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Case Report

Obstructive hydrocephalus and facial nerve palsy secondary to vertebrobasilar dolichoectasia: Case Report

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Abstract

Background: Symptomatic hydrocephalus due to vertebrobasilar dolichoectasia is a rare occurrence.

Case Description: We report a patient who presented with acute confusion and vomiting. Neuroimaging revealed elongated and tortuous basilar artery indenting and elevating the floor of third ventricle causing obstructive hydrocephalus. Initially, the patient was treated with external ventricular drain and then with ventriculo-peritoneal shunt.

Conclusion: We suggest prompt surgical intervention upon diagnosis as a first choice of treatment in order to avoid further complications.

Key Words: Facial nerve palsy, hydrocephalus, vertebro-basilar dolichoectasia, vp shunt

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INTRODUCTION

Vertebrobasilar dolichoectasia (VBD), also known as dilative arteriopathy, is tortuosity, elongation, and dilatation of the vertebral/basilar arteries or their junction. VBD is a known clinical entity with a patient presentation ranging from severe symptoms such as ischemic/hemorrhagic stroke, thromboembolic lesions, or with cranial nerve compression. They may also have varied clinical presentations, which includes tic douloureux, neuralgia, tinnitus, vertigo, motor or sensory deficits, ataxia, dementia, headache, migraine, leukoencephalopathy, central sleep apnea, and cerebellar dysfunctions; patients may also be asymptomatic[1] with most cases being diagnosed incidentally. Symptomatic hydrocephalus due to VBD is a rare occurrence.

CASE REPORT

Our patient is a 51-year-old Filipino female who had been recently diagnosed with hypertension, non-compliant with medication, presented to the emergency department with acute confusion for 1 day and repeated vomiting. There was no history of loss of consciousness or seizures. On admission, her blood pressure was 198/120 mm Hg, pulse was 92/min, and respiratory rate was 20/min. On examination, her glasgow coma scale (GCS) was 14/15, pupils were 3 mm, equal and reactive bilaterally. No evident cranial nerve palsy was observed. There was no papilledema.

Patient underwent CT scan brain and CT angiogram (CTA), which showed dilatation of both lateral and

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the third ventricles with periventricular hypo densities suggestive of transependymal cerebrospinal fluid (CSF) permeation. No intracranial hemorrhage was seen [Figure 1]. The fourth ventricle was normal. The patient underwent external ventricular drain (EVD) insertion for decompression of the ventricular system, following which the patient's GCS improved. Postoperatively, left facial palsy was noted.

The following day, we performed MRI with MR angiogram (MRA) and CSF flow studies. This revealed residual dilatation of both lateral and the anterior part of the third ventricle [Figure 2]. MRA revealed a dominant left vertebral artery, an elongated basilar artery reaching 18 mm above the dorsum sellae, which was elevating and compressing the posterior floor of the third ventricle and aqueduct, hence causing dilatation of the anterior third ventricle. The basilar artery was not significantly dilated and was slightly deviated off the midline. CSF flow studies did not show any flow in the third or lateral ventricles. Good flow was noted in the fourth ventricle, cistern magna, and pre-pontine cistern. Few incidental lacunar infarcts were seen in the thalamus and basal ganglia.

On retrospect, the third ventricular compression and the elevation of the basilar bifurcation was overlooked on the initial CTA. MRI and MRA were useful in settling the diagnosis.

Finally, the external ventricular drain was converted to ventriculo-peritoneal shunt and was subsequently discharged.

DISCUSSION

The incidence of intracranial dolichoectasia varies between 0.06–5.8%. Dolichoectasia is most common in the vertebral and basilar arteries.[3,11] Most of the time it is an incidental finding with 40% of the patients being asymptomatic. Hypertension is the most common co-morbidity found in patients with VBD.[6]

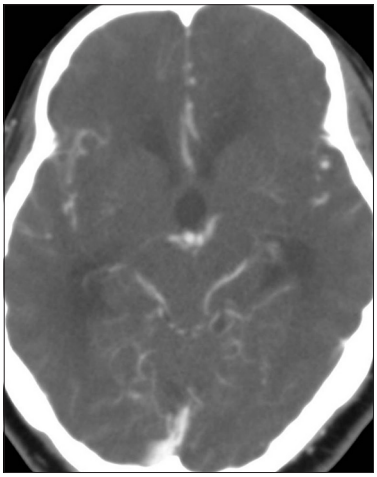

Figure 1: Axial Maximum Intensity Projection (MIP) image of a CT angiogram showing dilation of the frontal and temporal horns of the lateral ventricle and the anterior third ventricle (white arrow). The basilar bifurcation (black arrow) is seen at the posterior end of the third ventricle

Normally, the bifurcation of the basilar artery is located in the interpeduncular cistern adjacent to the dorsum sellae or in the suprasellar cistern below the level of the floor of third ventricle.

The diagnostic criteria for VBD is a basilar artery >4.5 mm in diameter, deviation of any portion of both arteries more than 10 mm from its shortest expected course, a basilar artery length >29.5 mm or intracranial vertebral length >23.5 mm.[8]

Vertical elongation of the vertebrobasilar arteries can be graded, according to Smoker *et al.*,[5] as

0: at or below dorsum sellae
1: Within suprasellar cistern
2: at the level of third ventricle floor
3: indenting and elevating floor of the third ventricle.

Passero *et al.*[6] in a detailed prospective study followed up 156 patients with VBD. In their study, VBD appears to be progressive with significantly increased morbidity and mortality. Most patients with VBD eventually developed a vascular event and stroke is the common cause of death in these patients. The cumulative proportion of VBD survivors, free of events is 54.1% at 5 years, 39.5% at 10 years, and 23.5% at 15 years.

Symptomatic patients with normal caliber but tortuous basilar artery are more likely to have isolated cranial nerve involvement, where as the patient with marked basilar artery dilation were far more likely to present with compressive or ischemic neurological deficits.[4]

VBD is an ischemic stroke risk factor. Intracranial VBD shows much more frequent occurrence of small vessel disease in stroke patients. The small blood vessel disease

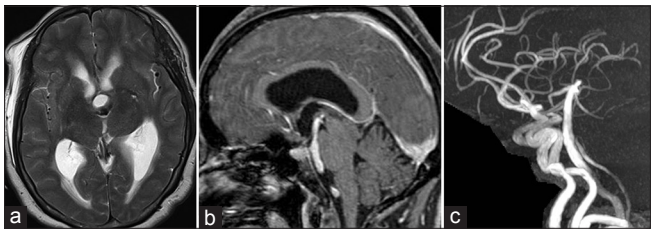

Figure 2: (a) Axial T2W, MRI image at almost a similar level showing, residual ventricular dilation and mild periventricular edema after insertion of the ventricular drain (small associated right frontal sub-galial collection). Note the basilar artery bifurcation (black arrow) indenting the posterior end of the dilated third ventricle. (b) Sagittal contrast-enhanced T1W image showing the elongated basilar artery (black arrow) elevating and causing compression and collapse of the posterior third ventricle. Contrast this with the dilated anterior third ventricle (white arrow). (c) Lateral MIP of a Time of Flight (TOF) MRA confirming the elevation of the basilar bifurcation (black arrow) above the carotid syphon (white arrow) normally situated lateral to the sella turcica

damage is dominated by multi-lacunar infarction, leukoriasis, and etat crible'. Intracranial bleeding in patients with VBD is not uncommon as previously believed.[7]

Hydrocephalus in VBD can be due to compression of the third ventricle due to ecstatic elongated basilar artery. Only few cases of hydrocephalus due to direct compression of the aqueduct, foramen of Monro, or the third ventricle have been reported in the literature. Another peculiar mechanism of hydrocephalus by "Water Hammering" effect due to the pulsatile blood in the ecstatic vessels which creates CSF flow impairment through the third ventricle has been described.[2]

Zisimopoulou *et al*. in their study suggested that the underlying mechanism for obstructive hydrocephalus due to vertebrobasilar dolichoectasia are both a *water-hammer effect* and a direct compression of adjacent structures.[9]

In our patient, there was a very high vertebral fusion to form the basilar artery. The basilar artery ran 18.4 above the dorsum sellae. Its maximum diameter reached 4.1mm. Since the obstruction was at the level of third ventricle, the patient benefitted from uni-ventricular shunt. Only few reported cases with obstruction at the level of foramen of Monroe required biventricular shunt.

Although the diagnosis was evident on the MR and MRA, it could not be seen on the initial CTA. Thin layer scans of high-resolution CT can to some extent avoid misdiagnosis caused by petrous bone artifacts - as suggested by Yong-Jie Yuan *et al*. in their study.[10] Hayder Kadhum Hassoun *et al*. also highlighted the need for high index of suspicion and special attention during brain imaging for the diagnosis of VBD.[1]

For an unclear reason, most of the cases of hydrocephalus complicating VBD have been reported from India. It is not clear whether VBD or its complications are more common in the Indian subcontinent or if this was simply a reporting bias.

Also, it is not clear why the VIIth nerve palsy was noted only following the ventricular shunt insertion. We believe that since it was subtle it might be there since start but was noticed only when the patient was evaluated thoroughly the following morning after the emergency shunt insertion. This is supported by the fact that the facial nerve palsy was on the right – the same to which the basilar artery is deviated.

CONCLUSION

Hydrocephalus is a rare but important presentation of VBD. This condition must be suspected whenever supra-tentorial hydrocephalus is evident in the presence of VBD. Review of the literature shows that VBD can have significant consequences and highlights the importance of identifying this finding, if present, whenever reporting on brain imaging studies. MRI or MRA are useful in confirming the diagnosis, if not digital subtraction angiography. Prompt surgical intervention upon diagnosis should be considered as a first choice of treatment in order to avoid further complications.

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Conflicts of interest
There are no conflicts of interest.

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