

A case of vertebrobasilar dolichoectasia with acute brainstem compression and obstructive hydrocephalus entrapped between dissecting aneurysm and cerebellar infarction

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Abstract

Vertebrobasilar dolichoectasia is relatively rare, and it can compress the brainstem and surrounding brain cistern due to its volume. It is frequently associated with arterial dissection, obstructive hydrocephalus, and infarction in the brainstem or cerebellum. We present a case of a 57-year-old man with a dissection localized to the basilar artery. However, the patient developed an infarction to the right posterior inferior cerebellar artery region, acute brainstem compression, and obstructive hydrocephalus caused by entrapment between the dissecting aneurysm and cerebellar infarction. Posterior fossa decompression and extraventricular drainage were performed immediately and alleviated his condition. We suggest that such treatments may be needed at early stages, and we also review existing cases in the literature.

Keywords: Dolichoectasia, vertebrobasilar artery dissection, cerebellar infarction, obstructive hydrocephalus, posterior fossa decompression craniotomy

INTRODUCTION

Vertebrobasilar dolichoectasia (VBD) is relatively rare, and it can compress the brainstem and surrounding brain cistern due to its volume; it is frequently associated with arterial dissection, obstructive hydrocephalus, and infarction in the brainstem or cerebellum. In VBD, because the dilated and meandering blood vessels tend to form thrombi, the penetrating branches tend to twist, and collateral circulation tends to develop¹⁻³. This may cause infarction in various parts of the posterior circulatory system and may present with acute brainstem compression or obstructive hydrocephalus entrapped between the dissecting aneurysm and infarction. In this case, although the dissection was localized to the basilar artery (BA), an infarction developed in the right posterior inferior cerebellar artery (PICA) region. It is predicted that the number of cases of dolichoectatic vertebrobasilar dissecting aneurysm should be potentially large, but there are few cases reported in the literature.

Herein, we present this rare case, and to investigate its clinical features, pathogenesis, and treatment effects. We performed an English

literature search and review of the PubMed database with the keywords *dissection* or *dissecting* and *dolichoectasia* or *dolichoectatic*.

CASE REPORT

A 57-year-old man presented with headache, vomiting, and right facial pain and was admitted to our hospital. Computed tomography showed a low-density area in the right cerebellar hemisphere and a 17-mm high-density area that appeared aneurysmal (Figure 1A).

Magnetic resonance imaging (MRI) with diffusion-weighted imaging (DWI) showed a fresh infarction in the right cerebellum and right lateral medulla in the right vertebral artery and posterior inferior cerebellar artery (PICA) region (Figure 1B). Magnetic resonance angiography (MRA) (Figure 1C), basi-parallel anatomical scanning (BPAS) MRI (Figure 1D), and cerebral angiography (Figure 1E, F) revealed basilar artery (BA) dilation and meandering. The right anterior inferior cerebellar artery (AICA) was occluded by a thrombotic dissecting aneurysm, and the right PICA was hypoplastic, all of which suggested that the right PICA was originally hypoplastic,

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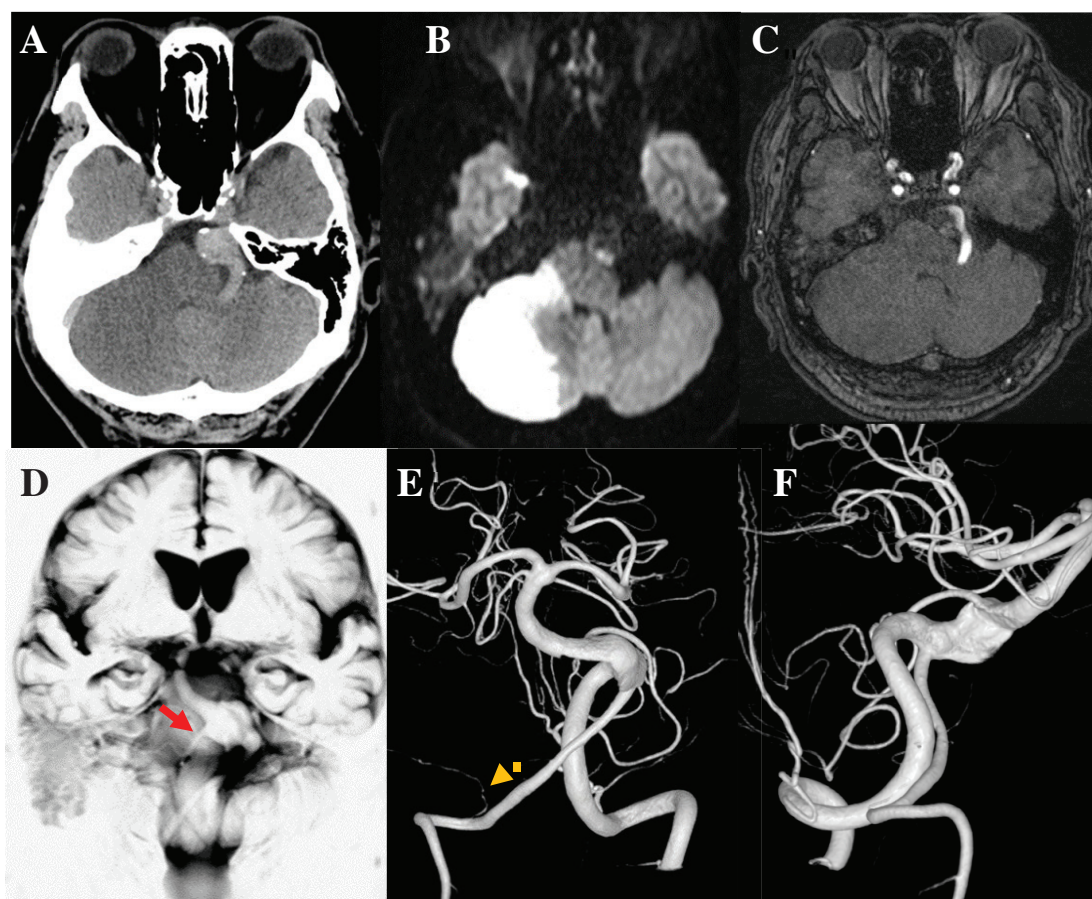


Figure 1 A-F, images acquired at the time of admission. Computed tomography (CT) showing a low-density area in the right cerebellar hemisphere and a 17-mm high-density area appearing aneurysmal (A). Magnetic resonance imaging (MRI) with diffusion-weighted imaging (DWI) showing fresh infarction in the right cerebellum and right lateral medulla (B). TOF (time of flight) MRA (C) showing signs of a double lumen in the basilar artery (BA). Basi-parallel anatomical scanning (BPAS) MRI (D) and magnetic resonance angiography (MRA) (E) showing BA dilatation and meandering, all of which suggested anterior inferior cerebellar artery (AICA); (red arrow) and posterior inferior cerebellar artery (PICA)(yellow arrow) occlusion due to thrombosed dissecting aneurysm associated with vertebrobasilar dolichoectasia (VBD).

and the right PICA region was mainly supplied from the right AICA, or the hypoplastic right PICA was twisted by transformation of VBD.

Because of clear consciousness (Glasgow Coma Scale score [GCS] of 15) on admission, we initially treated the patient conservatively with antiplatelet agents. However, 18 hours after onset, his consciousness level and respiratory condition rapidly deteriorated (GCS of 6). Since the MRI showed the brainstem and fourth ventricle were entrapped between the thrombosed aneurysm and a worsening edema caused by right cerebellar infarction (Figure 2A-C), urgent posterior fossa decompression and extraventricular drainage were performed.

After the operation, his consciousness level had improved to GCS of 14, and he was transferred

to a convalescent hospital for rehabilitation due to residual dysphagia. However, one month after transfer, he developed subarachnoid hemorrhage due to dolichoectatic vertebrobasilar dissecting aneurysm and died. Informed consent was obtained from the patient for publication of this case report and accompanying images.

DISCUSSION

Dolichoectasia is characterized by the dilatation, elongation, and meandering of an artery. One of the diagnostic criteria for VBD is an arterial dilatation to ≥ 4.5 mm in diameter, a meander ≥ 10 mm, and a basilar artery length ≥ 29.5 mm.⁴ As the present case met this criterion, a diagnosis of VBD was confirmed. Because the

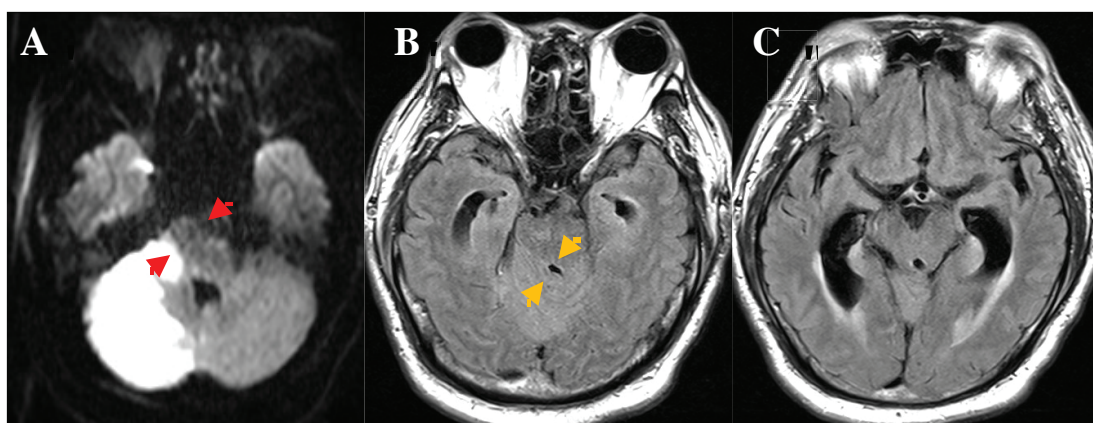


Figure 2. Diffusion-weighted imaging (DWI) showing brainstem compression between the thrombosed aneurysm and worsening edema of the cerebellar infarction (A) (red arrow). Fluid attenuated inversion recovery (FLAIR) images (B). (C) showing obstructive hydrocephalus because the fourth ventricle was entrapped between the thrombosed aneurysm and a worsening edema caused by right cerebellar infarction (yellow arrow).

vertebral arteries are usually asymmetrical, the hemodynamic stress from the larger vertebral artery will eventually cause meandering of the basilar artery, and these dilated and meandering vessels can compress adjacent structures such as the cranial nerves, brainstem, and the ventricle. Cranial nerve dysfunction causes the most common symptoms, such as dizziness, tinnitus, and auditory impairment, and unilateral facial convulsions, facial paralysis, and trigeminal neuralgia due to trigeminal nerve compression are also common.¹ Because of vessel wall thinning at the bends, cracking of the internal elastic lamina, degeneration of the tunica media, and loss of the internal elastic lamina due to wear and tear caused by turbulence in the distal stenosis, VBD is frequently associated with dissecting aneurysms.²

To investigate clinical features, pathogenesis, and treatment effects in patients who had dissecting VBD aneurysms, we performed an English literature search and review using the PubMed database with the keywords *dissection* or *dissecting* and *dolichoectasia* or *dolichoectatic*. Publications without detailed demographic descriptions, such as age or sex, or with no mention of dolichoectasia but reporting only dilated or tortuous vertebrobasilar arteries, were excluded. As Table 1 shows, only 12 cases, including the present case, have been reported.^{1, 2, 5-9}

The mean patient age was 58.8 (standard deviation, 7.1) years, all patients were male, and the BA was reported as the site of dissection. Of the 6 cases mentioned for complications of infarction, 2 cases including ours had cerebellar infarction even though the arterial dissection was localized to the BA.

In VBD, blood flow is reduced in the dilated blood vessels, and stasis of blood flow makes it easier for blood clots to form in the blood vessels. Abnormal dilation and tortuosity of the basilar artery causes extension and twist of the perforator. Gradual occlusion of the perforated artery by VBD may develop an anastomotic network that mobilizes collateral circulation in unaffected vascular segments of the basilar artery such as AICA / PICA or SCA, therefore, causing infarction in various parts of the posterior circulatory system.³

Ischemia has been frequently reported in dissecting vertebrobasilar arteries. Severe cerebellar infarction results in brainstem compression or obstructive hydrocephalus, and posterior fossa decompression or extraventricular drainage is effective in treating this condition. According to Jüttler *et al.*, median time from symptom onset to surgery in space-occupying cerebellar infarction is 4 days for extraventricular drainage alone and 3 days for posterior fossa decompression and extraventricular drainage groups.¹⁰ This might suggest that cerebellar infarction with dolichoectatic vertebrobasilar dissecting aneurysm may cause more acute brainstem compression or obstructive hydrocephalus.

In conclusion, VBD is easily associated with dissection and infarction in various parts of the posterior circulatory system. Because the volume of the aneurysm itself compresses the brainstem and ventricles, they are easily entrapped between the aneurysm and worsening cerebellar edema, leading to early brainstem compression or obstructive hydrocephalus.

Table 1: Summary of previously reported DVDA cases

	Author	Year	Age (years)/sex	Site of dissection	Type of stroke	Outcome	Recurrence
1	Mizutani ⁵	1992	51/M	BA	Infarction: L. cerebellum	Tetraparesis and lower cranial disturbance with progressive worsening	Yes
2	Mizutani ⁵	1992	53/M	BA	n.d.	NND	Yes
3	Mizutani ⁵	1992	73/M	BA	n.d.	n.d.	Yes
4	Mizutani ⁵	1992	56/M	BA	n.d.	n.d.	No
5	Mizutani ⁵	1992	52/M	R. VA, BA	n.d.	Ataxic gait and slurred speech with some recovery	No
6	Vieco ⁶	1997	60/M	BA	n.d.	Death	No
7	Nabika ⁷	2002	58/M	BA	n.d.	n.d.	Yes
8	Isa ²	2013	63/M	L. VA, BA	Infarction: pontine, thalamus, cerebellum, occipital lobe	Death	No
9	Yang ⁸	2017	51/M	BA	Infarction: pontine	R. hemiparesis, improved	Yes
10	Zhang ⁹	2018	60/M	L. VA, BA	Infarction: multifocal, SAH	Death	No
11	Wang ¹	2019	72/M	BA	Infarction: basal ganglia, L. thalamus, R. periventricular area	NND	No
12	Present case	2020	57/M	BA	Infarction: R. cerebellum and R. medulla	Moderately severe disability	Yes

M: male, BA: basilar artery, L.: left, n.d.: not described, NND: no neurological deficits, R.: right, VA: vertebral artery, DVDA: dolichoectatic vertebrobasilar dissecting aneurysm

Higher sample sizes would be necessary to establish more concrete recommendations with generalizability. Despite this limitation, this report presents important data that may benefit clinicians treating patients with cerebellar infarction associated with VBD or dolichoectatic vertebrobasilar dissecting aneurysm.

DISCLOSURE

Conflict of interest: None

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