3D DSA. As a main feeder was embolized, the dural branch (arrow head) of a new feeder became manifest, making the shunting point (arrow) clear. MIP: maximum intensity projection

Fig. 5  (a) Reconstruction of 3D angiography findings in which embolized intra-dural part of the main feeder cut as with surgery (b) Schematic drawing. arrow: Loop of main feeder embolized at the site of dural penetration; arrow heads: thin feeders that became manifest after main-feeder embolization; asterisk: Shunt point; solid double ended arrow: Site of clip occlusion; point arrow: A portion of the intradural main feeder cut during surgery. (c and d) Intraoperative images. asterisk: A drainer adjacent to the shunting point was occluded with a clip. arrow: Loop of a feeder embolized; VA: vertebral artery

CCJ dAVF (37.5%). Another study indicated that SAH related to CCJ dAVF was milder than that related to aneurysmal rupture (Hunt & Hess grade I/II, 83.3%). According to other studies, the direction of an outflow affects symptoms; hemorrhage initially occurred in many patients with ascending outflow tracts, and myelopathy in those with descending outflow tracts.2-4) The spontaneous course remains unclear due to an extremely small number of patients, and no study has reported the incidence or timing of additional hemorrhage. Treatment should be indicated for the patients with progressive myelopathy. When hemorrhage initially occurs, there may be no necessity of
emergency treatment, differing from aneurysmal rupture. However, a study reported an additional hemorrhage occurred 4 months after the initial onset; radical cure must be targeted. For diagnosis, CTA or MRA is primarily used. Angiography is essential for evaluating vascular anatomy. It is sometimes difficult to evaluate a detailed structure using conventional DSA; selective angiography or 3D DSA including MIP images are useful. In particular, the spinal artery is involved in CCJ perimedullary AVF to be differentiated, and radical cure is not achieved by drainer occlusion; thus, shunting point occlusion is required. Patients with both perimedullary and dural AVF have been reported. A shunting point on images is diagnosed based on rapid changes in the vascular diameter, but such a change of the vascular diameter is difficult to identify in some cases. In the present case, the shunting point could be more accurately evaluated by 3D DSA after embolization. For the detailed assessment of shunting point, there is an option of angiography under microballoon occlusion of the main feeder. Concerning treatment, surgical management is usually selected for radical cure. According to reviews of this disease, the craniotomy-related radical cure rate is approximately 100%, whereas the endovascular-treatment-related radical cure rate is approximately 70%. As a feeder is thin and tortuous, it is difficult to advance a microcatheter to the area adjacent to the shunting point and embolic materials cannot penetrate the shunting point. In the present case, our attempt resulted in main feeder occlusion. Furthermore, anastomoses with blood vessels perfusing the brainstem may induce unexpected complications related to aberrant embolic materials. No study has initially adopted the combination of endovascular treatment and surgical management, differing from dAVF in other sites. According to several studies, surgery was additionally performed when a shunt remained despite radical endovascular treatment, as demonstrated in the present case. For our patient, endovascular treatment was selected for the following reasons: surgical management might be impossible due to the unfavorable general condition; early treatment to reduce shunting blood flow was necessary after the onset of hemorrhage; and the possibility of embolic materials aberration into a spinal blood vessel was ruled out based on vascular anatomy. A study indicated that embolization of VA itself led to radical cure in a patient with an unfavorable general condition. However, the manifestation of a latent feeder, which is unclear before embolization, may occur in patients with dAVF, and radical cure may not be achieved. Recently, there has been reported that cases in which Onyx was used as an embolic material achieved radical cure. In the future, endovascular treatment may be considered as a first-line treatment of CCJ dAVF.

**Conclusion**

A patient with CCJ dAVF who initially developed SAH was performed endovascular treatment, followed by surgical management. The former alone did not lead to radical cure, but it decreased the shunting volume without any complications. This facilitated the more accurate
assessment of vascular anatomy, contributing to radical cure by surgical management.

Disclosure Statement

There is no conflict of interest for the main author and coauthors.

References