Case Report

Dolichoectasia of vertebrobasilar arteries leading to vascular dementia: A case report

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Abstract

The term dolichoectasia stands for a dilated, elongated and tortuous vessel. Dolichoectasia of the cerebral vasculature is rare but well-described and commonly involves vertebral and basilar arteries. It may be an incidental finding or the patient may present with local compression leading to hydrocephalus, cranial nerve palsies or with ischemic symptoms. We present a case of dolichoectasia of basilar artery contributing to multiple cerebral thromboembolisms where the patient presented with cerebellar symptoms and signs with vascular dementia.

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Introduction

Dolichoectasia is a condition in arteries characterized by weakening of its wall leading to a dilated, elongated, and/or tortuous vessel. Endothelial damage in the intima and the turbulent flow of blood through the abnormally dilated vessel can lead to intra-arterial clot formation that ultimately gets embolised distally. Dolichoectasia is commonly described in the cerebral vasculature, mostly involving the vertebra basilar system. Thus, it can lead to multiple cerebral infarctions in supplied territories of the brain. This may be further triggered by underlying patients' risk factors for athero-embolism, such as uncontrolled hypertension, underlying dyslipidaemia, advancing age, and behaviours such as smoking (1).

The prevalence is around 0.08% to 6.5% in the general population. There is a high prevalence among patients who had a stroke, which is 3% to 17% (2), and in general vertebra-basilar dolichoectasia (VBD) is asymptomatic; as a percentage, it's around 1.3% (1). Patients may present with ischaemic, haemorrhagic, or compressive features, including rare presentation as hydrocephalus. VBD can also be associated with other vascular risk factors, including advancing age, male gender, hypertension, history of adverse vascular events as myocardial infarction, lacunar infarcts. It's not associated with carotid atherosclerosis (3).

Vascular dementia is defined as any dementia that is caused by cerebrovascular disease or impaired cerebral blood flow. To establish the diagnosis of vascular dementia, there should be neuroimaging evidence of cerebral ischemia or infarcts. As VBD itself can give rise to cerebral infarctions, it can be considered a risk factor for the development of vascular dementia.

Case report

A 62-year-old male with hypertension was brought to the National Hospital, Kandy, Sri Lanka, by relatives due to memory impairment and confused behaviour over two days. The patient had complained of vertigo and headache for the same duration. The patient had struggled to identify close family members, to recall their names, and had been unable to recall his place of living. There was also a history of an episodic memory loss expanding for around two decades back from the date of admission. There were no clues in the history to suggest central nervous system infection, trauma, or substance abuse. He was conscious and oriented in time and place on examination but could not identify the bystander as his son-in-law. He was found to be hypertensive with a blood pressure of 160/100 mm Hg and had horizontal nystagmus in nervous system examination. The rest of the system examinations were normal.

His non-contrast computerized tomography(CT) brain showed multiple infarctions in the right thalamus, bilateral basal ganglia, right-side internal capsule (anterior limb), bilateral external capsule, and bilateral corona radiata with evidence of arterial thrombosis and ventricular enlargement. There were also hyper-densities and enlargement of the bilateral vertebral arteries and the basilar artery. His contrast-enhanced CT scan with CT cerebral angiography showed possible saccular aneurysm arising from the basilar artery and extending up to the right vertebral artery together with the above findings. As CT angiography was doubtful, further investigations proceeded with cerebral digital subtraction angiography, which showed dolichoectasia of the basilar artery and acute tapering of the left posterior cerebral artery with the absence of cortical branches suggestive of thromboembolism.

Since the patient was found to have multiple cerebral infarctions, further investigations were carried to rule out other causes leading to multiple cerebral infarctions. His electrocardiogram (ECG) was in sinus rhythm, and a two-dimensional echocardiogram didn't reveal any intracardiac sources of thrombo-emboli. His erythrocyte sedimentation rate (ESR) was 05 mm/hour, and thrombophilia screening was negative. However, his lipid profile was deranged with high triglycerides and Low-density lipoprotein (LDL) cholesterols.

The patient was diagnosed with vascular dementia with multiple cerebral infarcts precipitated by hypertension, impaired lipid profile, and underlying dolichoectasia of the basilar artery. He was started on an antiplatelet, statin, and antihypertensive therapy.

Discussion

Dolichoectasia of the vertebrobasilar system is rare, though a well-described entity. This condition is defined as dilatation, elongation, and tortuosity of vertebral and/or basilar arteries. This anomaly is usually characterised by a thin arterial wall with widened external diameter due to degeneration of the internal elastic lamina with multiple gaps and thinning of tunica media due to deficiency of reticular fibers together with smooth muscle atrophy (4).

The diagnosis depends on the vertebral or basilar artery diameter, which should be more than 4.5 mm, or basilar artery length of more than 29.5 mm, or intracranial portion of the vertebral artery length of more than 23.5 mm (5). The diagnosis is previously done by catheter angiography. Nowadays, it's mainly supported by CT scan and magnetic resonance angiography (MRA) findings.

Patients with VBD may be asymptomatic or present as acute ischemia of the vertebra-basilar artery territory, compressive features related to compression of cranial nerves, brain stem, or third ventricle, and disastrous outcome with vascular rupture.

Of the patients who present first with brain infarction, 10% are found to have intracranial arterial dolichoectasia (66.7% involving VBD) (6). There is a high risk of recurrence of stroke when associated with VBD, and the risk is proportionate to the diameter of the vertebra basilar artery (7).

Most of the time, the ischemic infarction is located in the brain stem, often involving pons (8). Although advancing age, hypertension, and male gender are common risk factors for both VBD and atherosclerosis, it is now well known that posterior circulation infarctions can occur by VBD itself without underlying atherosclerotic disease. Where previous acceptance was that VBD itself was a complication of atherosclerosis. Atherosclerotic changes observed in dolichoectatic arteries are due to the anatomical alteration in these vessels (4).

Mechanisms by which VBD leads to transient or permanent ischemia include reduced anterograde blood flow leading to distal territorial ischemia (9,10), thrombus formation inside the dilated arterial segment due to stagnation of blood flow, and reduction of blood flow of branching vessels due to closure or the narrowing of branching vessel openings. The GENIC study has shown a relationship between dolichoectasia and penetrating artery disease (small vessel disease) (4).

Vascular dementia is characterised by the development of vascular cognitive impairment in the background of a history of stroke or neuroimaging evidence of cerebrovascular pathology in a patient without a prior history suggestive of non-vascular cognitive disorder.

Management of an underlying VBD depends on the clinical manifestation it has given rise to. In ischemic cerebrovascular events and cerebral small vessel disease, the treatment goal is to prevent the risk of future events by lifestyle modifications, the achievement of optimal blood pressure and blood sugar control, and starting appropriate lipid-lowering agents and antiplatelets. If VBD is further complicated with vascular dementia, as in this case, both cognitive-behavioral and physical rehabilitation has a key role in patient management (11).

In this case, the patient was educated regarding his condition, importance of lifestyle modifications, including smoking cessation, and was started on antihypertensives, a single statin, and antiplatelet treatment. Cognitive rehabilitation was arranged.

Conclusion

It's important to be kept in mind that underlying VBD may be the aetiology for vascular dementia in a patient presenting with multiple territory infarctions, including posterior circulation and in cases of lacunar infarcts in cerebral small vessel disease.

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