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# Dolichoectasia of basilar artery presenting as non-communicating hydrocephalus-A rare presentation

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ABSTRACT

*Background:* Vertebrobasilar dolichoectasia is characterised by vascular elongation, tortuosity and widening of the vertebral or basilar arteries, with highly varied presentations. Hydrocephalus can rarely occur as a result of compression of the aqueduct of Sylvius or of the 3rd ventricle by the dolichoectatic basilar artery. Here, we present a case of basilar artery dolichoectasia leading to compression of the floor of the third ventricle resulting in obstructive hydrocephalus.

*Case summary:* A 67-year-old woman, hypertensive on regular treatment presented with a history of giddiness, difficulty while walking, slowness of gait and disorientation of two days duration. Her evaluation for metabolic and infective etiology were non-contributory. Magnetic resonance imaging of the brain done showed triventricular hydrocephalus with periventricular seepage of cerebrospinal fluid. Magnetic resonance angiogram of the brain with contrast showed dolichoectatic basilar artery compressing the floor of the third ventricle resulting in reduced flow of CSF through the aqueduct. Ventriculoperitoneal shunt (VP shunt) was inserted, following which there was a clinical improvement.

## 1. Introduction

Vertebrobasilar dolichoectasia (VBD) is a rare clinical entity characterised by significant elongation, dilatation and tortuous course of vertebral and basilar arteries with highly variable neurological signs and symptoms. The incidence of intracranial dolichoectasia ranges from 0.06% to 5.8%, with vertebrobasilar arteries being the most commonly affected segment [1].Rarely, the dilated and tortuous vertebral or basilar arteries may lead to compression of the aqueduct of Sylvius or, of the third ventricle resulting in a non-communicating hydrocephalus. Here, we present a rare case of an acute presentation of VBD as a non-communicating hydrocephalus.

## 2. Patient description

A 67-year-old woman, hypertensive on regular treatment presented with history of giddiness, difficulty while walking, slowness of gait and disorientation of two days duration. There was no history of fever, vomiting, dysuria, cough or seizures. She reported headache during the previous two weeks. On evaluation she was afebrile, drowsy with a GCS of E3V4M5 and blood pressure of 200/130 mm of Hg. Pupils were equal and reactive and she was moving all four limbs. She was admitted under Neuromedicine and worked up for metabolic and infective causes. Her metabolic parameters were within normal limits. Urine microscopic examination was normal. Her cerebrospinal fluid (CSF) study done to rule out infective causes showed 3 cells (all lymphocytes) with protein of 31 mg/dL and corresponding sugars. Her magnetic resonance imaging (MRI) of the brain done showed triventriculomegaly with periventricular seepage of CSF. (Fig. 1) Magnetic resonance angiogram of the brain with contrast showed dolichoectatic basilar artery compressing the floor of the 3rd ventricle resulting in reduced flow of CSF through the aqueduct. (Fig. 2)

Our patient had hypertension as a risk factor for VBD. The Neurosurgeon was called in to address the obstructive hydrocephalus as a result of VBD. The surgery decided upon was ventriculoperitoneal (VP) shunt. Endoscopic third ventriculostomy was not considered feasible as the introduction of the instruments to create a rent in the floor of the 3rd ventricle would be hazardous considering the close proximity of the basilar top which was impinging over the area. Bilateral VP shunt was not necessary, as the foramen of Monro was

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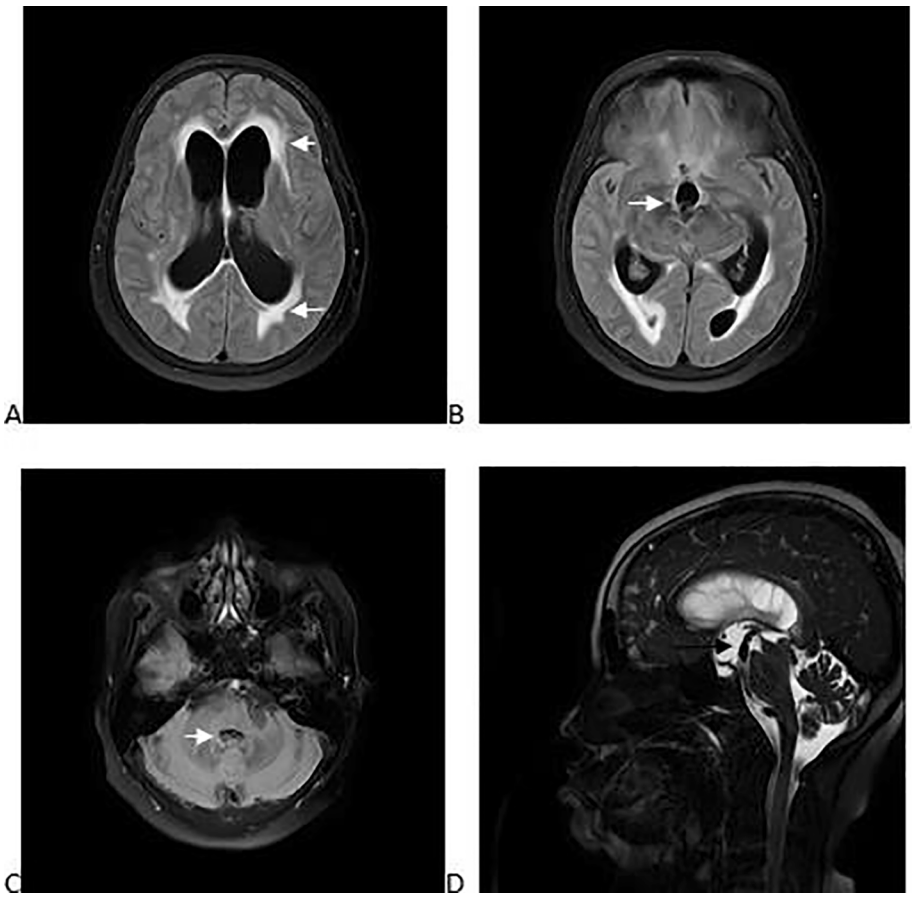


**Fig. 1.** Magnetic resonance T2 FLAIR axial images showing triventriculomegaly (Panel A, B and C). Panel A showing dilated lateral ventricles with periventricular seepage and panel B showing dilated 3rd ventricle(white arrow) with normal sized 4th ventricle(panel C). MRI T2 weighted ssagittal image showing dolichoectatic basilar artery (black arrow) compressing floor of 3rd ventricle with decreased CSF flow through the cerebral aqueduct (panel D).

patent. During surgery, the right lateral ventricle was hit at 4.5 cm depth. The entry point was through the right Keen's point. The CSF was clear and under high pressure and hence antibacterial coated medium pressure Chhabra shunt was used. (Fig. 2 Panel D) The post-operative period was uneventful. Her headache resolved during the postoperative period and she became ambulant and continent. She was discharged on the 10th post-operative day. She was doing well at 12-months follow up with the VP shunt functioning normally. Her cognitive assessment done at follow up, MMSE score 28/30 and MOCA 28/30 and there was no focal neurological deficits. Her blood pressure was normal, on three antihypertensive medications. Follow up CT brain imaging taken at 12-months after surgery showing reduction in ventricular size with no significant periventricular hypodensities, with VP shunt in situ, given in Fig. 3.

## 3. Discussion

Dolichoectasia is a rare clinical entity characterised by vascular elongation, widening, and tortuosity, with different clinical manifestations. The overall incidence of intracranial dolichoectasia ranges from 0.06% to 5.8%, of which vertebrobasilar arteries are the commonly affected segment. [1,2] The prevalence of vertebrobasilar involvement is 4.4%, with basilar artery segment being affected in 40% and vertebral arteries affected in 16% of cases. [3]

The basilar artery lies in the pontine cistern and near the dorsum sellae in the prepontine cistern it bifurcates or in the suprasellar cistern below the floor of the 3rd ventricle. [2] The mean diameter of the basilar artery ranges from 1.5 to 4 mm. The diagnostic criteria for VBD are arterial diameter more than 4.5 mm and above 10 mm deviation from the shortest expected course, basilar artery length more than 29.5 mm, or intracranial vertebral artery length more than 23.5 mm. [2,3]

Two subtypes of dolichoectasia were suggested, juvenile and senile subtypes, even though, the exact pathophysiological mechanism is not clear. The pathoanatomic findings in dolichoectatic arteries encompass abnormally large diameter and a thin arterial wall, with degeneration of the internal elastic lamina, thinning of the media and smooth muscle atrophy. The senile subtype is associated with arterial hypertension and atherosclerosis. Hypertension causes continued stress on the vessel wall resulting in degradation of the vessel wall leading to loosening of the collagen and elastin meshwork of the intima. [4] The juvenile type is not associated with either atherosclerosis or hypertension, but mainly associated with connective tissue disorders like Ehlers-Danlos syndrome and Marfan syndrome.

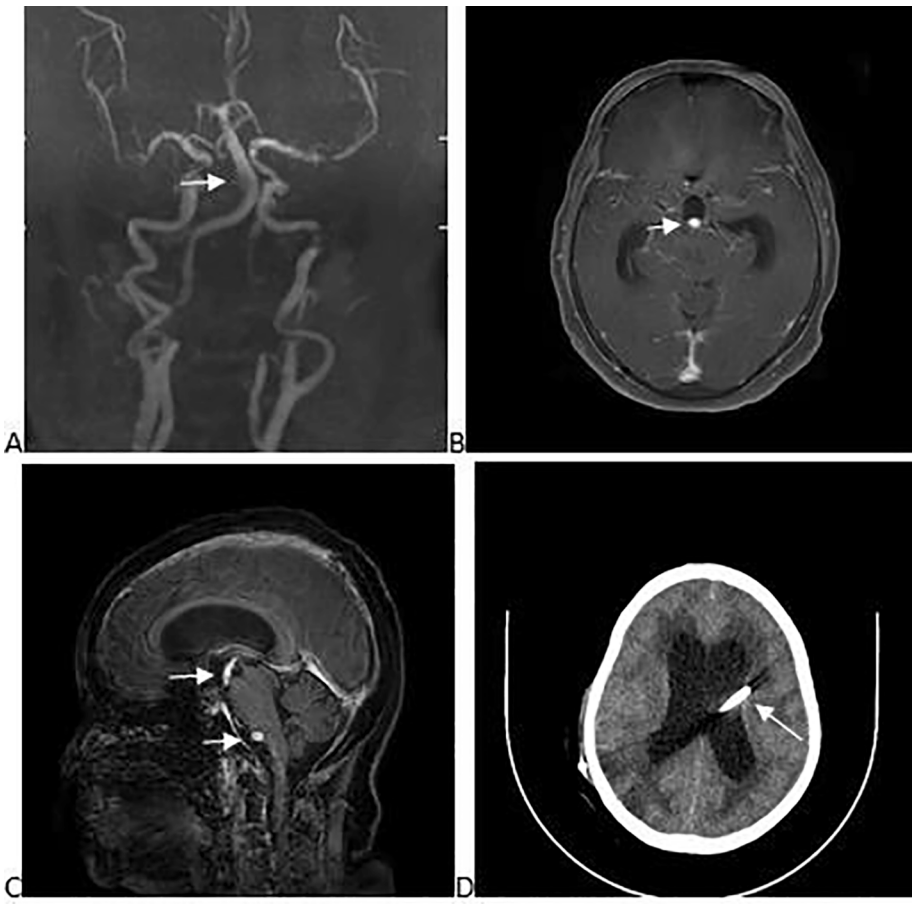


**Fig. 2.** Magnetic resonance angiogram showing dolichoectatic basilar artery (Panel A) shown by white arrow. MR Contrast axial images depicting basilar artery in the floor of third ventricle shown with white arrow in panel B and in sagittal images (panel C). Panel D showing post-operative plain CT image showing ventriculo-peritoneal shunt in the lateral ventricle (white arrow).

Vertebrobasilar dolichoectasia presents with ischemic stroke, haemorrhage or due to mass effect leading to multiple cranial neuropathies. Most cases of VBD are asymptomatic. Common sites of ischemic stroke are brainstem (41%), posterior cerebral artery territory (29%) and thalamus (22%). [5] VBD can produce mass effect over the brainstem. [6] The most common symptoms of cranial nerve involvement are trigeminal neuralgia and hemifacial spasm, which are caused by pulsatile compression of the trigeminal and facial nerve root by the ectatic vessels. [7] The occurrence of hydrocephalus as a result of VBD has been reported in a few studies. In a study by Ikeda et al, a total of 7345 adult subjects were investigated and 96 of them had asymptomatic VBD and only four of them had radiological evidence of hydrocephalus. [6]

Most cases of hydrocephalus in VBD are not obstructive but normal-pressure hydrocephalus. The “water-hammering” effect on the foramen of Monro or the third ventricle floor due to pulsating blood in the ectatic arteries, impairs CSF outflow through the 3rd ventricle further result in a communicating hydrocephalus. Non-communicating hydrocephalus in VBD had been rarely reported, in particular, an ectatic basilar artery compressing floor of the 3rd ventricle leading to aqueductal obstruction such as in our case is extremely rare. [7]

Management of VBD depends predominantly on the clinical symptoms. Various electrodiagnostic tests like blink reflex, brainstem auditory-evoked responses and motor-evoked potentials may be utilised for long-term monitoring in asymptomatic subjects as well as in the decision-making process prior to the surgical approach. Obstruction of the third ventricle or cerebral aqueduct can be managed with a unilateral VP shunt. A biventricular shunt may be needed when there is obstruction at the level of the foramen of Monro. [8] Endoscopic third ventriculostomy may be technically difficult in these cases due to the odd anatomy of the basilar trunk.

In our patient, hypertension was the risk factor for vertebrobasilar dolichoectasia. The VBD itself can be managed with good blood pressure control. Since the patient has no other comorbidities and her BP is well controlled, definitive management of the VBD was not considered. If VBD becomes symptomatic later in the form of TIA, stroke, aneurysm development or cranial neuropathies, definitive management in the form of endovascular intervention or bypass procedure may be considered.

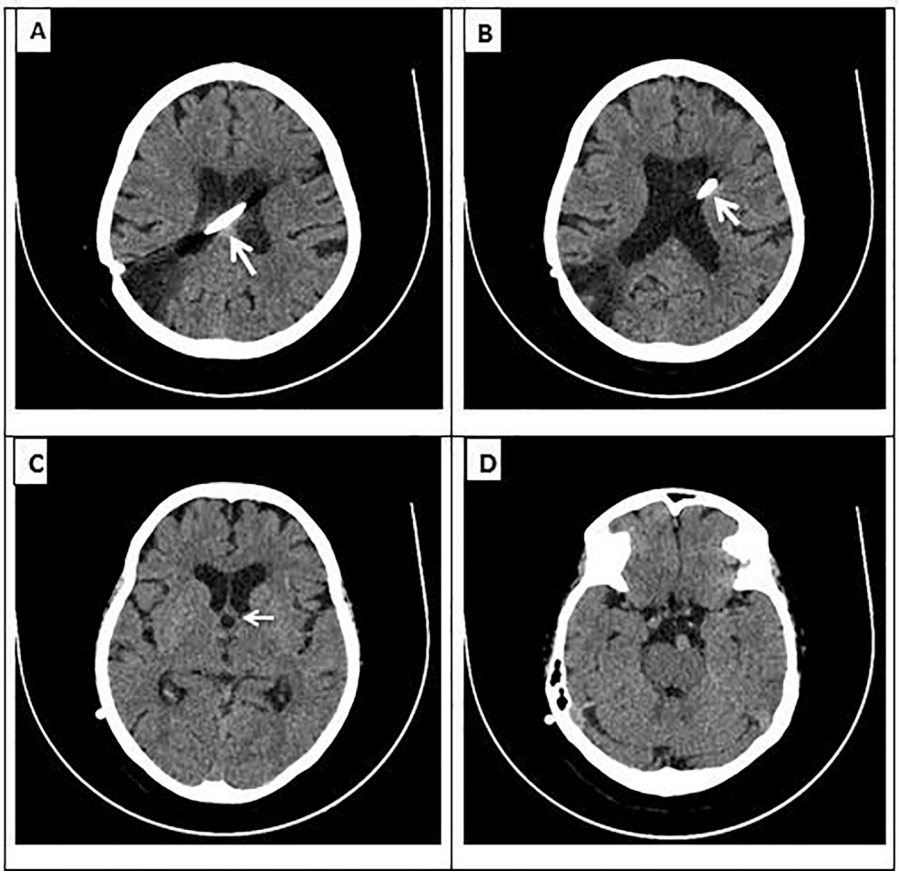


**Fig. 3.** Follow up CT brain image taken after 1 year with VP shunt in situ (shown in panel A and B marked with white arrows. Ventricular size has reduced with no significant periventricular hypodensities. 3rd ventricle marked in panel C with white arrow.

## 4. Conclusion

Non-communicating hydrocephalus due to compression of the basilar artery on the floor of the third ventricle is documented, though rare. In our case, obstructive hydrocephalus resulted from compression of the floor of third ventricle by the dolichoectactic basilar artery. Our case highlights, the physicians as well as neurologists to keep a low threshold to consider the possibility of such an etiology for an acute presentation of hydrocephalus, as an adequate intervention can result in a better outcome.

## Declaration of Competing Interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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