The dilemma of treating vertebrobasilar dolichoectasia

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Abstract

Vertebrobasilar dolichoectasia (VBD) is a common phenomenon among people over 50 years old, and the related clinical expressions are varied. One of our VBD patients presented with brainstem infarction initially, received low molecular weight heparin treatment, and developed rupture of the dolichoectasia segment. Another patient with a similar-sized VBD experienced recurrent brainstem infarction three times over 2 years, despite higher bleeding tendency and long-term antplatelet treatment. The third patient with a smaller-sized VBD, had left hemiplegia and received intravenous recombinant tissue plasminogen activator within 3 h, totally recovered with no lesions detected on brain Magnetic Resonance Imaging (MRI). The pathophysiology of VBD is unique, its prevalence and risks of ischemic stroke and intracranial hemorrhage both increase as the degree of arterial dolichoectasia extends, making the strategy of management quite a challenge. The best management of VBD is controlling arterial hypertension and following up with image studies regularly to detect the early extension of VBD degree.

Case Reports

Case #1

A 58-year-old man was admitted to our emergency room (ER) because of dizziness and right arm weakness. His family reported that he had intermittent swallowing difficulty and easily choked over the past year. The patient had hypertension without prior medication control, and his blood pressure (BP) measured at our ER was above 200/100 mmHg. The next day, brainstem signs including dysarthria, nystagmus, dysphagia, right hemiplegia and hypesthesia, and bilateral positive Babinski signs developed. The brain computed tomography (CT) showed vertebrobasilar dolichoectasia with hyperdense basilar signs, indicating basilar thrombosis (Figure 1). CT angiography revealed dolichoectasia of the left vertebral artery and basilar artery with intraluminal thrombus (Figure 2). We gave the patient aspirin (324 mg) for one day and then started low-molecular-weight-heparin (LMWH, Clexane, 60 mg q12h) injection under the diagnosis of basilar artery occlusion. The patient’s muscle power with regards to his right limbs improved within the subsequent 2 days of starting treatment, but high BP (>200/100 mmHg) still persisted despite the use of multiple anti-hypertensive agents treatment. Four days after onset, the patient experienced a sudden severe headache with vomiting and further elevated BP (>210/120 mmHg), followed by absence of brainstem reflexes. Emergent brain CT demonstrated massive brainstem hemorrhage, the location indicated rupture of the dolichoectasia segment (Figure 3). The patient expired 9 days after onset.

Case #2

A 72-year-old man was incidentally found to have basilar artery dolichoectasia when he was admitted for a nasal hemangioma operation in 2005. He had hypertension (HTN), liver cirrhosis, esophageal varicose (EV) and gastrointestinal (GI) bleeding history, and had not been taking anti-hypertensive agents or antithrombotic agents previously. In April 2006, he experienced acute onset of dizziness, left limb numbness and weakness. The initial systolic BP when he came to ER was 180 mmHg. Left central facial palsy, dysarthria, and dysphagia appeared on day 3. Brain CT showed VDE without hemorrhage or prominent hypodense lesions (Figure 4), and he was prescribed clopidogrel during hospitalization. All the symptoms improved completely on day 8, and he stopped taking medications by himself and was lost to follow-up after discharge.

Then, he came again to our hospital in May 2007, with symptoms of right side weakness, slurred speech, and dysphagia. His systolic BP at ER was 155 mmHg, and was lower than 150 mmHg during hospitalization. Brain CT showed suspicious hypodense lesion over left pons, and VDE that was the same size as the previous study (Figure 5). We gave the patient 300 mg of aspirin daily. The patient’s symptoms recovered to baseline conditions within one week, and he continued the aspirin treatment and the use of calcium channel blockers for BP control. Five months later, another episode of left hemiplegia and dysphagia developed, accompanied with left central facial palsy with his tongue deviating to the left side. Systolic BP was initially 175 mmHg, and not above 150 mmHg afterwards. Brain imaging study demonstrated acute right pons infarction (Figure 6), with the same-sized VDE indentation to left pons (Figure 7). Clopidogrel was then added to his medication regimen, and his left limb muscle power improved partially. The patient was prescribed clopidogrel at the outpatient department follow-up. In February 2008, acute right pons infarction occurred again, with the symptoms of left limbs weakness, slurred speech, and diminished right
facial sensation. His systolic BP did not go above 150 mmHg through the course, and brain magnetic resonance imaging (MRI) confirmed the ischemic stroke, without differences in VDE characteristics as compared to previous study (Figure 8). 

**Case #3**

A 59-year-old woman had hypertension that was under control with the regular use of medications. She experienced sudden onset of left hemiplegia, left facial palsy, and tongue deviating to the left side. She was sent to our ER within 1 h, and BP measured in the ER was 170/91 mmHg. Brain CT demonstrated hyperdensity within basilar artery at the segment of medullo-cervical junction level, suggesting thrombus formation (Figure 9). The patient’s initial National Institute of Health stroke scale was 9, and we started intravenous recombinant tissue plasminogen activator (rt-PA) 2 h after symptoms onset, and monitored the patient’s condition closely, controlling her BP to be under 185/105 mmHg. The patient’s symptoms improved within 37 h after stroke onset. Followed-up brain MRI showed dilated and tortuous vertebrobasilar artery with compression on the left medullo-cervical junction, but no hyperdense lesion on diffusion weighted-MRI (Figure 10). The patient’s prognosis was favorable, and she returned home without any sequelae.

**Discussion**

The pathophysiology of intracranial dolichoectasia has been extensively studied, and the related vascular risk factors, the association with other stroke types, the relation to systemic arterial disease, and the genetic factors in the young have also been thoroughly researched. Cerebral infarction of dolichoectasia is caused by luminal thrombi that obstruct arterial branches, and the thin dilated arterial wall may also break and this may lead to intracranial hemorrhage. This is different from the pathophysiology of atherosclerosis and aneurysm. Many studies which compare
intracranial dolichoectasia to atherosclerosis and aneurysm also demonstrate different prevalence and prognosis regarding stroke. The first patient had an initial presentation of VDE with fluctuated brainstem symptoms for one year, which may have been caused by brainstem compression of vertebrobasilar occlusion. This could be the early warning sign of VDE enlargement or thrombus formation. The rapidly progressed vertebrobasilar territory infarction by basilar thrombus responded well to low-molecular-weight heparin initially, but soon developed into catastrophic VDE wall rupture. The patient’s HTN was never treated before admission, and the baseline BP was unknown; during hospitalization, BP was difficult to control and was always above 200/100 mmHg. The second patient was quite different. He had repeated neurological deficits within 3 years, was proven to have brainstem infarction during his most recent two events. We could not confirm the exact etiology of the first two episodes - ischemic stroke or brainstem compression - since they improved dramatically within one week. The diameter of VDE was about 8.7 mm (as compared to 8.4 mm in the first patient), and did not dilate or become more tortuous in serial image study. His systolic BP was never above 185 mmHg, and was always below 155 mmHg after taking anti-HTN drugs treatment for years. The patient had a higher bleeding tendency than the first patient because of liver cirrhosis, he had EV and GI bleeding history, and his platelet count was always below the lower border range; prolonged international normalized ratio was also detected in the most recent two years. However, quite unusually, the patient still experienced recurrent ischemic stroke despite a tendency towards bleeding and long-term antiplatelet agent usage, and hemorrhagic stroke did not occur in this case.

The third patient had neurological symptoms and signs compatible with left medullo-cervical junction ischemia, and recovered totally after rt-PA treatment. The third patient’s VBD diameter was 6.5 mm, which was smaller than the first two patients, but VBD tortuous level was similar. We reviewed the related literature regarding VBD cases undergoing rt-PA or thrombolysis treatment, and they all reported having a poor outcome with rupture of the basilar artery and subarachnoid hemorrhage. Such findings are different than those in our case. There are many factors to consider with regards to the management of intracranial dolichoectasia. The risks of ischemic stroke in VDE patients are associated with older age (being more than 60 years old), male sex, arterial hypertension, and previous history of myocardial infarction. On the other hand, the occurrence of hemorrhagic events in patients with VDE increases with female sex, the degree and enlargement of ectasia (maximal diameter of basilar artery >6.4 mm), lateral displacement of basilar artery (score >1), arterial hypertension, and the use of antiplatelet/anticoagulant agents. As the studies indicate, the prevalence of ischemic stroke and intracranial hemorrhage both increase in patients with VBD as compared to healthy adults, and as the degree of ectasia on basilar artery extends, the risks of both ischemic stroke and intracranial hemorrhage also increase. Many neurologists focus on the prevention and treatment of recurrent infarction - they have good reason to do so as ischemic stroke contributes the most to the mortality rate of VDE patients - and they prefer to prescribe long-term antiplatelet or anticoagulant agents for patients with VDE. If thrombus is seen on an image study, no one would doubt the benefits of performing thrombolytic or anticoagulant therapy, whether it is symptomatic or not. However, it should be noted that this strategy does have its drawbacks, for dolichoectasia is an arteriopathy rather than atherosclerosis, and as such the response to thrombolytic or anticoagulant therapy is not as efficient as atherosclerotic or embolic infarction, and it would increase the potential of VBD rupture.
patients with cerebral ischemia, the control of associated risk factors and the prevention of potential hemorrhage are not discussed. As to our management of the first patient, the usage of LMWH may be worth reconsidering because at present LMWH is not recommended in cases of basilar artery occlusion, stroke in evolution, or VDE. Whether or not it will be effective in cases of cerebral infarction and increase the risk of hemorrhage, as conventional antiagulant use for VBD patients is uncertain at present. On the contrary, the prevention effect of antiplatelet drugs on the second patient was quite poor, and we did not try anticoagulants because of his bleeding tendency, which raises the question that if anticoagulants are indeed better than antiplatelet drugs for recurrent ischemic stroke prevention. The good prognosis after rt-PA injection in our third patient suggests another point-of-view, that it is not the treatment agents that influence the outcome, but the size of VBD and the control of BP, which will determine the patient’s prognosis.

As to our opinion, the best conventional management of VDE may be the control of arterial hypertension. According to series studies, arterial hypertension not only plays a role in the formation and enlargement of intracranial dolichoectasia, but also contributes to the increased rupture potential and the occurrence of infarction. Series image study could determine the enlargement or thrombus formation of VDE at an early stage, and help to identify when to intervene in the evolution of VDE. In conclusion, there is still plenty of room for research on intracranial dolichoectasia, with more large series studies commencing at this time, and we hope that more evidence for managing this distinct vascular disorder will emerge in the very near future.

References