SNI

SURGICAL NEUROLOGY INTERNATIONAL

SNI: General Neurosurgery

OPEN ACCESS

For entire Editorial Board visit : http://www.surgicalneurologyint.com

Editor: C. David Hunt, M.D. Marquette General Neurosurgery, Brooklyn, NY, USA

Case Report

Hydrocephalus as a rare compilation of vertebrobasilar dolichoectasia: A case report and review of the literature

Keveh Ebrahimzadeh, Mehrdad H. Bakhtevari, Misagh Shafizad[1], Omidvar Rezaei

Department of Neurosurgery, Loghman e Hakim hospital, Shahid Beheshti University of Medical Sciences, Tehran, [1]Department of Neurosurgery, Emam Khomeini Hospital, Mazandaran University of Medical Sciences, Sari, Iran

E-mail: Keveh Ebrahimzadeh - dr.kavehebrahimzadeh@gmail.com; *Mehrdad H. Bakhtevari - mhbakhtevari@yahoo.com; Misagh Shafizad - shafizad@gmail.com; Omidvar Rezaei - info@drrezaee.ir
*Corresponding author

Received: 27 April 17 Accepted: 17 August 17 Published: 24 October 17

Abstract

Background: Vertebrobasilar dolichoectasia (VBD) is a rare disease characterized by significant expansion, elongation, and tortuosity of the vertebrobasilar arteries. Hydrocephalus is a rare compilation of VBD.

Case Description: In this study, we report a 68-year-old male presenting with headache, progressive decreased visual acuity, memory loss, imbalance while walking, and episodes of urinary incontinency. The patient was diagnosed with dolichoectasia of basilar artery causing compression of the third ventricular outflow and thus, presenting with hydrocephalus documented with brain computed tomography scan and brain magnetic resonance imaging. The patient underwent surgical operation and ventriculoperitoneal shunt placement.

Conclusion: In the case of hydrocephalus or normal pressure hydrocephalous, VBD should be considered as a differential diagnosis.

Key Words: Dolichoectasia, hydrocephaly, vertebrobasilar, water hammer, Windkessel

Videos available on: www.surgicalneurologyint.com

Access this article online

Website: www.surgicalneurologyint.com

DOI: 10.4103/sni.sni_162_17

Quick Response Code:

INTRODUCTION

Dolichoectasia is defined as a rare disorder of vascular elongation, widening, and tortuosity, with highly variable neurological symptoms and signs. Its prevalence in the adult population is estimated at 0.06–5.8% and affects most frequently the basilar and vertebral arteries, followed by the internal carotid artery and the middle cerebral artery.[1,25]

The main criteria for diagnosis of vertebrobasilar dolichoectasia (VBD) are a basilar artery or vertebral artery diameter >4.5 mm, deviation of more than 10 mm from the shortest expected course, basilar length >29.5 mm, or intracranial vertebral artery length >23.5 mm in magnetic resonance angiography.[5,18,20]

VBD can develop without any clinical symptoms, and most cases are asymptomatic. Symptoms of VBD can be divided into ischemic, hemorrhagic, and mass effect, with ischemic stroke the most common symptom and also the most common cause of VBD-related death.[4,14,24] In a study about common sites of ischemic stroke in VBD patients, it was estimated that 41% of infarcts occur in the brainstem, 29% in the posterior cerebral artery

How to cite this article: Ebrahimzadeh K, Bakhtevari MH, Shafizad M, Rezaei O. Hydrocephalus as a rare compilation of vertebrobasilar dolichoectasia:A case report and review of the literature. Surg Neurol Int 2017;8:256.
http://surgicalneurologyint.com/Hydrocephalus-as-a-rare-compilation-of-vertebrobasilar-dolichoectasia:-A-case-report-and-review-of-the-literature/

territory, 22% in the thalamus, 2% in the cerebellum, and 2% in other regions.[14]

The second most common symptom of VBD patients is brainstem and cranial nerve compression which can manifest as weakness, hypertension, cranial nerve palsy, and intracranial pseudospace-occupying lesions.[7,9] VBD progression rate is slow and symptoms of brainstem compression are uncommon; however, some minor changes, such as the blink reflex latency and changes in motor-evoked potentials in limbs, can be detected.[13,15] The most common symptoms of cranial nerve palsy are trigeminal neuralgia and hemifacial spasm, which are caused by VBD pulsatile compression of the trigeminal nerve root and facial nerve root.[3,23] Another symptom in VBD patients is headache. VBD-caused headaches are rare but are observed in clinical practice. In recent studies headache is shown as a clinical manifestation in VBD patients.[6,11]

CASE REPORT

A 68-year-old male presented with headache and progressive visual loss over 12 months. He also complained of memory loss, gait disturbance, and episodes of urinary incontinency for the past 4 months. The patient's past medical and psychological history were normal. In physical examination, there was a significant decrease in visual acuity of both eyes (able to detect finger count in <1 m distance). Neurological examination revealed bilateral optic atrophy as a sign of increased intracranial pressure (ICP) on fundoscopic ophthalmic examination.

Due to his presentation, brain computed tomography (CT) scan was performed, which showed severe hydrocephalus with periventricular edema. Further evaluation with magnetic resonance imaging (MRI) of the brain demonstrated dilated lateral ventricles and dilated basilar artery, which was extending into the suprasellar cistern compressing floor of the third ventricle.

The patient was diagnosed with dolichoectasia of vertebrobasilar arteries which caused hydrocephalus [Figure 1].

Because of the severe hydrocephalous, we recommended the patient for placement of a ventriculoperitoneal shunt. We avoided endoscopic third ventriculostomy because of the high risk of basilar artery injury. Endoscopic septostomy followed by unilateral ventriculoperitoneal shunting was performed and both Monro foramina and basilar pulsation were seen during endoscopic septostomy [Figure 2; Videos 1 and 2].

The postoperative period was uneventful, and the patient was discharged from hospital on the fifth postoperative day. Patient's headache resolved during postoperative period, gait imbalance, memory disturbance, and urinary incontinency improved gradually during 6 months. The visual acuity remained unchanged. Follow-up imaging after 6 months revealed relief of the hydrocephalus with normal-sized ventricles [Figure 3].

DISCUSSION

Mechanisms of hydrocephalus

It should be noted that hydrocephalus is a rare compilation of VBD. Estimated 5-year risk of progressive hydrocephalus is 3.3%.[22] Hydrocephalus is divided into obstruction-visible and obstruction-invisible based on radiological criteria.[24] Obstructive hydrocephalus is a rare complication of VBD. It is predominantly due to cerebrospinal fluid (CSF) circulation disorders caused by direct or indirect compression of the bottom of the third ventricle or midbrain aqueduct.[2,7,10,12,17,26] It is assumed that obstructive-visible hydrocephalus in VBD

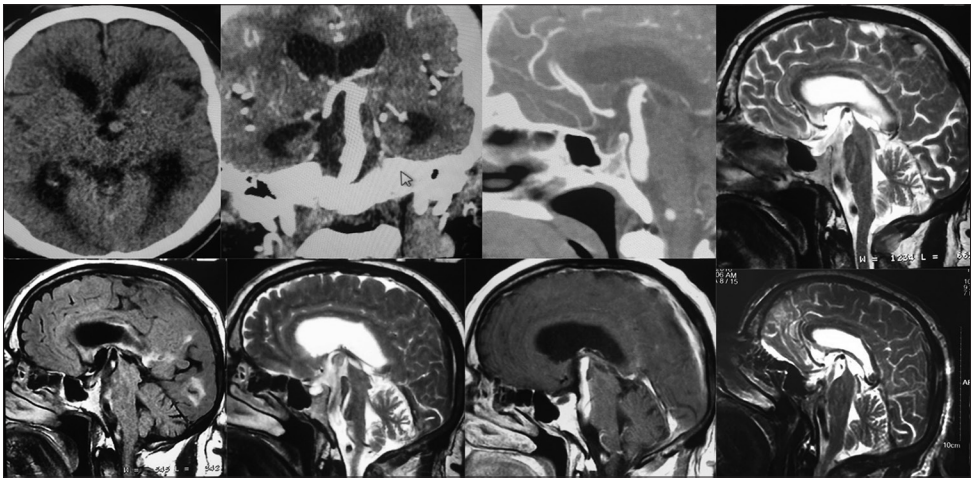

Figure 1: Brain imaging demonstrated dolichoectatic basilar with severe hydrocephalous. MRI demonstrated the presence of CSF in the posterior third ventricle and aqueduct

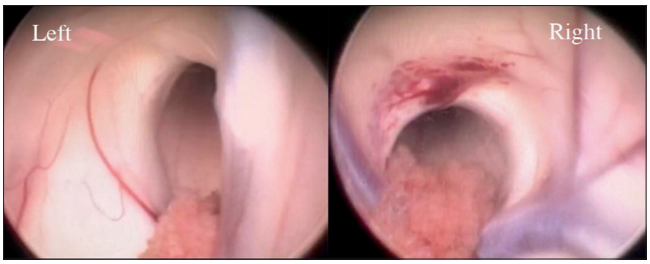

Figure 2: Intraoperative photograph of Monro foramen

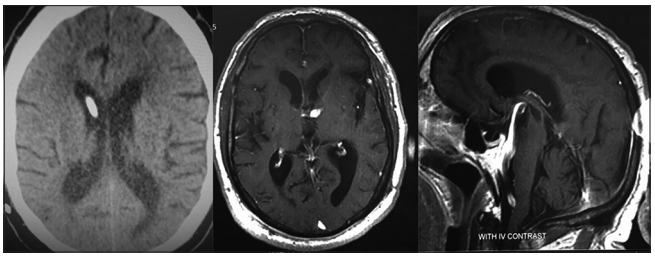

Figure 3: Postoperative imaging after 6 months revealed relief of the hydrocephalus with normal-sized ventricles

is a result of compression of the foramen of Monro, the cerebral aqueduct, or the third ventricle.[26] However, some other studies declare a different mechanism for obstructive-invisible hydrocephalus; that is, a combination of increased CSF pulse pressure and impairment of outward CSF flow by counter current pulsations of the basilar artery. The proposed mechanism for obstruction-invisible hydrocephalus is a "water hammer" effect within the bottom of the third ventricle or foramina of Monro, which is generated by pulsatile blood flow in the dolichoectatic arteries and results in hydrocephalus under normal pressure.[2,19,24] Strong CSF pulsations become a water hammer, wear out the brain, and cause ventricular dilation. In other words, a basilar artery extending into the floor of the third ventricle exerts a water hammer pulse transmitted toward the foramina of Monroe, resulting in an impairment of outflow from the lateral ventricles and initiate distension.[10,12]

To confirm a possible water hammer effect causing the dilation of the ventricles, we should have confirmatory imaging such as ventriculogram or MRI Cine study demonstrating CSF flow through the aqueduct. In our case, the relatively rapid clinical course, the loss of visual acuity, and the marked ventriculomegally involving the lateral and third ventricles while sparing the fourth ventricle, all argue in favor of obstructive hydrocephalus produced by compression of the posterior third ventricle and/or aqueduct by the dolichoectatic basilar artery. The rapid resolution of the ventriculomegally with shunting would also be extremely unusual for NPH and supports the diagnosis of obstructive hydrocephalus.

Literature review

There are few studies which have reported hydrocephalus as a consequence of VBD. In a study of Ikeda et al., 7345 adult subjects were investigated. They found that 96 of them had asymptomatic VBD [Table 1]. Among these 96 subjects, only four subjects had hydrocephalus as the neuroradiological finding.[4]

In 1998, Marinescu *et al*. reported a case in which an aneurysm of the basilar artery was revealed by an ischemic stroke in a 65-year-old man. Hydrocephalus developed during the following months. The MRI studies showed

Table 1: Literature review of vertebrobasilar dolichoectasia-induced hydrocephalous

Author/Year	Age/ Sex	Treatment
Breig *et al*. (1967)	3 cases	Not known
Ekbom *et al*. (1969)	6 cases	VA shunt
Rozario *et al*. (1978)	57/M	Unilateral VP shunt
Healy *et al*. (1981)	50/M	Unilateral VP shunt
Branco *et al*. (1993)	58/M	Not known
Aiba T and Nakazawa T (1995)	72/F	Unilateral VP shunt
Ricci *et al*. (2000)	66/M	Bilateral VP shunt
Weber *et al*. (2002)	69/M	Bilateral VP shunt
Thiex R and Mull M (2006)	54/M	Unilateral VP shunt
Siddiqui *et al*. (2008)	71/F	Unilateral VP shunt
Kansal *et al*. (2011)	60/M	Unilateral VP shunt
Seshadri *et al*. (2012)	45/M	Unilateral VP shunt
Çelik et al. (2013)	47/M	Unilateral VP shunt and endoscopic exploration
Zisimopoulou *et al*. (2015)	48/M	Refused surgical treatment
Our case (2016)	68/M	Endoscopic septostomy and unilateral VP shunt

that it could not be explained merely by a permanent compression. However, the patient improved clearly after a ventriculoperitoneal derivation.

Aneurysms of the basilar artery can cause hydrocephalus due to compression of the third ventricle or the Sylvian aqueduct. The observation of a particular case led to discuss another possible mechanism of hydrocephalus. The hydrocephalus could be explained by a "water hammering" effect due to the pulsating blood in the ectatic vessel, which created a CSF outflow impairment through the third ventricle.[10]

Breig *et al*.[2] reported three cases of VBD-induced hydrocephalus without obvious obstruction of the ventricular system.

In 2008, Siddiqui *et al*. described a case of a 71-year-old woman presenting with clinical features of raised ICP due to VBD producing obstructive hydrocephalus. The patient underwent emergency ventricular drainage, with resulting decompression of the ventricles and resolution of symptoms.[17]

In 2011, Kansal *et al.*[8] reported a case of a 60-year-old male with dolichoectasia of the basilar artery causing compression of the third ventricular outflow and, thus, presenting with noncommunicating obstructive hydrocephalus.

In a prospective study, Passero *et al.* investigated 156 patients with VBD for an average follow-up period of 11.7 years. Hydrocephalus occurred in only two cases (1.3%) of VBD. On the contrary, ischemic stroke occurred in 59 cases (37.8%), cranial nerve and brainstem compression occurred in 31 cases (19.9%), and cerebral hemorrhage occurred in 21 cases (13.5%).[13,23]

In 2015, Zisimopoulou *et al.* reported a 48-year old male with a profound dilatation of the ventricular system due to a dolichoectatic basilar artery. They mentioned that the patient suffered from longstanding hydrocephalus and presenile dementia. They suggested that the dilated basilar artery initially caused nonobstructive or obstructive-invisible hydrocephalus under the water hammer effect by impairment of CSF flow. That resulted to the aforementioned symptoms of evolving cognitive impairment and personality changes. Secondarily, as the VBD evolved by direct compression of the third floor ventricle, hydrocephalus worsened.[26]

According to our literature review, among the reported cases, Ricci *et al.*[16] and Weber *et al.*[21] managed hydrocephalous by planting bilateral ventriculoperitoneal shunt, and the other cases managed by one-sided ventriculoperitoneal shunting.

VBD-induced hydrocephalus is a very rare condition, which is commonly managed with ventriculoperitoneal shunt [Table 1]. The treatment of hydrocephalus secondary to VBD depends on the site of obstruction. Patients with bilateral obstruction of foramen of Monroe could be managed with bilateral ventriculoperitoneal shunt. In obstruction of the third ventricle or aqueduct of Sylvius, a unilateral ventriculoperitoneal shunt is sufficient.[17,19,24]

CONCLUSION

In the case of hydrocephalus or NPH, VBD and other vascular malformations should be considered as a differential diagnosis. We proposed that as in the NPH, Windkessel effect could be one of the probable cause of hydrocephalous in vertebrobasilar dolichoectatic-induced hydrocephalous by interfering with normal CSF circulation and pulsation.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

REFERENCES

1. Adams Jr HP, Hachinski VC, Norris JW. Ischemic cerebrovascular disease: Oxford University Press; 2001.
2. Breig A, Ekbom K, Greitz T, Kugelberg E. Hydrocephalus due to elongated basilar artery. A new clinicoradiological syndrome. Lancet 1967;1:874-75.
3. El-Ghandour NM. Microvascular decompression in the treatment of trigeminal neuralgia caused by vertebrobasilar ectasia. Neurosurgery 2010;67:330-7.
4. Flemming KD, Wiebers DO, Brown RD, Jr., Link MJ, Huston J, 3rd, McClelland RL, *et al.* The natural history of radiographically defined vertebrobasilar nonsaccular intracranial aneurysms. Cerebrovasc Dis 2005;20:270-9.
5. Giang DW, Perlin SJ, Monajati A, Kido DJ, Hollander J. Vertebrobasilar dolichoectasia: Assessment using MR. Neuroradiology 1988;30:518-23.
6. Huang L, Yu C-Y, Wang B-N, Zhang H-M, Li L-Y, Wang Y. Vertebrobasilar dolichoectasia causing a presentation resembling basilar-type migraine. Clin Neurol Neurosurg 2013;115:784-6.
7. 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.
8. Kansal R, Mahore A, Dange N, Kukreja S. Dolichoectasia of vertebrobasilar arteries as a cause of hydrocephalus. J Neurosci Rural Pract 2011;2:62-4.
9. Lou M, Caplan LR. Vertebrobasilar dilatative arteriopathy (dolichoectasia). Ann N Y Acad Sci 2010;1184:121-33.
10. Marinescu M, Remy A, Dufour H, Derome P, Cambon H. [A peculiar mechanism of hydrocephalus: The "water-hammering" effect]. Neurochirurgie 1998;44:117-20.
11. Moreira I, Mendonça T, Monteiro JP, Santos E. Hypnic headache and basilar artery dolichoectasia. The Neurologist 2015;20:106-7.
12. Nishizaki T, Tamaki N, Takeda N, Shirakuni T, Kondoh T, Matsumoto S. Dolichoectatic basilar artery: A review of 23 cases. Stroke 1986;17:1277-81.
13. Passero S, Rossi S, Giannini F, Nuti D. Brain-stem compression in vertebrobasilar dolichoectasia. A multimodal electrophysiological study. Clin Neurophysiol 2001;112:1531-9.
14. Passero SG, Rossi S. Natural history of vertebrobasilar dolichoectasia. Neurology 2008;70:66-72.
15. Pereira-Filho A, Faria M, Bleil C, Kraemer JL. Brainstem compression syndrome caused by vertebrobasilar dolichoectasia: Microvascular repositioning technique. Arq Neuropsiquiatr 2008;66:408-11.
16. Ricci G, Lenzi J, Esposito V. Hydrocephalus caused by dolichoectatic basilar artery. Case report. J Neurosurg Sci 2000;44:155-8.
17. Siddiqui A, Chew NS, Miszkiel K. Vertebrobasilar dolichoectasia: A rare cause of obstructive hydrocephalus: Case report. Br J Radiol 2008;81:27603660.
18. Smoker WR, Corbett JJ, Gentry LR, Keyes WD, Price MJ, McKusker S. High-resolution computed tomography of the basilar artery: 2. Vertebrobasilar dolichoectasia: Clinical-pathologic correlation and review. AJNR Am J Neuroradiol 1986;7:61-72.
19. Thiex R, Mull M. Basilar megadolicho trunk causing obstructive hydrocephalus at the foramina of Monro. Surg Neurol 2006;65:199-201.
20. Ubogu EE, Zaidat OO. Vertebrobasilar dolichoectasia diagnosed by magnetic resonance angiography and risk of stroke and death: A cohort study. J Neurol Neurosurg Psychiatry 2004;75:22-6.
21. Weber MA, Steiner T, Fiebach J. [Unusual cause of acute impairment of consciousness. Decompensated hydrocephalus caused by blockage of the foramen Monro due to enlarged basilar artery]. Radiologe 2002;42:833-6.
22. Wolters FJ, Rinkel GJ, Vergouwen MD. Clinical course and treatment of vertebrobasilar dolichoectasia: A systematic review of the literature. Neurol Res 2013;35:131-7.
23. Yang XS, Li ST, Zhong J, Zhu J, Du Q, Zhou QM, *et al.* Microvascular decompression on patients with trigeminal neuralgia caused by ectatic vertebrobasilar artery complex: Technique notes. Acta Neurochir 2012;154:793-7.
24. Yuan YJ, Xu K, Luo Q, Yu JL. Research progress on vertebrobasilar dolichoectasia. Int J Med Sci 2014;11:1039-48.
25. Yuh SJ, Alkherayf F, Lesiuk H. Dolichoectasia of the vertebral basilar and internal carotid arteries: A case report and literature review. Surg Neurol Int 2013;4:153.
26. Zisimopoulou V, Ntouniadaki A, Aggelidakis P, Siatouni A, Gatzonis S, Tavernarakis A. Vertebrobasilar dolichoectasia induced hydrocephalus: The water-hammer effect. Clin Pract 2015;5:749.