Clinical Case Report

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Severe vertebrobasilar dolichoectasia as a cause of obstructive hydrocephalus

A case report

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Abstract

Rationale: Dolichoectasia of the vertebrobasilar artery is a vascular anomaly characterized by marked elongating, widening, and tortuosity of the arteries. Although this anomaly is usually asymptomatic, it may present with ischemic symptoms or mass effect involving brainstem or cranial nerves.

Patient concerns: A 52-year-old male was admitted with headache and visual field defect.

Diagnoses: Computed tomography and magnetic resonance imaging showed noncommunicating hydrocephalus due to vertebrobasilar dolichoectasia.

Interventions: The patient underwent right-side ventriculoperitoneal shunt.

Outcomes: The patient's symptoms improved gradually, although visual symptoms persisted.

Lessons: Neurosurgeons need to keep in mind vertebrobasilar dolichoectasia as a rare cause of obstructive hydrocephalus for accurate diagnosis and swift treatment.

Abbreviations: CT = computed tomography, ICA = internal carotid artery, VBD = vertebrobasilar dolichoectasia.

Keywords: dolichoectasia, obstructive hydrocephalus, vertebrobasilar artery

1. Introduction

Vertebrobasilar dolichoectasia (VBD) is a condition characterized by marked elongation, dilatation, and tortuosity of the vertebral and basilar arteries.[1,2] Although this anomaly is relatively uncommon and generally asymptomatic, occasionally it may manifest clinically in compression of the cranial nerves and/or brainstem, ischemic symptoms, or intracranial bleeding.[3–5] Rarely, the dolichoectatic vertebral or basilar arteries may result in compression of the third ventricle or cerebral aqueduct, thus manifesting as noncommunicating hydrocephalus.[3,6–8] We here present a patient with a rare case of noncommunicating hydrocephalus due to VBD.

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2. Case report

A 52-year-old man presented to our outpatient department with a 2-week history of headache; he also complained of disturbed visual field. On physical and neurological examination, there was no abnormal finding except severe headache. All laboratory tests were within normal limits, and he had no previous medical history. On the presentation day, the patient underwent nonenhanced computed tomography (CT), CT angiography of the brain, and magnetic resonance imaging of the brain. On nonenhanced CT and CT angiography, there were elongated, wide, tortuous vertebrobasilar arteries and fusiform aneurysmal dilation of the left supraclinoid portion of the internal carotid artery: vertebral artery – 12 mm, basilar artery – 16 mm, and midsection of left supraclinoid fusiform aneurysm – 13 mm. Brain magnetic resonance imaging revealed dilated and thrombosed basilar artery adjacent to the right anterolateral aspect of the diencephalon (Fig. 1A–C).

The patient was discharged after supportive care for 1 week without any signs or symptoms. Two years after discharge, he visited the emergency room with chief complaints of severe headache, memory impairment, gait disturbance, and visual field impairment. The visual field examination revealed left-side homonymous hemianopsia (Fig. 2), and on brain CT scan, the frontal horns of both lateral ventricles and periventricular low densities were higher than they had been on previous CT (Fig. 3A and B). The diameter of the ectatic basilar artery increased to 26 mm, and it seemed to be compressing the right optic tract near the diencephalon (Fig. 3C). Sagittal CT scan showed that an expanded and elongated ecstatic basilar artery had compressed the midbrain and obstructed the cerebrospinal outflow at the level of the cerebral aqueduct (Fig. 3D). The patient underwent right-side ventriculoperitoneal shunt. Six months later, the symptoms gradually improved, although visual symptoms persisted.

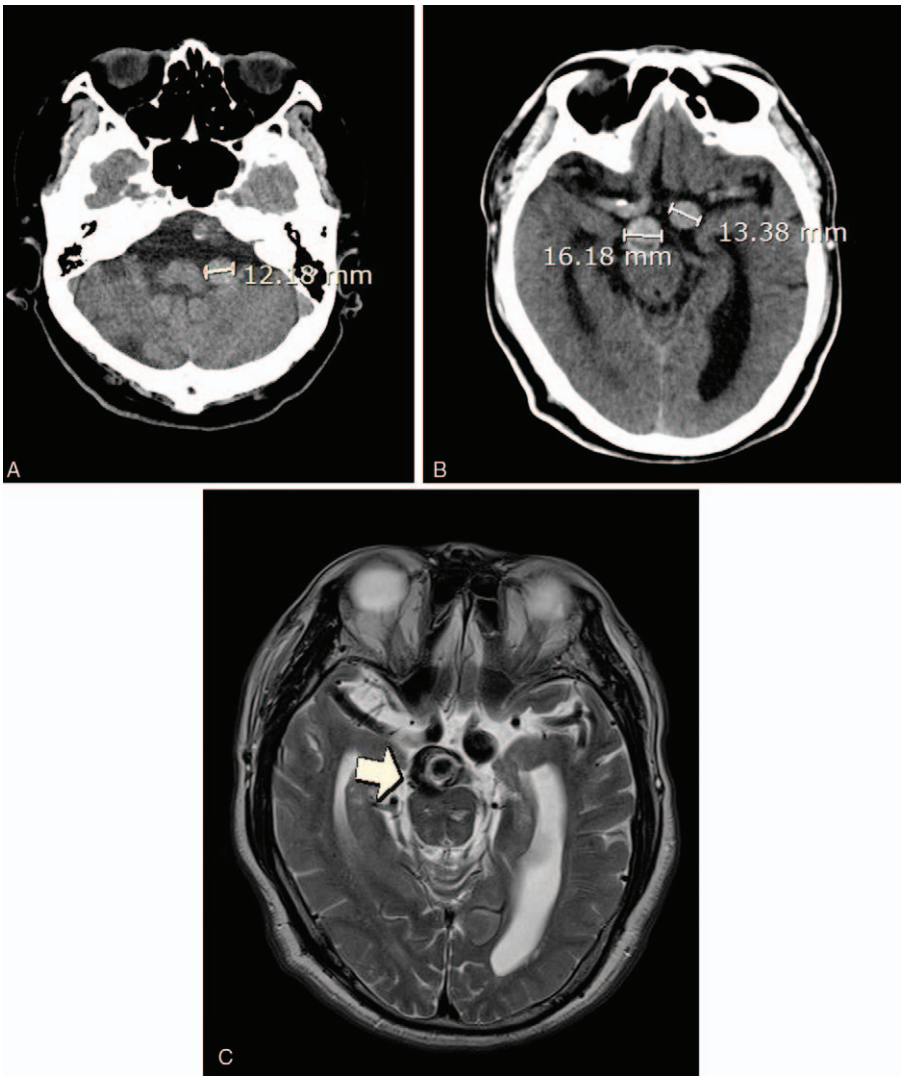

Figure 1. Nonenhanced CT revealed wide vertebrobasilar artery and fusiform aneurysmal dilation of the left supraclinoid portion of the ICA: vertebral artery – 12 mm, basilar artery – 16 mm, and midsection of left supraclinoid fusiform aneurysm – 13 mm (A, B). Brain MR T2-weighted imaging (C) revealed dilated and thrombosed basilar artery adjacent to the right anterolateral aspect of the diencephalon (arrow). CT = computed tomography, ICA = internal carotid artery.

3. Discussion

VBD is a rare but well-recognized vascular anomaly characterized by tortuosity, elongation, enlargement, and dilatation of vertebrobasilar arteries and subsequent hemodynamic changes.[1,2] The incidence of intracranial dolichoectasia ranges from 0.06% to 5.8%, with vertebrobasilar involvement being the most common segment affected.[2,9] The prevalence of VBD is 4.4%, and the primary location is the basilar artery only (40%), followed by the bilateral vertebral arteries, the basilar artery (22%), and both vertebral arteries (16%).[5] The bifurcation of the basilar artery is located in the interpeduncular cistern adjacent to the dorsum sellae or in the suprasellar cistern below the floor of the third ventricle, and the basilar artery lies in the pontine cistern within a space delimited by the lateral margins of the clivus and the dorsum sellae.[9,10] The mean diameter of the basilar artery ranges from 1.5 to 4 mm.[11] The diagnostic criteria for VBD are arterial diameter of over 4.5 mm at any location along its course and deviation of any portion of diameter higher than 10 mm from the shortest expected course, basilar length of over 29.5 mm, or intracranial vertebral artery length of over 23.5 mm.[5,9,10]

Although the pathophysiology of dolichoectasia is not clear, 2 distinguishable types were suggested: senile and juvenile. The senile type is associated with visible advanced atherosclerotic change such as aneurysms of the peripheral vascular system; the juvenile type is not associated with either atherosclerosis or hypertension. On histological examination, loss of internal elastic membrane and tunica media in the vessel walls was observed.[12] Therefore, it is supposed that this condition is related to inborn biochemical–histopathological abnormalities such as Marfan or Ehlers–Danlos syndromes.[4,12]

VBD is usually asymptomatic, and fewer than 10% of patients have neurologic symptoms.[13] Two types of symptoms were found with VBD: ischemic events and the symptoms resulting from compression of structures adjacent to the abnormal vessels. The ischemic symptoms resulted from recurrent thrombosis caused by hemodynamic and hemostatic changes within ectatic

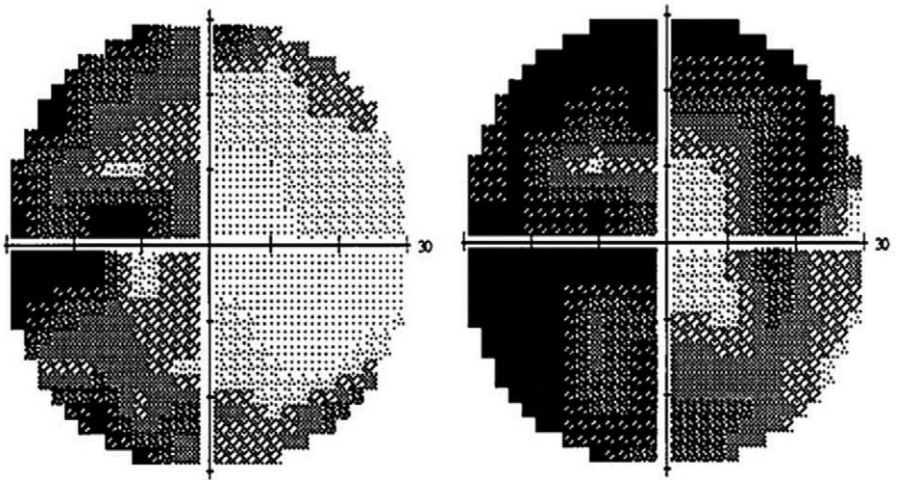

Figure 2. Visual field examination revealed left-side homonymous hemianopsia.

vessels.[1,2,7] Clinical presentations of compressive symptoms may be due to compression of the cervicomedullary junction or brainstem by ectatic vessels; compression at these regions may induce cranial nerve palsies or cerebellar or brainstem dysfunctions.[4,7,8,13] Based on analyses, in most cases of VBD, symptomatic cranial nerve compressions are facial nerve paralysis, trigeminal neuralgia, or lower cranial nerve involvement such as sleep apnea.[3,7,12] Interestingly, in our case, the

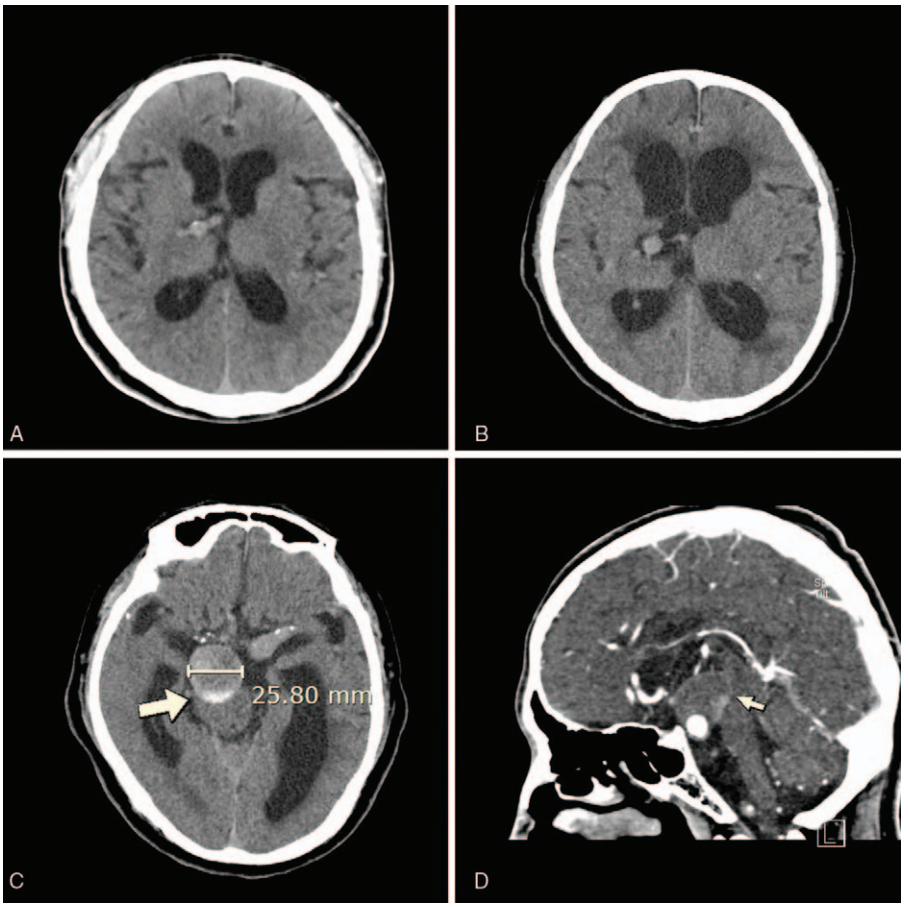

Figure 3. CT scan of the brain 2 years later (A) compared with initial CT (B). The frontal horns of both lateral ventricles and periventricular low densities were larger. The diameter of the ectatic basilar artery increased to 26 mm (C), and the right optic tract near the diencephalon was compressed (arrow). Sagittal CT scan (D) showed thrombosed and expanded ecstatic basilar artery compressed at the midbrain, resulting in obstructed cerebrospinal outflow at the cerebral aqueduct level. CT = computed tomography.

right optic tract was compressed by a thrombosed ectatic basilar artery, and to our knowledge, this is the first report of an optic pathway insulted by VBD.

Hydrocephalus is an uncommon complication of VBD, and most cases are not obstructive but normal-pressure hydrocephalus.[3,6–8] This is because hydrocephalus in VBD can be caused not only by direct obstruction of the foramen of Monro or the cerebral aqueduct by ectatic, elongated, and tortuous basilar arteries but also by pulsatile compression of the third ventricle or foramen of Monro. The mechanism of communicating hydrocephalus onset is supposed to be a "water-hammering" effect on the foramen of Monro or the third ventricle floor due to pulsating blood in the ectatic arteries, which impairs cerebrospinal fluid outflow through the third ventricle.[6,14] Obstructive hydrocephalus in VBD had been rarely reported; in particular, hydrocephalus due to aqueduct obstruction via compression of midbrain such as our case is extremely rare.[6,8,15]

Hydrocephalus due to obstruction of cerebrospinal fluid outflow by VBD can be treated by ventriculoperitoneal shunting, but the shunting method differs depending on the level of obstruction. Patients with hydrocephalus due to obstruction at the foramen of Monro need biventricular shunts, but in cases of obstruction at the aqueduct level, single ventriculoperitoneal shunt insertion is sufficient.[8,16]

In summary, VBD is a well-known disease entity whose clinical expression may be due to compression of the cranial nerve or brainstem or to thromboembolic events in vertebrobasilar arterial territory. Very rarely, severe compression of the midbrain with obstruction of cerebral aqueduct by VBD may result in obstructive hydrocephalus, and in this situation, emergent ventricular decompression is clinically required. As such, neurosurgeons need to keep in mind VBD as a rare cause of obstructive hydrocephalus for accurate diagnosis and swift treatment.

Author contributions

Investigation: Jong Myong Lee.
Methodology: Jung Soo Park.
Supervision: Eun Jeong Koh.
Writing – original draft: Jong Myong Lee.
Writing – review and editing: Jung Soo Park, Eun Jeong Koh.

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