Spontaneous Occlusion of a Large Fusiform Basilar Artery Dissecting Aneurysm in a Child Complicated with Brainstem Infarction: a case report

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ABSTRACT

Basilar artery dissecting aneurysms, although very rare in children, may be associated with severe morbidity and mortality. The management of these vascular lesions depends on clinical presentation of patients. Large fusiform vertebro-basilar dissecting aneurysms are exceedingly rare and can exert mass effect on the brainstem and cerebellum. We herein report the neuroimaging findings, including MRI, MR angiography, diffusion-weighted imaging, and conventional angiography, in a four-year-old boy with a large fusiform basilar artery dissecting aneurysm complicated with brainstem infarction. The implications of neuroimaging findings on the decision to treat this patient are also discussed.

Basilar artery dissecting aneurysms (BDA) are very rare in the pediatric population [1]. However, they may be associated with severe morbidity and mortality because of the possibility of generating strokes. In patients with BDA, conditions requiring aggressive intervention mainly comprise arterial dissection and aneurysm formation. Thrombus in a patent pseudolumen of the dissection may cause ischemic insults to distal arterial territories, while a aneurysm poses a risk of hemorrhage [2-4]. Giant vertebro-basilar dissecting aneurysms are exceedingly rare and can exert mass effect on the brainstem and cerebellum [5]. We herein report the neuroimaging findings, including MRI, MRA, DWI, and conventional angiography, in a four-year-old boy with a large fusiform basilar artery dissecting aneurysm complicated with acute brainstem infarction.

CASE REPORT

A 4-year-old boy had been well until several hours before admission when he suddenly fell down from a bicycle. Right hemiplegia and right facial palsy soon developed. There were no congenital or other acquired risk factors for cerebrovascular disease. Neurological examination revealed decreased muscle power of the right extremities to grade 1/5, with increased deep tendon reflexes and positive Babinski sign. Laboratory data, including hemogram and biochemistry, were unremarkable. MRI showed loss of vascular flow void of the distal basilar artery with an intramural hematoma composed of mixed signal intensities and peripheral curvilinear hyperintensity on both T1-weighted and T2-weighted images (Fig. 1). A T2-hyperintense lesion was seen in the left pontine region,
with reduced water diffusion demonstrated by diffusion-weighted imaging. MRA showed complete occlusion of the distal basilar artery. These imaging findings were consistent with a basilar artery dissecting aneurysm complicated with acute brainstem infarction. Transfemoral angiography of the vertebro-basilar system confirmed complete occlusion of the basilar artery occlusion. Angiography of the bilateral internal carotid arteries showed patent bilateral posterior cerebral arteries (PCAs) but no opacification of the distal basilar artery or superior cerebellar arteries. Without evidence of opacification of the aneurysm, further intervention was not performed. He was conservatively treated with anti-coagulation, and transferred to rehabilitation. Muscle power of his right extremities gradually improved. Follow-up MRI ten months later showed shrinkage of the aneurysm (Fig. 2) and persistent occlusion of the distal basilar artery. The brainstem was less compressed. At clinical follow-up three years later, he had a nearly complete recovery with mild numbess of the right extremities.

**DISCUSSION**

Vertebro-basilar dissecting aneurysms (VDA) in children and adolescents has a worse prognosis than the extracranial dissection [6-8]. Trauma represents the most...
common predisposing factor for VDA during childhood. Other predisposing factors include fibromuscular dysplasia, inheritance of intracranial aneurysms, connective tissue disorders such as Marfan’s syndrome, Moyamoya disease, systemic lupus erythematosus, syphilitic arteritis, polyarteritis nodosa, degeneration of the media, and oral contraceptives [6-8].

On MRI, VDA is seen as a signal void representing a patent lumen, surrounded by a crescent-shaped intramural hematoma of variable signal intensities depending on its stage [9]. The chronological changes of the intramural hematoma parallel those of an intracerebral hematoma, showing isointensity on T1WI in the acute stage, staying hyperintense from the subacute stage to approximately 2 months, and becoming isointense or unrecognizable within 6 months [10]. Intraluminal hematoma could be differentiated from an atheroma, also an non-enhancing intraluminal lesion within an aneurysmal artery, such that atheroma exhibits a relatively linear outer surface, a lack of chronological signal change on MRI, and an absence of features of an atherosclerotic plaque including a fibrous cap overlying a necrotic core, calcified regions, luminal irregularity, and ulceration [11]. Moreover, an atheroma is unlikely to occur in a child. Contrast enhancement of the dissecting aneurysm may represent a vulnerable lesion (Fig. 1c); on follow-up imaging, it may indicate persistent aneurysmal dilatation [12]. Conventional angiographic findings of dissecting aneurysms include luminal stenosis or occlusion, aneurysmal dilatation preceding or following a focal vascular narrowing, true and false lumens, isolated fusiform dilatation, and intimal flap [13]. Diffusion-weighted imaging has been known to be able to differentiate between cytotoxic and vasogenic edema [14]. In our case, the brainstem was severely compressed by the large aneurysm. While the T2WI showed subtle hyperintensity in the pons, DWI provided a greater imaging contrast of this brainstem abnormality and further confirmed this abnormality to be acute infarction. The dissection, thrombus and complicated acute brainstem infarction in this boy were believed to arise from a pre-existing fusiform aneurysm, suggested by smooth indentation of the adjacent pons, insignificant narrowing of the prepontine cistern, patency and hypertrophy of the bilateral PCAs and posterior communicating arteries (PComAs), and atrophy of the bilateral vertebral arteries.

Because the risk of recurrent bleeding in a VDA is high, aggressive surgery or endovascular treatment should be performed in most patients when possible [15, 16]. When treating a dissecting aneurysm, to increase the likelihood of a favorable outcome the operator should consider the location of the aneurysm, dominance of the vertebral arteries, and adequacy of collateral circulation. Surgical techniques include clipping, trapping, and wrapping [17]. Endovascular treatment, by using detachable silicon balloons or Guglielmi detachable coils, can be performed to embolize or trap the dissecting aneurysm [18], or occlude the parent artery [15] in carefully selected cases, for example, in a patient having a proximal basilar artery dissecting aneurysm with adequate collateral circulation from the bilateral PCAs and PComAs. Also, angioplasty can be performed with stents to preserve the parent artery [19]. However, spontaneous healing of the dissecting basilar artery has been reported [2].

In the present case, MRI showed the size and location of the dissection of the basilar artery suggested by its mural hematoma, and the thrombosed aneurysm. No obvious intracranial hemorrhage associated with rupture of the BDA was identified. Diffusion-weighted imaging showed acute infarction of the pons, suggesting that the ischemic insult may be an on-going process which required aggressive management to prevent or stabilize further insult. Moreover, the presence of vascular flow within the dissected arterial segment or the aneurysm could not be excluded by using MRI. Conventional angiography of the vertebrobasilar arteries was subsequently performed and showed occlusion of the basilar artery and no opacification of the dissected segment or the aneurysm. Also, carotid angiography showed no retrograde opacification of the BDA. The risk of rupture of the aneurysm tended to be low because it was almost completely thrombosed and no vascular flow was identified within both the dissected artery and aneurysmal sac. We decided not to

**Figure 2.** Follow-up axial T2-weighted MR image performed 10 months later shows shrinkage of the thrombosed aneurysm (arrows) and an old pontine infarct (double arrows).
perform aggressive intervention. Follow-up MRI showed persistent occlusion of the BDA. The decision of conservative management of this BDA appeared to be justified.

In conclusion, we demonstrated the neuroimaging findings in a 4-year-old boy having a large fusiform BDA with spontaneous thrombosis and complicated with acute brainstem infarction. Prompt neuro-intervention is mandatory in patients with a VDA because of high risk of recurrent bleeding. Location of the aneurysm, presence of thrombosis or associated infarction, and adequacy of collateral circulation are important aspects to consider when managing a VDA.

REFERENCES