Vertebrobasilar dolichoectasia presenting as symptomatic obstructive hydrocephalus: A case report with review of literature

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ABSTRACT

Vertebrobasilar dolichoectasia (VBD) is a common, but usually asymptomatic condition characterized by marked elongation, dilatation, and tortousity of the vertebral and basilar arteries. VBD can sometimes present with symptoms related to mass effect like cranial nerve palsies, or with ischemia or hemorrhage. Hydrocephalus is an extremely uncommon presentation of VBD. We describe here a patient with VBD who presented with symptomatic hydrocephalus due to third ventricular obstruction, which was relieved by ventriculoperitoneal shunt.

Key words: CTA, hydrocephalus, MRI, shunt, vertebrobasilar dolichoectasia

INTRODUCTION

Vertebrobasilar dolichoectasia (VBD) is usually an asymptomatic condition characterized by marked elongation, dilatation, and tortousity of the vertebral and basilar arteries. Although diagnosed incidentally on imaging, they can sometimes present with either symptoms related to mass effect like cranial nerve palsies, or with ischemia or hemorrhage.[1] Hydrocephalus is an extremely uncommon presentation of VBD. We here report a patient with VBD presenting with obstructive hydrocephalus secondary to compression of the third ventricle.

CASE REPORT

A 45-year-old gentleman presented with complaints of headache and vomiting for 2 weeks. On examination, his blood pressure (BP) was 200/110 mmHg. He was conscious, alert and oriented, and had bilateral papilledema. He had no other neurological deficits. CT head revealed dilatation of both lateral ventricles and the anterior part of the third ventricle with a normal fourth ventricle [Figure 1]. Contrast CT demonstrated a severely ectatic, tortuous, and elongated basilar artery compressing the body of the third ventricle causing obstructive hydrocephalus. MRI brain demonstrated a dolichoectatic basilar artery bifurcating above the level of dorsum sella, elevating the floor of the third ventricle and compressing it against its roof, posterior to the foramen of Monro [Figure 2]. The left vertebral artery was also dolichoectatic. The maximum diameter of the left vertebral artery was 8 mm, and that of the basilar artery was 13 mm, near the vertebrobasilar junction. There was bilateral symmetric extensive periventricular white matter T2 and FLAIR hyperintensities, probably secondary to hypertension. There were no infarcts but numerous punctate foci of blooming suggestive of hemorrhages were present, predominantly in the posterior circulation territory.

He was initially put on intravenous antihypertensives to control the BP, and then maintained on oral antihypertensives. A right-sided ventriculoperitoneal (VP) shunt was performed to relieve the hydrocephalus. His headache resolved, and he was discharged on oral antihypertensives and aspirin. A CT angiogram performed 3 months after the VP shunting revealed decompressed ventricles indicating good functioning of the shunt, and the dolichoectatic vertebrobasilar arteries [Figure 3]. However, 4 months after presentation, the patient died of congestive cardiac failure.

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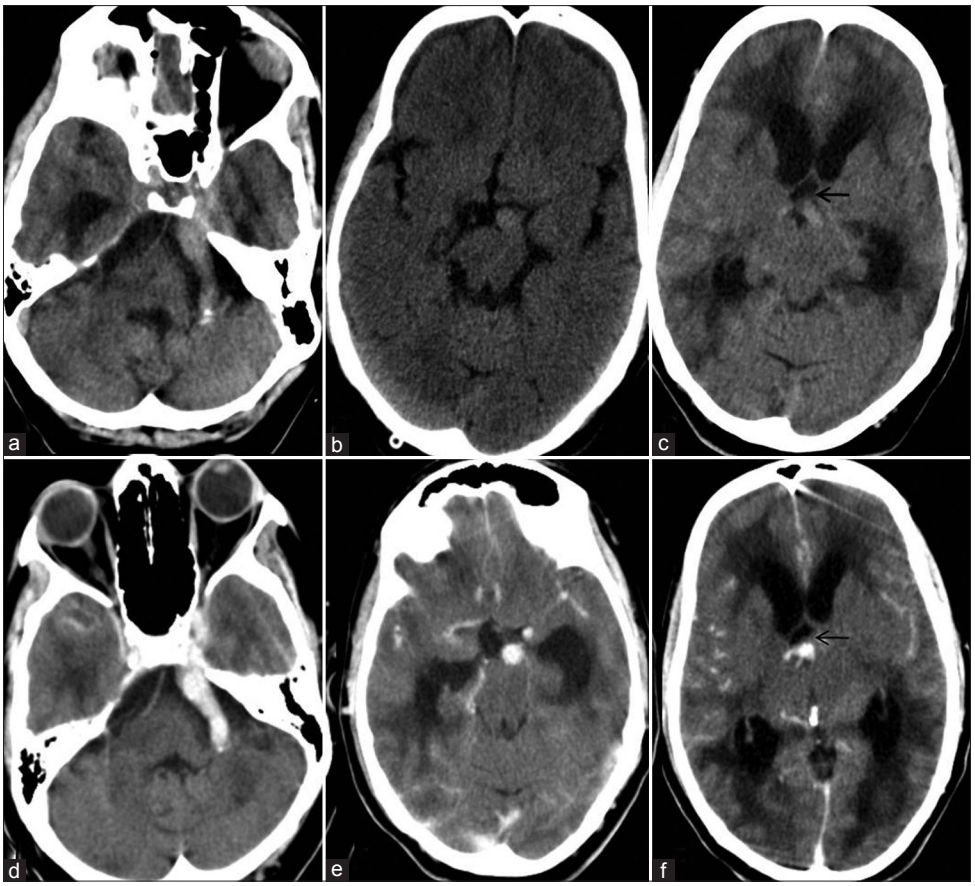

Figure 1: Plain (a–c) and contrast enhanced CT (d–f). Left vertebral artery (a, d) is dolichoectatic. Basilar artery (b, c, e, f), which is also dolichoectatic is compressing the third ventricle, causing dilatation of the anterior third ventricle (arrow) and bilateral lateral ventricles. There is periventricular diffuse hypodensities due to hydrocephalus

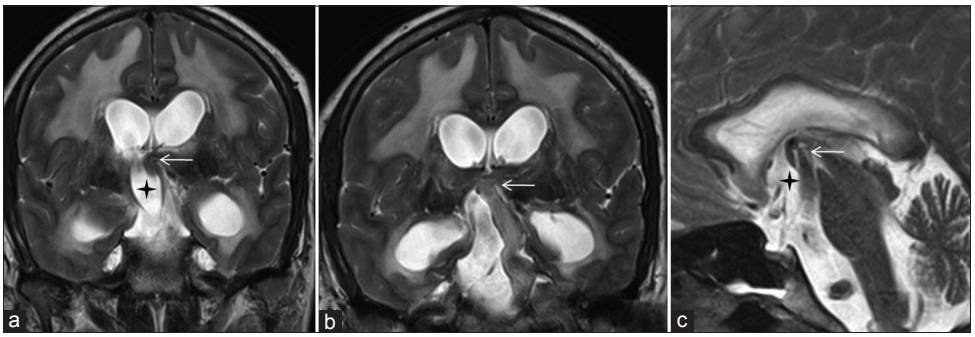

Figure 2: Coronal (a, b) and sagittal (c) T2WI. Dolichoectatic basilar artery is elevating the third ventricular floor, at the level of the interthalamic adhesion, posterior to the foramen of Monro, and compressing it against its roof, causing obstructive anterior third ventricular (star) and bilateral lateral ventricular hydrocephalus. Extensive white matter T2 hyperintensities are evident indicating interstitial edema

DISCUSSION

VBD is characterized by elongated, ecstatic, and tortuous vertebral and basilar arteries. The incidence of intracranial dolichoectasia is 0.06–5.8%, and is commonest in vertebral and basilar arteries.[2,3] A diameter of more than 4 mm of the vertebral or more than 4.5 mm of basilar artery is regarded as ectasia. Vertical elongation of the vertebrobasilar arteries can be graded as: 0 (at or below the dorsum sella), 1 (within suprasellar cisterns), 2 (at the level of third ventricle floor), and 3 (indenting and elevating floor of third ventricle). Our patient had grade 3 VBD. The basilar artery is considered elongated (dolicho) if it lies lateral to the margin of the clivus or dorsum sellae.

VBD is usually asymptomatic. The commonest clinical manifestation of VBD is ischemic stroke, most commonly in the pons.[4] Various pathophysiologic mechanisms have been described for the ischemic stroke, including

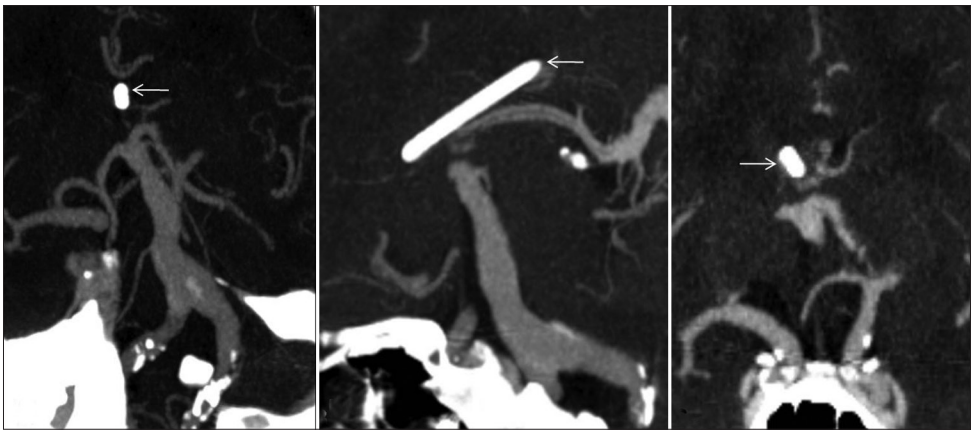

Figure 3: CT angiogram performed 3 months after placement of VP shunt (arrow). Vertebrobasilar arteries are dolichoectatic. Calcifications can be seen in bilateral vertebral arterial walls. Basilar artery is seen bifurcating much above the suprasellar region and approximating the floor of the third ventricle to its roof. VP shunt (arrow) is seen with its tip in the frontal horn of the right lateral ventricle

atherosclerosis, artery-to-artery embolism, and stagnant flow in the dilated and ectatic vessels. They can also present with hemorrhage, and cranial nerve compressive symptoms like hemifacial spasm and trigeminal neuralgia.[5] The symptoms and prognosis are related to the severity of ectasia and elongation of the arteries. Patients have a higher incidence of stroke and Transient ischemic attacks, and also cardiac complications because of co-morbidities. Stroke is the commonest cause of death in these patients.

Hydrocephalus is a very uncommon presentation of VBD. There are two different pathophysiologic mechanisms of hydrocephalus secondary to VBD. One is obstructive hydrocephalus secondary to elongation of the basilar artery and subsequent compression of the foramen of Monro, the cerebral aqueduct, or the third ventricle.[6-8] The other mechanism was described by Marinescu *et al*.,[9] where the 'water-hammering' effect on the foramen of Monro or the third ventricle floor due to pulsating blood in an ectatic artery causes CSF flow impairment and leads to normal pressure hydrocephalus. Ikeda *et al*.[10] evaluated 7345 adults with physical examination and MRI brain, and found VBD in 96 patients (1.3%). All the patients were asymptomatic, and MR evidence of hydrocephalus was found in four patients. Symptomatic hydrocephalus is exceedingly uncommon manifestation of VBD, and only 11 cases have been described in the literature so far. Hypertension was a common co-morbidity in these patients. In eight of these patients, the compression was at the level of the aqueduct, while in two patients, the compression was at the foramen of Monro (not specified in one patient). In our patient, the compression was unusual because it was at the middle of the third ventricle just anterior to the interthalamic adhesion. This is the first patient in which such a compression has been described in the literature.

The treatment of hydrocephalus secondary to VBD depends on the site of obstruction. Obstruction at the level of foramen of Monro may require biventricular shunts for decompression of both ventricles, while obstruction posteriorly or at the aqueduct will require univentricular shunt. In the present patient, the obstruction was posterior to the foramen of Monro, and hence a univentricular shunt was placed. There is no class I evidence about starting all these patients on antiplatelets for the primary prevention of stroke. However, as per the current management guidelines, aspirin should be used for the secondary prevention of stroke. Since these patients are also at risk for hemorrhage, long-term treatment with dual antiplatelets and anticoagulants is not recommended. If VBD presents with cranial nerve symptoms like trigeminal neuralgia or hemifacial spasm, microvascular decompression gives good clinical results.

CONCLUSION

VBD is usually asymptomatic. Symptomatic hydrocephalus is exceedingly uncommon manifestation of VBD resulting from severe elongation and ectasia of the basilar artery with subsequent compression of foramen of Monro, third ventricle or aqueduct of sylvius. VP shunting gives good relief of symptoms resulting from hydrocephalus, though the longterm outcome depends on the thromboembolic or hemorrhagic complications resulting from VBD, as well as the complications secondary to the long standing hypertension.

REFERENCES

1. Resta M, Gentile MA, Di Cuonzo F, Vinjau E, Brindicci D, Carella A. Clinical-angiographic correlations in 132 patients with megadolichovertebrobasilar anomaly. Neuroradiology 1984;26:213-6.

2. Giang DW, Perlin SJ, Monajati A, Kido DJ, Hollander J. Vertebrobasilar dolichoectasia: Assessment using MR. Neuroradiology 1988;30:518-23.
3. Smoker WR, Price MJ, Keyes WD, Corbett JJ, Gentry LR. High-resolution computed tomography of the basilar artery: 1. normal size and position. AJNR Am J Neuroradiol 1986;7:55-60.
4. Passero S, Filosomi G. Posterior circulation infarcts in patients with vertebrobasilar dolichoectasia. Stroke 1998;29:653-9.
5. Titlić M, Vrebalov-Cindro V, Lahman-Dorić M, Buca A, Jukić I, Tonkić A. Hemifacial spasm in vertebrobasilar dolichoectasia. Acta Neurol Belg 2006;106:23-5.
6. Thiex R, Mull M. Basilar megadolicho trunk causing obstructive hydrocephalus at the foramina of Monro. Surg Neurol 2006;65:199-201.
7. Rozario RA, Levine HL, Scott RM. Obstructive hydrocephalus secondary to an ectatic basilar artery. Surg Neurol 1978;9:31-4.
8. Siddiqui A, Chew NS, Miszkiel K. Vertebrobasilar dolichoectasia: A rare cause of obstructive hydrocephalus: Case report. Br J Radiol 2008;81:e123-6.
9. Marinescu M, Remy A, Dufour H, Derome P, Cambon H. A peculiar mechanism of hydrocephalus: The "water-hammering" effect. Neurochirurgie 1998;44:117-20.
10. Ikeda K, Nakamura Y, Hirayama T, Sekine T, Nagata R, Kano O, *et al*. Cardiovascular risk and neuroradiological profiles in asymptomatic vertebrobasilar dolichoectasia. Cerebrovasc Dis 2010;30:23-8.

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