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Case Report

A rare case of spinal dural arteriovenous fistula mimicking malignant glioma of the medulla oblongata: Significance of cerebral angiography for accurate diagnosis of brain stem region

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ABSTRACT

Background: The findings of a hyperintense sign on T2-weighted imaging (T2-WI) and gadolinium (Gd) contrast enhancement on magnetic resonance imaging (MRI) of the brain stem suggest malignant glioma. However, this pathological condition is probably uncommon, and it may be unknown that a dural arteriovenous fistula (DAVF) can imitate this radiological pattern. In addition, it is extremely rare to be caused by a spinal DAVF. Here, a rare case of spinal DAVF that mimicked malignant glioma of the medulla oblongata is presented.

Case Description: A 56-year-old woman was admitted with a progressive gait disturbance, vertigo, and dysphasia. MRI showed a hyperintense signal in the medulla oblongata on fluid-attenuated inversion recovery (FLAIR) and moderate contrast enhancement on Gd-enhanced MRI. Interestingly, Gd-enhanced MRI demonstrated abnormal dilated veins around the brain stem and cervical spinal cord. Cerebral angiography showed spinal DAVF at the left C4/C5 vertebral foramen fed by the C5 radicular artery. The fistula drained into spinal perimedullary veins and flowed out retrograde at the cortical vein of the posterior cranial fossa. Therefore, surgical disconnection of the spinal DAVF was performed by a posterior approach. The patient's postoperative course was uneventful. Cerebral angiography showed complete disappearance of the DAVF, with marked reductions of the hyperintense sign of the medulla oblongata on FLAIR.

Conclusion: This important case illustrates MRI findings mimicking brain stem glioma. In cases with the hyperintense sign-on T2-WI associated with contrast enhancement suspicious of brainstem glioma, careful checking for perimedullary abnormal vessels and additional cerebral angiography should be performed.

Keywords: Cerebral angiography, Lower cranial nerve palsy, Mimicking medulla oblongata glioma, Spinal dural arteriovenous fistula, Venous congestion

INTRODUCTION

Accurate diagnosis of the brain stem or cervical spinal cord lesions is very difficult on routine magnetic resonance imaging (MRI).[2,4,6] The reason why is that, in this region, various pathological conditions present, for example, tumors (gliomas, lymphoma, and so on), inflammatory diseases, degenerative disorders, vascular diseases, and processes secondary to systemic conditions, and

there are no specific findings on MRI to differentiate them.[2,4,6] In addition, faster and more accurate diagnosis of pathology is required than for other parts of the body due to the severity of symptoms, and an inadequate diagnosis can lead to an immediately life-threatening condition.[2,6] Here, a rare case of spinal dural arteriovenous fistula (DAVF) presenting as a contrast-enhanced lesion with T2 hyperintensity mimicking malignant glioma of the medulla oblongata is presented.

CASE DESCRIPTION

A 56-year-old woman with a 2-month history of nausea and progressive gait disturbance visited our department after experiencing gradual worsening of swallowing difficulty over a 2-week period. On admission, she complained of severe nausea and truncal ataxia, and she had left glossopharyngeal and vagal nerve palsies. MRI of the brain showed hypointensity on diffusion-weighted imaging (DWI) and hyperintensity on fluid-attenuated inversion recovery (FLAIR) of the medulla oblongata and cerebellar peduncle. Gadolinium (Gd) administration showed moderate enhancement mainly within the medulla oblongata [Figure 1]. At first glance, the images were considered compatible with an infiltrative malignant glioma, and the issue was the early introduction of chemoradiotherapy treatment. However, a more careful analysis using MRI emphasized that there were abnormal dilated veins clearly demonstrated around the brain stem and in front of the cervical spinal cord [Figure 2a and b], and a hyperintense signal on T2-weighted imaging (T2-WI) in the medulla, with an abnormal signal flow void extending to the cervical portion of the spinal cord [Figure 2c]. This characteristic radiological finding suggested the possibility of a vascular malformation due to a DAVF, and cerebral angiography was thus performed. Cerebral angiography and three-dimensional contrast-enhanced computed tomography images showed a left-sided spinal DAVF at the location of the C4/C5 vertebral foramen with arterial supply through the C5 radicular artery arising from the left vertebral artery. The draining vein was single, and it drained into the cortical vein localized retrograde along the left aspect of the medulla oblongata through spinal perimedullary veins [Figure 3]. Thus, the involvement of the brain stem and cerebellar peduncle was due to venous congestive injury. Therefore, the surgical disconnection of the spinal DAVF by a posterior approach was performed [Figure 4]. The patient's postoperative course was uneventful, and the lower cranial nerve palsy and nausea were markedly improved over a few days after the treatment, and 3 weeks later, she returned to her normal lifestyle with slight truncal ataxia. Cerebral angiography 2 weeks after surgery showed complete disappearance of the spinal DAVF

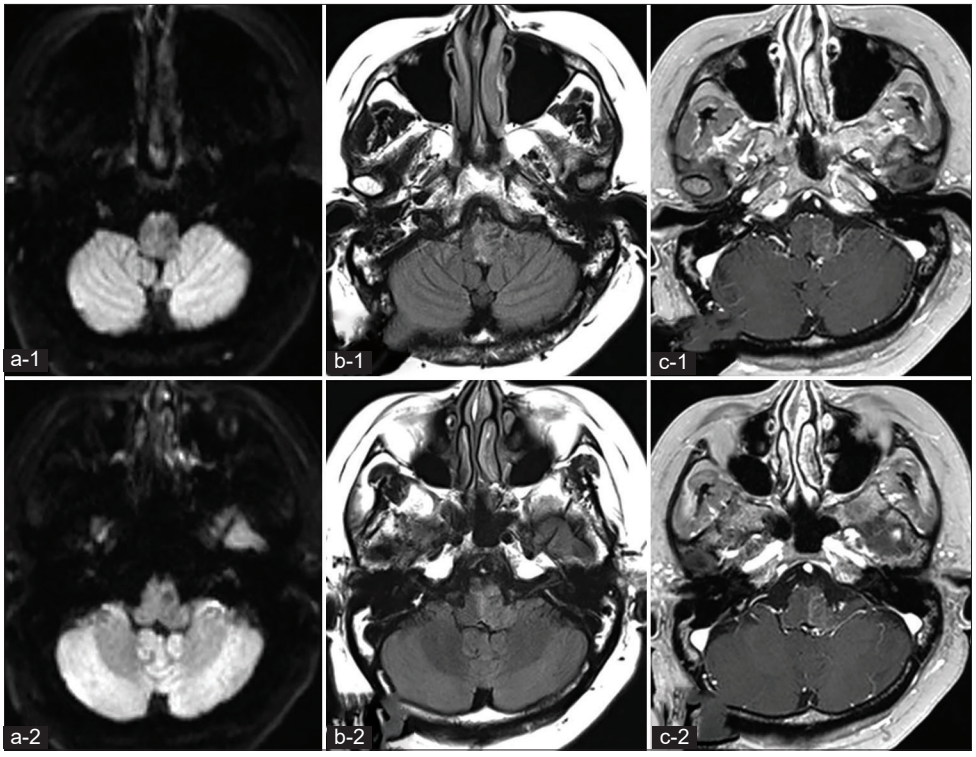

Figure 1: Preoperative axial magnetic resonance imaging. (a) Diffusion-weighted image (DWI), (b) fluid attenuated inversion recovery (FLAIR), and (c) gadolinium (Gd)-enhanced T1-weighted images showing a lesion in the medulla oblongata. The lesion is hypointense on DWI hyperintense on FLAIR and shows moderate enhancement on Gd administration.

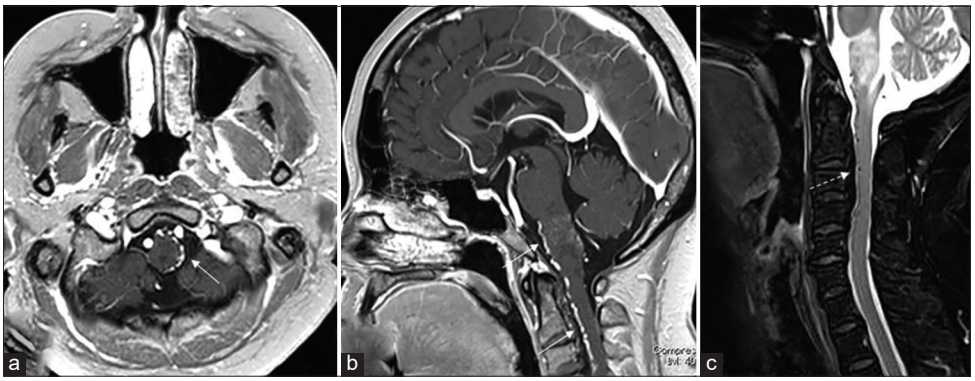

Figure 2: Gadolinium (Gd)-enhanced T1-weighted MRI (a: axial and b: sagittal) clearly showing abnormal dilated veins around the brain stem and in front of the cervical spinal cord (white arrow). T2-weighted sagittal image showing hyperintensity of the medulla and an abnormal signal flow void extending to the cervical portion of the spinal cord (white dashed arrow) (c).

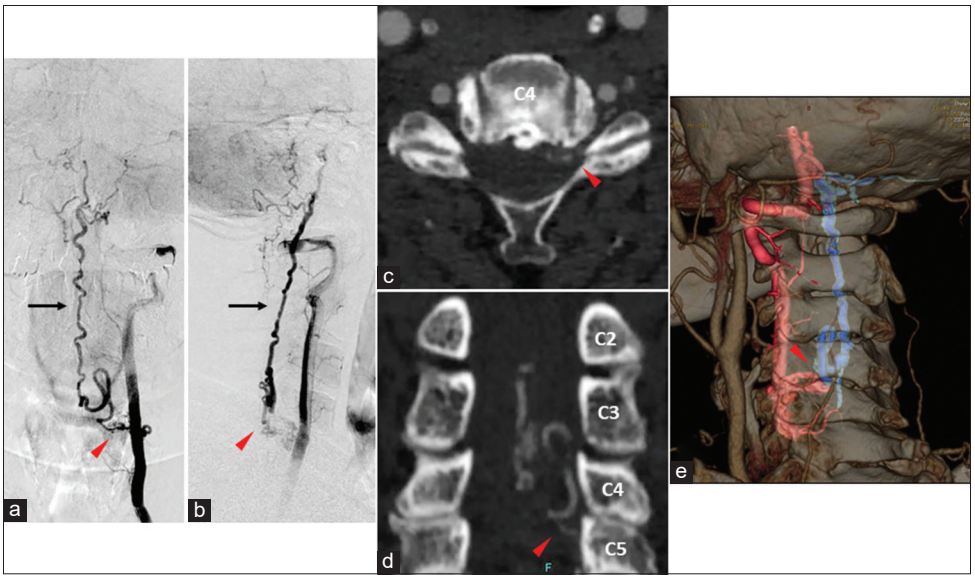

Figure 3: Left vertebral artery angiographic findings. (a) Anteroposterior view and (b) lateral view showing a spinal dural arteriovenous fistula (red arrowhead) supplied by the C5 radicular artery arising from the left vertebral artery draining into the spinal perimedullary veins (black bald arrow) reaching the cortical vein localized along the left aspect of the medulla oblongata. Computed tomography three-dimensional fusion digital subtraction angiography demonstrating the exact location of the arterio-venous shunt point (red arrowhead). (c) Axial view, (d) coronal view, and (e) reconstruction fusion model (red blood vessels are arteries and blue are draining veins)

[Figure 5a and b], resulting in marked reductions of the hyperintense sign of the medulla oblongata on FLAIR and disappearance of the atypical perimedullary vein on Gd-enhanced T1-WI on MRI [Figure 5c and d].

The clinical study of the above-mentioned case report was approved by the Ethics Committee for Clinical Research of Ehime University Hospital, and informed consent was obtained from the patient before initiating the study.

DISCUSSION

The precise diagnosis of the brain stem and cervical cord lesions is often difficult in the field of neurosurgery.[2,4,6] In particular, vascular disease in this area, for example, DAVF, is even more difficult to diagnose accurately, and appropriate therapeutic interventions often tend to be delayed.[1,2,8,10] Thus, we should take utmost care when treating pathological lesions in this part.

It is commonly accepted that a hyperintense sign-on T2-WI or FLAIR and T1 contrast enhancement of Gd on MRI of the brain stem and cervical cord is mainly related to malignant gliomas.[2,6,8] However, for a pathological condition in this area, in particular the medulla oblongata, it is challenging to perform a surgical biopsy for an accurate diagnosis. Thus, this pathological condition could be misdiagnosed as a malignant glioma based on MRI findings alone, and chemoradiotherapy

could easily be administered incorrectly. On the other hand, it is so far uncommon and probably unknown that the edematous lesion caused by venous congestion with an intracranial DAVF can produce a similar radiological pattern to that of malignant glioma, especially Gd contrast enhancement and spinal cord enlargement.[1,2,6,8,10] In the case of a DAVF in the posterior cranial fossa, the venous drainage is through a unique cisternal arterialized vein, leading to venous congestion. This congestion occurs due to the increasing venous pressure, responsible for slowing drainage of the venous routes of the brain, and spinal cord.[2,6,8] In addition, perimedullary venous drainage of DAVFs presents a high risk of ischemic or hemorrhagic lesions, unlike DAVFs with sinus drainage.[2,6,7,8] Therefore, we think that a DAVF in this area requires immediate and appropriate therapeutic action.

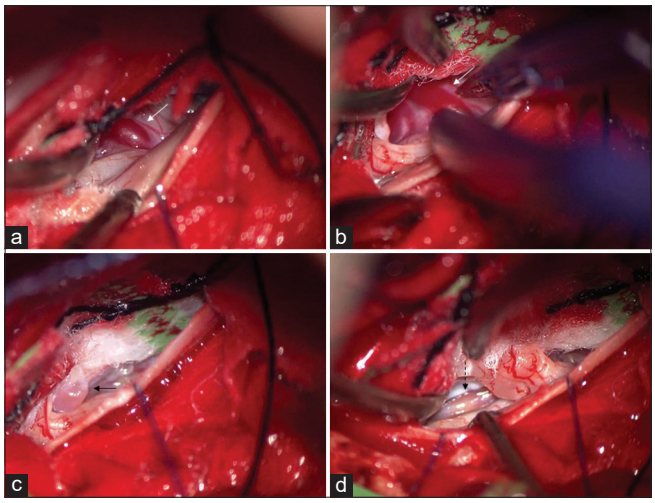

Figure 4: Intraoperative views (opening of the dura mater, left side), showing the shunt location and the single draining vein (white arrow) (a and b), which is disconnected with bipolar coagulation (black arrow: disconnected vein) (c). The draining vein is degenerated with shunt disconnection (black dashed arrow) (d).

Making the diagnosis of spinal or intracranial DAVF can be very difficult because the usual MRI protocol can miss an abnormal dilated vessel, and because its appearance can mimic that of a neoplasm at its initial presentation.[6,7] In fact, the presence of a hyperintense brainstem signal on T2-WI or FLAIR suggests the possibility of brainstem glioma.[2,6,8,9] However, in this setting, it is necessary to perform a careful workup searching for evidence of vascular, inflammatory, or infectious processes.[2,6,9] The identification of atypical perimedullary vessels on MRI is the most important hallmark of a DAVF in the posterior cranial fossa and spinal cord.[2] Experienced neurovascular radiologists or neurosurgeons performing an accurate review of the images of the present case showing the dilated venous pattern would likely favor a diagnosis of DAVF.[2] In their recent study, Haryu *et al*. found perimedullary flow-related signal voids in only 37% of patients with Cognard Type V

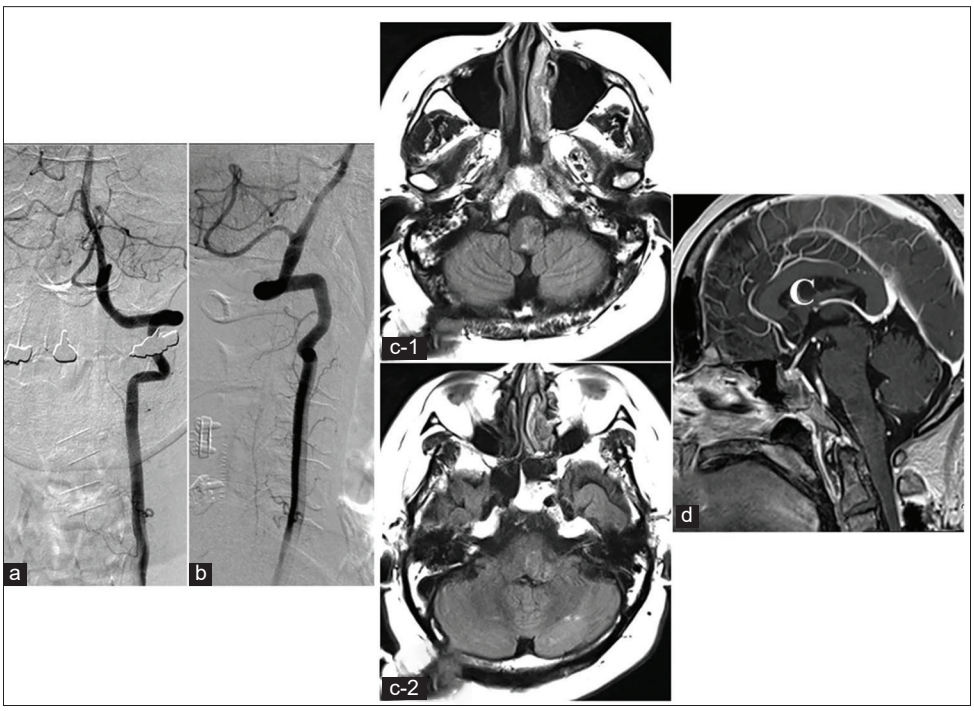

Figure 5: Left vertebral artery angiographic findings. (a) Anteroposterior view and (b) lateral view demonstrating complete disappearance of the dural arteriovenous fistula. Postoperative MRI shows marked reductions of the hyperintense sign of the medulla oblongata on fluid-attenuated inversion recovery (c) and the disappearance of the atypical perimedullary vein on gadolinium-enhanced T1-WI (d).

DAVFs.[3,5] Thus, simply looking for the flow void signal on T2-WI is insufficient for making the diagnosis of spinal DAVF. However, these abnormal perimedullary vessels with the dilated venous pattern are apparently detected on Gd-enhanced T1-WI of MRI.[2] Le Guennec *et al*. reported that when both T2-weighted MRI and T1-weighted MRI (in particular contrast-enhanced T1-WI) are carefully examined, the detection rate of atypical perimedullary vessels with MRI reaches 76%.[6] In the present case, the first impression of the pathological condition was a malignant tumor. However, it was possible to diagnose DAVF by noticing the abnormal blood vessels on MRI, in particular, the sagittal view of Gd-enhanced T1-WI [Figure 2b].

In the previous reports, intracranial DAVF in the posterior cranial fossa has often been reported as the cause of signal changes on T2-WI and of Gd enhancement in this part.[2,6,8] However, the present case showed an ischemic lesion in the brainstem caused by the spinal DAVF. This pathological condition that is the location of the arterio-venous fistula could not be confirmed by MRI alone. In other words, cerebral angiography is essential for accurate diagnosis of shunt location and planning the therapeutic strategy. Therefore, in patients who have brain stem symptoms and who show an ischemic lesion over a wide brain stem area on MRI suggestive of DAVF, cerebral angiography should be performed, taking spinal DAVFs into consideration. To the best of our knowledge, there has been no report of spinal DAVF patients who initially presented with brain stem ischemia. Further experience with this pathology and results from longer patient follow-up is required.

CONCLUSION

A rare case of spinal DAVF identified by recognizing venous congestion of the medulla oblongata mimicking a malignant glial tumor was presented. This is an extremely important case report that describes MRI findings mimicking brain stem glioma. In cases with the hyperintense sign-on T2-WI associated with contrast enhancement suspicious of brainstem glioma, a careful check for perimedullary abnormal vessel dilatation and additional cerebral angiography should be mandatory.

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.

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