Vertebrobasilar Dolichoectasia Complicated with Arterial Dissection and Acute Posterior Circulation Infarctions

HHC Tsang, SSM Lo, WL Poon, KW Tang
Department of Radiology and Imaging, Queen Elizabeth Hospital, Jordan, Hong Kong

ABSTRACT
Vertebrobasilar dolichoectasia is defined as elongation, dilatation, and tortuosity of the basilar and vertebral arteries. We describe two patients with vertebrobasilar dolichoectasia, one who presented with acute posterior circulation infarct and subsequent arterial dissection, and the other who presented with acute posterior circulation infarct, arterial dissection, and hydrocephalus at presentation. These cases serve as a reminder of the clinical significance of the presence of vertebrobasilar dolichoectasia and its potential complications.

Key Words: Basilar artery; Intracranial arteriosclerosis; Ischemic attack, transient; Stroke; Vertebrobasilar insufficiency

中文摘要
椎基底動脈延長擴張症併發動脈夾層和急性後循環梗塞
曾凱晴、羅尚銘、潘偉麟、鄧國穎
椎基底動脈延長擴張症是指椎動脈和基底動脈的異常延長、擴張和迂曲。本文描述椎基底動脈延長擴張症的兩個病例：其中一例起病為急性後循環梗塞，繼發動脈夾層；另一例症狀為急性後循環梗死，動脈夾層和腦積水，本報告指出椎基底動脈延長擴張症的臨床重要性以及其潛在的併發症。

INTRODUCTION
Vertebrobasilar dolichoectasia (VBD) is defined as elongation, dilatation, and tortuosity of the basilar and vertebral arteries. VBD is a condition with a variable clinical course, which may be asymptomatic or present with various complications. Known complications of VBD include compression of the cranial nerves or brainstem related to mass effect, ischaemia or haemorrhage in the vertebrobasilar territory, and obstructive hydrocephalus.1-4

CASE REPORTS
Case 1
A 43-year-old man with a history of hypertension, hyperlipidaemia, and obstructive sleep apnoea presented to Queen Elizabeth Hospital in Hong Kong with sudden onset of left-sided weakness and numbness, which resolved on admission in October 2012. Physical examination revealed no focal neurological deficit. Urgent computed tomography (CT) of the brain showed ectatic basilar, bilateral internal carotid and
middle cerebral arteries (Figure 1). Urgent magnetic resonance imaging (MRI) of the brain confirmed ectatic intracranial arteries without any evidence of arterial dissection. Ectatic vertebral arteries and acute pontine infarct were also noted (Figure 2). There was significant upward shift of the basilar tip causing compression on the brainstem and cerebral aqueduct resulting in obstructive hydrocephalus. Medical therapy with an antiplatelet agent, namely aspirin (160 mg daily orally), was started. After 6 months, the patient presented with acute onset of left-sided weakness, which persisted after admission. Another MRI of the brain was performed, which revealed increased mural thrombus at the distal basilar artery (Figure 3). In view of the newly developed symptoms and clinical suspicion of basilar artery dissection, a diagnostic cerebral angiogram was performed, which depicted the presence of VBD and acute dissection of the right vertebral artery with extension into the basilar artery (Figure 4). Given the newly developed acute dissection of the vertebral and basilar arteries, anticoagulation with warfarin (2 mg daily orally) was started. Follow-up MRI of the brain performed after 7 months demonstrated an enlarging, abnormal contrast out-pouch at the basilar artery, suspicious of pseudoaneurysm formation (Figure 5). After multidisciplinary meeting between neurosurgeons and neuroradiologists, it was decided to continue with conservative management with warfarin (2 mg daily orally).

Case 2
In January 2014, a 58-year-old man presented to Queen Elizabeth Hospital in Hong Kong with dizziness, vertigo, and vomiting that had increased in severity over 3 days. Physical examination revealed right sixth and seventh cranial nerve palsies. There was no limb weakness on examination. An urgent unenhanced CT scan of the brain showed markedly ectatic basilar and right vertebral arteries (Figure 6). MRI of the brain showed VBD and suspected basilar artery dissection. Acute pontine infarct was also noted (Figure 7). A diagnostic cerebral angiogram was performed and confirmed the clinically and MRI-suspected acute dissection of the basilar artery. The patient was given antiplatelet and anticoagulation therapy with aspirin (80 mg daily orally) and warfarin (2.5 mg daily orally). Follow-up 6 months after initiation of treatment did not reveal any evidence of complications such as further ischaemic stroke.

DISCUSSION
VBD is a disorder characterised by elongation and dilatation of the basilar and vertebral arteries in the posterior circulation. Vertical elongation of the vertebrobasilar system can be graded as: 0 (at or below the dorsum sella), 1 (within the suprasellar cisterns), 2 (at the level of the third ventricle floor), and 3 (indenting and elevating the floor of the third ventricle). The basilar artery is considered elongated if it lies lateral to the margin of the clivus or dorsum sella, or if it bifurcates above the plane of the suprasellar cistern. Ectasia can be considered when the basilar artery is dilated to a diameter of >4.5 mm or >4 mm in the vertebral artery.

Digital subtraction angiography is the gold standard for diagnosis of cerebrovascular disease. For the diagnosis of VBD, however, non-invasive methods with CT and MR angiogram are sufficient. Although CT angiogram was once shown to overestimate the size of the basilar artery dilatation in VBD, this error has been overcome by the availability of thin-slice source images. With the aid of three-dimensional volume-rendered reconstruction, the anatomical relationships between the dilated vertebrobasilar system and adjacent structures can be readily and clearly demonstrated. In

Figure 1. An axial unenhanced computed tomography image of the brain of case 1 shows ectactic bilateral internal carotid (white arrows), bilateral middle cerebral (white arrowheads), and basilar (white dotted arrow) arteries.
both of these patients, digital subtraction angiography was performed not only for the diagnosis of VBD, but also for the clinical suspicion of arterial dissection.

VBD has variable clinical manifestations. Although some patients are asymptomatic and VBD is discovered only as an incidental finding, some patients present with neurological symptoms. The most common symptom is ischaemic stroke, followed by brainstem and cranial nerve compression, hydrocephalus, and cerebral
haemorrhage.1,5-7 Passero and Rossi1 studied the location of the ischaemic infarcts and found that 42% were in the brainstem, followed by 32% in the thalamus, 12% in the posterior cerebral artery territory, 11% in the cerebellum, and 2% in other regions. Of the ischaemic infarcts involving the brainstem, most were localised within the pons.5 In the systematic review by Wolters et al,7 it was found that patients with VBD are at high risk of neurological deterioration during a 5-year follow-up, with ischaemic stroke and progressive brainstem compression being the most commonly observed conditions.
Different pathophysiological mechanisms have been described for ischaemic events in patients with VBD, including atherosclerosis, artery-to-artery embolism, dissection, and stagnant / turbulent flow within the tortuous and ectatic vessels. The two patients presented here displayed several complications as a result of VBD, namely acute ischaemia in the posterior circulation territory, cranial nerve compression, arterial dissection, dissecting aneurysm, and obstructive hydrocephalus. It was observed that dissection within the vertebral arteries extending to the basilar arteries had resulted in acute pontine infarction in both patients. Postulated pathophysiological mechanisms include occlusion of the small perforating branches of the basilar artery caused by a thrombosed false lumen or embolism. In the second patient, it could be argued that there was a previous dissection of the vertebral artery resulting in fusiform dilatation. However, the development of a post-dissection fusiform aneurysm would require a long period of time and it is unusual for a patient to remain asymptomatic after the initial dissection. Therefore, in view of the acute presentation of the infarct, it was more
likely to be a pre-existing VBD complicated by acute dissection.

**Management of VBD varies with patient’s symptoms.** To our knowledge, there is no consensus on the treatment of VBD in the literature. Medical treatment with anticoagulants may be considered, as ischaemic events are the commonest complication, but anticoagulant therapy carries an increased risk of haemorrhagic stroke. Antiplatelet agents such as aspirin may be an alternative treatment, although this is also associated with elevated risk of intracerebral bleeding.19 Surgical means may be employed in the event of obstructive hydrocephalus, with insertion of a ventriculoperitoneal shunt shown to be effective in some case reports.10,13 In this era of minimally invasive procedures, treatment of VBD by endovascular means has also been proposed. Wu et al14 performed coildo assisted reconstruction of VBD in nine patients. These authors were able to halt the natural morphological progression and prevent ischaemic strokes in about half of the patients up to 24 months after reconstruction. It is believed that stent reconstruction of the diseased ectatic arteries can modify the haemodynamics and, to some extent, normalise the morphology of dolichoectatic arteries. With such changes, blood flow can be restored to normal in the previously distorted branches, which may lead to ischaemia if left untreated. However, in view of the small sample size in Wu et al’s series,14 further studies with more patients and longer follow-up are required to validate the long-term effect of endovascular reconstruction of VBD in preventing stroke. Besides the use of coils and stents, with flow-diverting stents gaining popularity in treatment of aneurysms in the internal carotid arteries, their application in the vertebrobasilar system has also been studied. Siddiqui et al15 found substantial morbidity and mortality in the seven patients with fusiform vertebrobasilar aneurysms whom they treated with flow-diversion devices. This may be due to the inherent perforator-rich characteristics of the basilar system in comparison to the internal carotid arteries. Thus, further understanding of the haemodynamic effects of flow-diversion devices on brainstem perforators is needed.15

**CONCLUSION**

VBD is a condition that can be silent until complications develop. The two patients in this report illustrate the most common complication of VBD, namely ischaemic insult in the territory supplied by the dolichoectatic arteries. Although there is no consensus on the treatment of VBD, once the diagnosis is made, the management plan should include prevention of complications and monitoring of disease progression clinically and radiologically. When complications arise, treatment should then be tailored according to the type of complication and symptoms.

**REFERENCES**