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Case Report

Estimation of the rupture point of the craniovertebral junction intradural arteriovenous fistula with vessel wall magnetic resonance image and its pathological findings: A case report

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ABSTRACT

Background: Arteriovenous fistulas (AVFs) of the craniocervical junction (CCJ) and intradural AVFs are often associated with aneurysms and varics, and it is sometimes difficult to identify the ruptured point on radiological images. We report a case in which vessel wall magnetic resonance image (VW-MRI) was useful for identifying the ruptured point at the CCJ AVF.

Case Description: A 70-year-old man presented with a sudden onset of headache. He had Glasgow Coma Scale E4V5M6, world federation of neurosurgical societies (WFNS) Grade I. Fisher group 3 subarachnoid hemorrhage and hydrocephalus were found on head computed tomography. Cerebral angiography showed a spinal AVF at the C1 level of the cervical spine. Magnetic resonance image-enhanced motion sensitized driven equilibrium (MSDE-method showed an enhancing effect in part of the AVF draining vein, but the vascular architecture of this lesion was indeterminate. We performed continuous ventricular drainage for acute hydrocephalus and antihypertensive treatment. Cerebral angiography was performed 30days after the onset of the disease, and was revealed an aneurysmal structure in a portion of the AVF draining vein, which VW-MRI initially enhanced. On the 38th day after onset, he underwent direct surgery to occlude the AV fistula and dissect the aneurysmal structure. Histopathology showed that the aneurysmal structure was varices with lymphocytic infiltration, and hemosiderin deposition was observed near the varices.

Conclusion: Recently, VW-MRI has been reported to show an association between the enhancement of varices in dural AVF and rupture cases. VW-MRI, especially the enhanced MSDE method, may be useful in estimating the ruptured point in arteriovenous shunt disease.

Keywords: Arteriovenous fistula of craniocervical junction, Ruptured point, Subarachnoid hemorrhage, Varices, Vessel wall magnetic resonance image, CCJ dAVF, Pathological findings

INTRODUCTION

Arteriovenous fistula (AVF) of the craniocervical junction (CCJ) is a rare spinal vascular lesion classified as dural, intradural, or extradural according to vascular anatomy. Among these, intradural AVFs are reported to be more frequently associated with subarachnoid hemorrhage

(SAH) (43% vs 82% vs 43%) and aneurysm-like dilation or varices in the lesion (14% vs. 74% vs. 14%).[8] However, when aneurysms and varices are not evident on radiological imaging examinations, the estimation of the bleeding point may be difficult. Recently, the usefulness of vessel wall magnetic resonance image (VW-MRI) for identifying the ruptured point has been reported. We have reported the usefulness of VW-MRI in identifying ruptured aneurysms in multiple aneurysms.[13] In this study, we report a case of intradural AVF in which VW-MRI was useful for estimating the ruptured point in the CCJ-AVF, including a review of the literature.

CASE PRESENTATION

A 70-year-old male was brought to our emergency department with a complaint of sudden onset headache. He had Glasgow Coma Scale E4V5M6, WFNS grade I, at the time of arrival. A head computed tomography (CT) showed Fisher group 3 SAH and acute hydrocephalus [Figures 1a and b]. CT angiography did not reveal an arterial aneurysm, but abnormal vessels were observed around the right vertebral artery of the intracranial portion [Figure 1c], suspecting AVF. Cerebral angiography was performed on the same day. An arteriovenous (AV) shunt was observed, with the C1 radicular artery and anterior spinal artery of the right vertebral artery as the inflow vessels and the intradural vein as the drain aging vein. The outflow vein drained cranial and caudal intradural veins. No arterial aneurysm or varices was evident in the lesion [Figure 2a] at the initial cerebral angiography. Magnetic resonance image (MRI) contrast-enhanced motion-sensitized driven equilibrium (MSDE) showed a faintly enhanced area in front of the spinal cord that was presumed to be a vascular component [Figures 2b-e]. MRI contrast spoiled gradient recalled echo (SPGR) did not show any obvious vascular architecture in the same area [Figure 2f].

We diagnosed SAH due to ruptured intradural AVF at the CCJ. Feeding arteries were the C1 radicular artery, anterior spinal artery, and draining veins outflow to cranial and caudal directions. In the acute period, continuous ventricular drainage was performed. Considering the possibility that the hematoma might obscure the shunt point and that other lesions might be revealed as a source of bleeding, we decided on a wait-and-see surgical and to review radiological examinations.

Cerebral angiography was performed again on the 26th day. A varicose structure was seen in part of the draining vein compared with initial angiography [Figures 3a and b]. The same site coincided with a vascular component that appeared to be an outflow vein previously by MRI-enhanced MSDE [Figures 3c and d]. MRI contrast-enhanced SPGR also showed an aneurysmal structure [Figure 3e]. We considered that this aneurysmal structure had ruptured, and we planned direct surgery to occlude AVF.

On the 38th day, he underwent a right suboccipital craniotomy and hemilaminectomy of the right C1. After the dural incision, a shunt point was found at the cranial side of the first cervical nerve, which was considered to be the radicular AVF [Figures 4a and b]. The inflow vessel, including the shunt point, was coagulated. Moreover, we observed the ventral side of the spinal cord, and the varices were found, so we coagulated and removed its lesion [Figures 4c and d]. Intraoperative indocyanine green angiography confirmed the disappearance of AV shunt flow. The postoperative course was good, and cerebral angiography was performed on postoperative day 9, confirming the disappearance of AV shunt flow. On the 14th day, he underwent a ventriculoperitoneal shunt for secondary hydrocephalus and was transferred to a rehabilitation hospital with a modified Rankin scale 2 on the 30th day.

Pathological findings

The lesion contained a muscular artery with an internal elastic lamina, a vein with a sparse muscular layer, and a

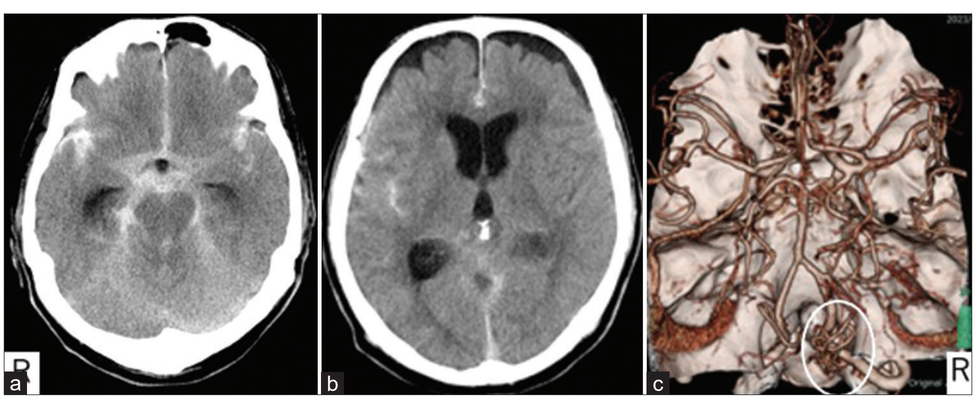

Figure 1: (a and b) Computed tomography (CT) showed subarachnoid hemorrhage and hydrocephalus. (c) CT angiography showed abnormal vessels around the right intracranial vertebral artery (within a circle).

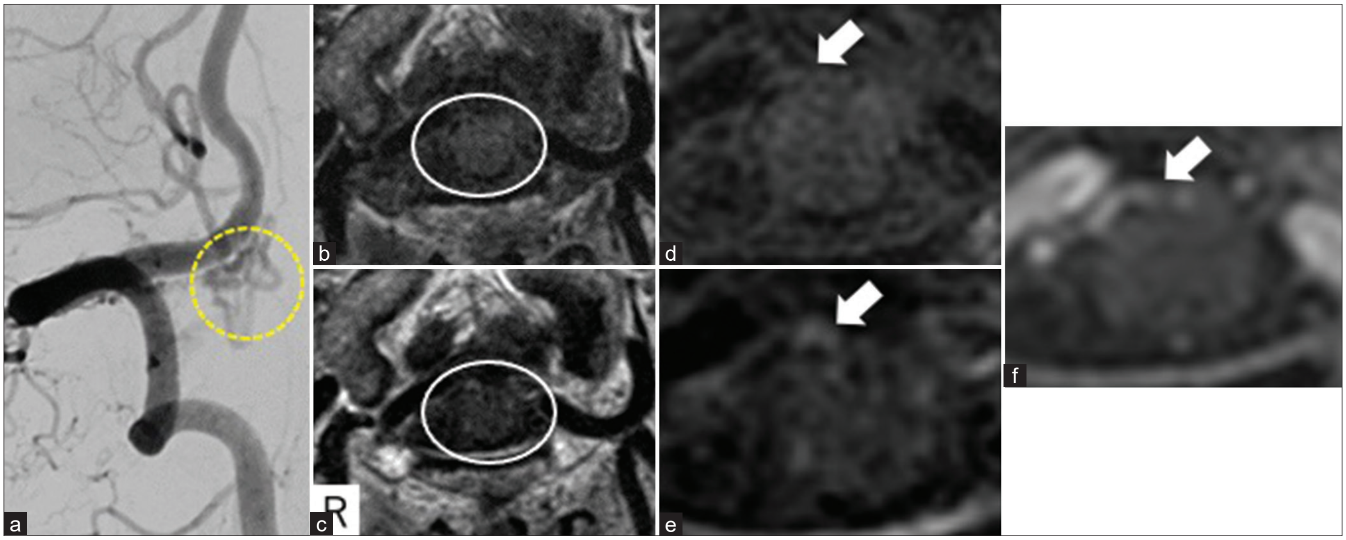

Figure 2: Initial radiological images. (a) Right vertebral angiography showing an arteriovenous fistula at the craniocervical junction. (A-P arterial phase, yellow dotted circle). (b-e) Motion-sensitized driven equilibrium shows contrast effect ventral to the spinal cord (white arrow); b and d are plain, c and e are contrast-enhanced, and d and e are magnified images of b and c (within a circle). (f) Contrast spoiled gradient recalled echo at the same level of b and c did not clearly show vascular structure (white arrow).

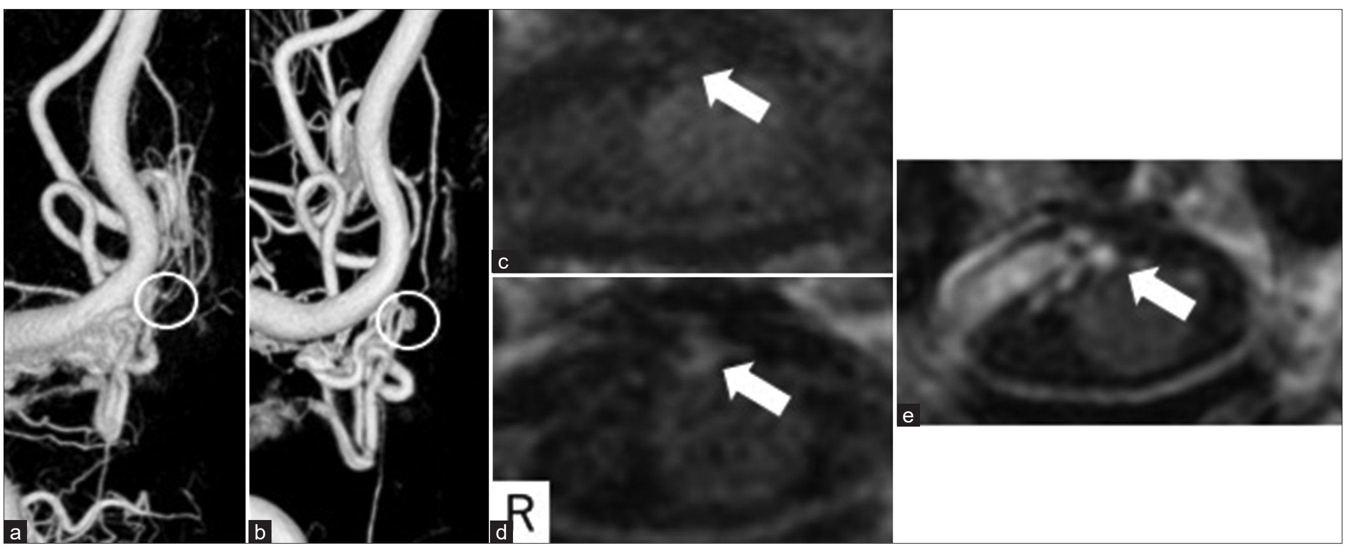

Figure 3: Changes in radiological images. (a) Day of onset; (b–e) 26 days after onset. (a and b) Three-dimensional digital subtraction angiography showed an aneurysmal structure in some of the draining veins (white circle). (c-e) Contrast spoiled gradient recalled echo also showed an aneurysmal structure (e), and the same level of motion-sensitized driven equilibrium showed an enhancing effect (c and d) white arrow (c-e).

vascular component with a thin wall that appeared to be a varices [Figure 5a]. Both the muscular artery and vein had enlarged areas of the lumen that were presumed to be the transition to the dilated vessels and were thought to be an AV shunt point. Elastica Masson staining showed that the walls of suggested varices structures were highly thinned and lacked internal elastic lamina [Figure 5b]. The vein wall also showed irregular thickening of the muscular layer, suggesting arterialization that causes abnormal pressure load [Figure 5c]. Immunohistochemical staining results for Figure 5d is presented in Figure 6. Although there was no obvious ruptured point in the specimen, there were lymphocytic cells infiltrated around the varicose vein [Figures 5d and 6a], suggesting inflammation and hemosiderin deposition [Figure 6b], which may have occurred after bleeding. CD68 staining showed positive cells around blood vessels [Figure 6c], which appeared to be macrophages. Myeloperoxidase staining was negative [Figure 6d].

DISCUSSION

Recently, AVFs of the CCJ have been increasingly reported, but its diagnosis is difficult due to its complex vascular structure. Hiramatsu *et al*. reported the vascular anatomy and clinical characteristics of each and classified AVF into five categories based on the location of the feeding artery and AV shunt in the vascular anatomy.[7] In the present case, intraoperative findings revealed the presence of a shunt point on the nerve root of C-1 in the dura mater, which was considered the radicular AVF. Clinically, radicular AVF is often associated with hemorrhagic events (94%), and our case also had an SAH. The risks of hemorrhage include ascending drainage, varicose veins, aneurysms, radicular AVF, intramedullary descending drainage, dural branches of vertebral arteries, and epidural drainage.[7,10] Our case had ascending drainage, varices, and intramedullary ascending drainage, and variceal structures were confirmed by preoperative images, and the intraoperative diagnosis was radicular AVF, suggesting that the lesion was at high risk for bleeding. Treatment includes direct surgery only in 35% of cases and combined treatment with direct surgery and endovascular treatment in 29%, with more than 60% of cases including direct surgery.[7]

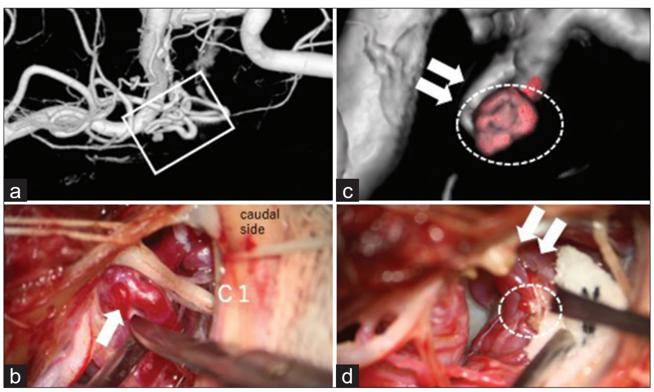

Figure 4: Intraoperative view. (a) The inside of the frame is an operation view (Three-dimensional digital subtraction angiography, white square). (b) A shunt point was noted on the cranial side of the first cervical nerve (arrow). (c and d) Double arrows indicate a draining vein. A circle indicates an aneurysmal structure.

The presence of aneurysms and varices is associated with the development of bleeding, and some reports suggest that they are treated at the same time as the shunt point occlusion.[4,5,12] On the other hand, aneurysm and varices formation may be hemodynamic and flow-dependent. Since the varices in our case were located ventral to the spinal cord, we decided to determine whether the varices could be safely removed after the AVF was occluded.

After the AVF occlusion, the varices were removed after determining that the procedure could be performed safely. In our case, the lesion that was inferred to be a vascular component on initial VW-MRI could be determined to be varices on later examination, and this lesion was judged as the bleeding point intraoperatively.

It has been reported that VW-MRI cannot suppress the blood flow signal in low-velocity blood flow, such as veins and shows high signal intensity.[11] Therefore, findings of the venous component of VW-MRI have not been studied.

However, Cord *et al*. reported on VW-MRI of an intracranial dural AVF (dAVF) with hemorrhagic onset with varices. In all three cases, the vein wall near the hematoma component showed a selective enhancement and was presumed to be the ruptured point.[3] However, all of these reports were treated with endovascular therapy; the pathology had not been examined.

In our case, the reason why the draining vein of intracranial dAVF showed as an enhanced vascular component was considered to be due to arterialization of the draining vein

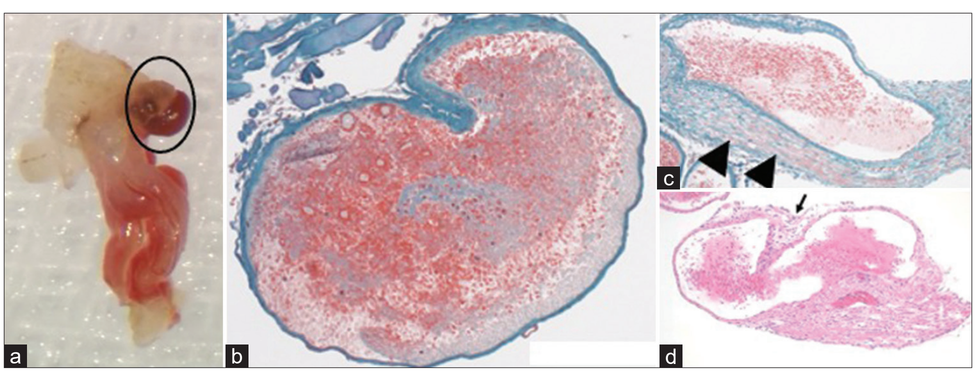

Figure 5: Histological findings. (a) Arteriovenous fistula and aneurysmal structure (circle). (b) The aneurysmal structure was considered to be a varix because there was no internal elastic plate in the vessel wall. The vessel wall was highly thinned. (Elastica-Masson; ×40). (c) The draining vein around the varix was arterialized(arrowhead). (Elastica-Masson; ×10). (d) Lymphocytic infiltration and edematous changes in the wall of the draining vein surrounding the varix were observed (arrow). (Hematoxylin-Eosin; ×10).

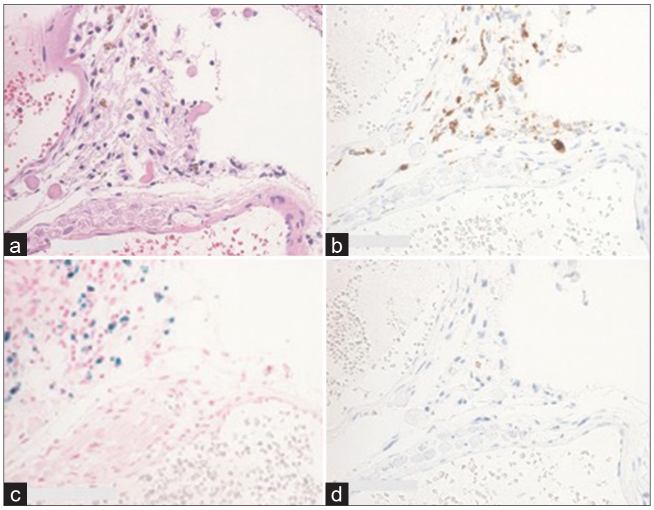

Figure 6: Histological finding with immunohistochemical staining. (a) Draining vein around varix (Hematoxylin-Eosin; ×40). (b) Hemosiderin deposition surrounding draining veins (Berlin blue; ×40). (c) CD68-positive cells are seen in the area of hemosiderin deposition (CD68; ×40). (d) MPO staining is negative (Myeloperoxidase; ×40).

near the shunt. Moreover, we got the pathological finding of arterialization of a continuous venous component from the varices, which supported the findings by Cord *et al.*[3]

VW-MRI is used to identify ruptured cerebral aneurysms and estimate the vulnerability of aneurysm, and inflammation of the aneurysm wall is believed to be the cause of the enhancement. Myeloperoxidase staining, which is secreted by neutrophil granulocytes, was positive in 4 out of 5 unruptured cerebral aneurysms that showed vessel wall enhancement by VW-MRI.[9]

Other inflammatory cells, such as macrophages, have also been implicated. Inflammation is reported to be involved in hemorrhage of cerebral AV malformations as well as cerebral aneurysms,[2] and Hasan *et al*. have attempted to delineate macrophages using ferumoxytol-enhanced MRI in cerebral AV malformations. In two of four cases, the low signal was observed in the nidus. In one of these cases, CD-68-positive macrophages were identified within the vessel wall of the nidus.[6] Bhogal *et al*. reported that VW-MRI of a hemorrhage-onset cerebral AV malformation showed enhancement in the vessel wall of the nidus, but no findings that suggested inflammation were obtained on immunostaining.[1]

In our case, a vascular lesion that had shown an enhancement on VW-MRI was found later to have a distinct vascular structure as a varices, which was presumed to be the ruptured point.

Although myeroperoxidase (MPO) staining was negative in our case, lymphocytic infiltration and CD 68 positive cells were observed near the rupture point, and the involvement of inflammation was also inferred as a factor in the positive VW-MRI finding.

Moreover, the arterialization of the venous component, which was identified in the pathology, may have contributed to the enhancement, suggesting that VW-MRI may be useful in estimating the rupture point in AV shunt disease as well.

CONCLUSION

We have experienced a case of AVF of the CCJ with SAH in which VW-MRI could identify the rupture point.

VW-MRI has the potential to identify the ruptured point not only in cerebral aneurysms but also in AV shunt disease.

Ethical approval

The Institutional Review Board approval is not required.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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