

# Target Sign of Third Ventricle in Basilar Dolichoectasia with Multiple Clinical Presentations: A Case Report

Akshay S. Patil<sup>1</sup> Sandeep Iratwar<sup>1</sup> Chetan Rathod<sup>1</sup> Paresh Korde<sup>1</sup>

<sup>1</sup>Department of Neurosurgery, Jawaharlal Nehru Medical College and Acharya Vinoba Bhave Rural Hospital, Sawangi, Wardha, Maharashtra, India

**Address for correspondence** Akshay S. Patil, MS, MCh, Department of Neurosurgery, Jawaharlal Nehru Medical College and Acharya Vinoba Bhave Rural Hospital, Sawangi, Wardha, Maharashtra 442001, India (e-mail: dr.akshayarbat@hotmail.com).

Indian J Neurosurg;2020;9:199–203

## Abstract

### Keywords

- basilar dolichoectasia
- hydrocephalus
- subarachnoid hemorrhage

Basilar dolichoectasia (BDE) is an uncommon anatomical variant usually detected incidentally or during stroke evaluation. BDE can occasionally become symptomatic and may present with stroke (infarct or hemorrhage), raised intracranial pressure due to obstructive hydrocephalus, or with cranial nerve palsies. We present a unique case of BDE presenting with obstructive hydrocephalus, stroke, and cranial nerve palsy in single patient and propose a radiological sign (target sign of third ventricle), which could aid in imaging diagnosis and further management.

## Introduction

Basilar dolichoectasia (BDE) is a rare anatomical variant of basilar artery with variable clinical presentation. It is usually detected incidentally and is essentially a radiological diagnosis. BDE may rarely present as posterior circulation ischemic stroke, intracerebral bleed subarachnoid hemorrhage (SAH), cranial nerve palsies, and rarely with obstructive hydrocephalus due to compression on midbrain, aqueduct, and floor of third ventricle or the foramen of Monro.<sup>1</sup> We are documenting a unique case of a 38-year-old male presenting with raised intracranial pressure (ICP) in the form of obstructive hydrocephalus and right facial lower motor neuron palsy. The imaging characteristics and diagnostic clues are also discussed.

## Case History

A 38-year-old male, laborer by occupation, presented with complaints of raised intracranial since 5 days. Neurological examination revealed papilloedema with right lower motor neuron type facial (VII nerve) palsy (H B grade III) and gait ataxia. Computerized Tomogram (CT) of the brain plain (► **Fig. 1**) revealed biventricular hydrocephalus with hyperdense lesion in the basal cistern and floor of third ventricle. Magnetic resonance imaging (MRI and MR angiogram; ► **Fig. 2**) revealed a dolichoectatic high-riding basilar artery (BA) compressing the

mid-third of the floor of third ventricle with resultant biventricular hydrocephalus. T1- and T2-weighted and fluid-attenuated inversion recovery (FLAIR) axial images of dolichoectatic BA showed target sign (BA along the floor of third ventricle with elevating the floor; ► **Fig. 1**). MRI brain also showed blooming on susceptibility-weighted images (SWIs) sequence along bilateral basal ganglia suggestive of micro bleed. The patient further underwent digital subtraction angiogram (DSA ► **Fig. 2**), which revealed an elongated high-riding BA 2.5 cm above posterior clinoid (Smoker's grade 3) and BA diameter of 6.4 mm was noted. Patient underwent right ventriculoperitoneal (VP) shunt in view of biventricular hydrocephalus following which his headache improved. He was initiated on antiplatelet prophylaxis to prevent further ischemic events.

## Discussion

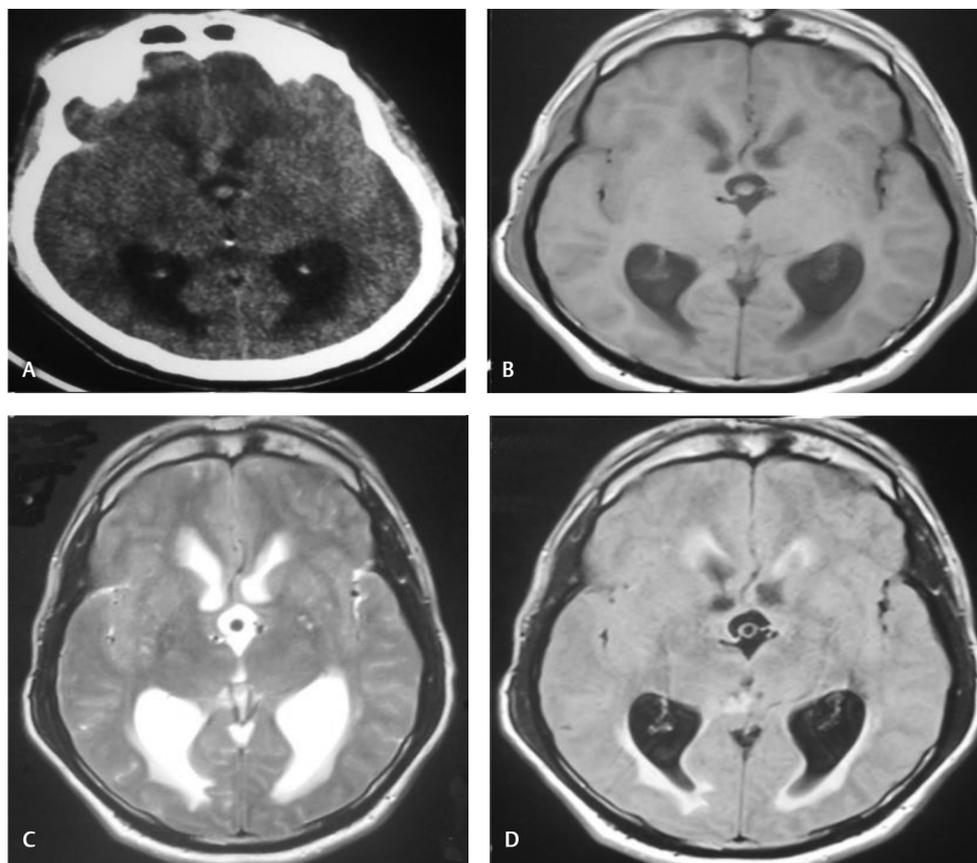
BDE refers to an anatomical variant characterized by dilatation and elongation of the BA. Knowledge regarding the natural history and progression of this rare entity is limited and progressive stroke is common cause of morbidity and mortality depending upon severity of condition at diagnosis.<sup>1,2</sup> The incidence ranges from 0.06 to 5.8% with a prevalence of 3.1 to 4.4%.<sup>1,3,4</sup> Males are more commonly affected and they become symptomatic mostly in the sixth to eighth decade.

DOI <https://doi.org/10.1055/s-0039-3402536>  
 ISSN 2277-954X.

© 2020. Neurological Surgeons' Society of India.

This is an open access article published by Thieme under the terms of the Creative Commons Attribution-NonDerivative-NonCommercial-License, permitting copying and reproduction so long as the original work is given appropriate credit. Contents may not be used for commercial purposes, or adapted, remixed, transformed or built upon. (<https://creativecommons.org/licenses/by-nc-nd/4.0/>)

Thieme Medical and Scientific Publishers Pvt. Ltd., A-12, 2nd Floor, Sector 2, Noida-201301 UP, India

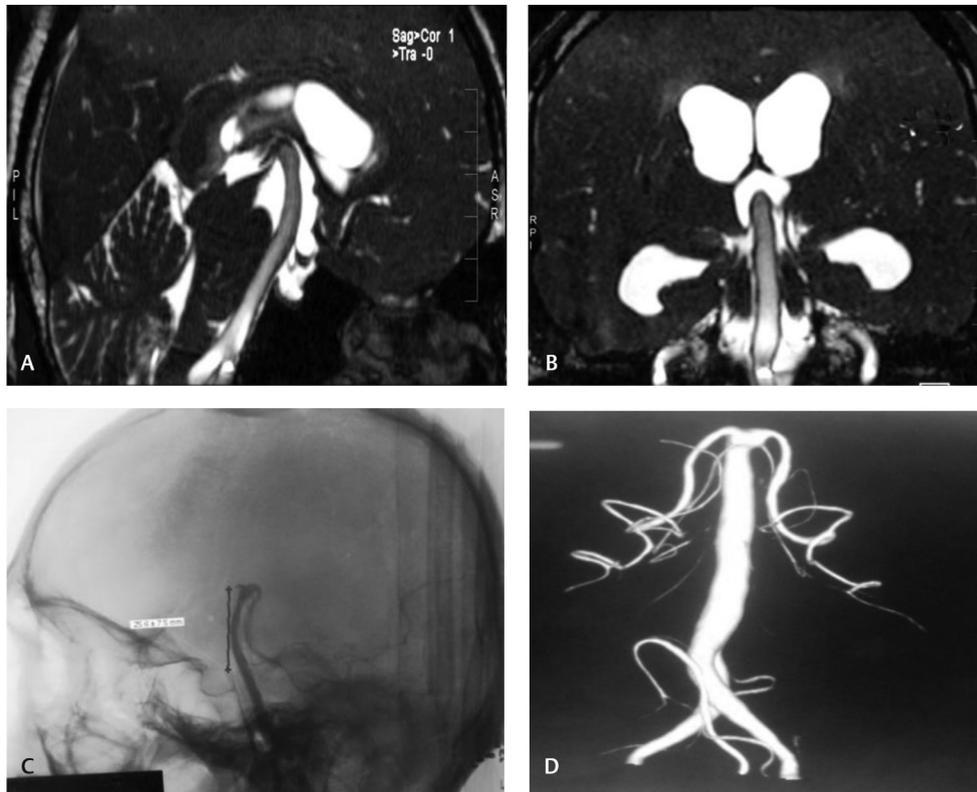


**Fig. 1** (A) CT brain axial images reveal hyperdense basilar artery in the floor of third ventricle appears as bull's eye or target sign. (B–D) Axial T1-weighted MR images reveal iso- to hyperintense signal in this lesion with signal void on T2 and FLAIR images. On T2-weighted MR images, this appearance may mimic a “target sign” with central flow void surrounded by hyperintense CSF signal. CSF, cerebrospinal fluid; CT, computed tomography; FLAIR, fluid-attenuated inversion recovery; MR, magnetic resonance.

The clinical manifestation can vary from a benign asymptomatic course to an extremely debilitating condition. When symptomatic, an ischemic pontine infarct is the most common presentation of BDE (20–30%). Other modes of presentation include intracerebral hemorrhage, cranial nerve palsies, obstructive hydrocephalus, subarachnoid hemorrhage, trigeminal neuralgia, and hemifacial spasm.<sup>1,4</sup> The etiopathogenesis of ischemic stroke in BDE is related to the stretching and distortion of branches of BA leading to stasis and reduction in forward flow in a dilated artery and superimposed atherosclerosis resulting in thromboembolic phenomenon.<sup>5,6</sup> In addition, some recent studies have shown an association between small vessel disease and BDE which could be another causative factor for lacunar infarcts.<sup>1</sup> Cranial nerve palsies and neuralgia probably result from direct compression of tortuous and elongated vessel.<sup>7</sup> The ectatic vessels are often associated with aneurysmal dilatation which may present with SAH or bleed.<sup>6,8</sup> Hydrocephalus is one of the rare presentation of BDE and may be communicating (postsubarachnoid hemorrhage) or noncommunicating obstructive hydrocephalus.<sup>9–14</sup> BDE can cause obstruction at cerebral aqueduct, midbrain, third ventricle, or as high as the foramen of Monro. Noncommunicating hydrocephalus or normal pressure hydrocephalus could be due to water hammer effect of BA on the foramen of Monro or third ventricle.<sup>12</sup>

CT scan will reveal the hydrocephalus and a hyperdense lesion in the prepontine cistern which may be confused as a mass lesion as in our case. CT angiogram or DSA helps in visualizing the exact morphology and to rule out aneurysmal dilatations. MRI further delineates the anatomy and also helps to visualize the presence of small infarcts, and quantify the mechanical compression of cranial nerves if any. BA with flow void in the middle-third floor of third ventricle before bifurcation has typical appearance like a bull's eye. Target sign of third ventricle as we wish to name, it is a new finding which helps to exclude mass lesion or aneurysm in floor of third ventricle. It has not been described in literature so far.

BDE is defined as a BA diameter greater than 4.5 mm or deviation of any portion of the BA more than 10 mm from the shortest expected course or as a BA of more than 29.5 mm in length.<sup>7</sup> Level of BA is variable and had been graded by Smoker et al<sup>15</sup> into the following grades: grade 1 (bifurcation within the supra sellar cistern), grade 2 (bifurcation at the level of the third ventricle floor), and grade 3 (bifurcation indenting the third ventricle floor). Similarly, the BA lateral displacement too has been graded as follows: grade 1 (BA located in the medial to lateral margin of the clivus or the dorsum sellae), grade 2 (BA located lateral to those landmarks), and grade 3 (BA located in the cerebellopontine angle). Our patient had grade-III bifurcation which is very rare and grade-I displacement. Our patient had obstruction



**Fig. 2** (A, B) MRI sagittal and coronal images showing dolichoectatic basilar artery elevating and compressing the floor of the third ventricle. (C, D) DSA confirms the presence of a dolichoectatic basilar artery reaching 25 mm above the level of posterior clinoid. Maximum diameter was 6.5 mm. No aneurysms were seen. However, there was stagnation of flow through the basilar artery. DSA, digital subtraction angiogram; MRI, magnetic resonance imaging.

**Table 1** Case reports of symptomatic hydrocephalus due to BDE refused surgery

No.	Name	Year of publication	Age (y)/sex	Management
1	Breig et al <sup>16</sup>	1967	3 cases, no description	No details
2	Ekbom et al <sup>11</sup>	1969	6 cases, no data	No details
3	Rozario et al <sup>17</sup>	1978	57/M	Unilateral VP shunt
4	Healy et al <sup>18</sup>	1988	50/M	Unilateral VP shunt
5	Braneo et al <sup>19</sup>	1993	58/M	No details
6	Aiba T et al <sup>9</sup>	1995	72/F	Unilateral VP shunt
7	Ricci et al <sup>13</sup>	2000	55/M	Bilateral VP shunt
8	Weber et al <sup>20</sup>	2002	69/M	Bilateral VP shunt
9	Thiex et al <sup>21</sup>	2006	54/M	Unilateral VP shunt
10	Siddiqui et al <sup>14</sup>	2008	71/F	Unilateral VP shunt
11	Kausal et al <sup>22</sup>	2011	60/M	Unilateral VP shunt
12	Seshadri et al <sup>23</sup>	2012	45/M	Unilateral VP shunt
13	Celik et al <sup>24</sup>	2013	47/M	Unilateral VP shunt + endoscopic exploration
14	Zisimopoulou et al <sup>25</sup>	2015	48/M	Refused Sx
15	Ebrahinzadeh et al <sup>26</sup>	2016	68/M	Endoscopic septostomy and Unilateral VP shunt
16	Present case	2018	38/M	Unilateral VP shunt

Abbreviations: F, female; M, male; VP, ventriculoperitoneal.

to cerebrospinal fluid (CSF) flow at midpart of the third ventricle, which is an extremely rare site for obstruction and ours study is probably the second such case reported in literature. A definite etiology for the lower motor neuron (LMN) facial

palsy could not be explained, and we presume it to be caused by microemboli with minor brain stem stroke or due to the direct compression of the ectatic vessel or stretching on the nuclear or cisternal portion of the seventh cranial nerve.

Till date, only 15 cases (►Table 1) have been reported with symptomatic hydrocephalus as presentation of BDE. Our case is unique showing multiple presentations (hydrocephalus, microbleed/facial nerve palsy) in single patient with BDE. Symptomatic patients with hydrocephalus need emergency CSF diversion procedure either VP shunt or endoscopic septostomy with shunt according to the level of obstruction. Till date, no other permanent procedure for hydrocephalus due to BDE has been published endoscopic third ventriculostomy can be dangerous in situation like ours where the BA bifurcation is at the mid-third of the floor of the third ventricle and close to the foramen of Monro. Endovascular procedures still not established as treatment due to longer curvature and course. Role of antiplatelets and antiaggregants are in use but still debatable. Microvascular decompression can be helpful in neuralgia and cranial nerve palsy but need further endorsement with more studies. Randomized trials comparing efficacy of different treatment are lacking and needed urgently.

## Conclusion

BDE is a rare anatomical condition which can be having various presentations of which obstructive hydrocephalus necessitate immediate surgical intervention. Target sign of the third ventricle is a useful radiological sign, which helps us to give diagnosis of BDE. VP shunt is preferred over endoscopic third ventriculostomy to avoid injuring the ectatic and tortuous BA. Endoscopic septostomy, followed by VP shunt, is also another surgical option. Definitive treatment for hydrocephalus has not established but needed further randomized trials.

### Funding

None.

### Conflict of Interest

None declared.

## References

- 1 Passero SG, Rossi S, Giannini F. Natural history of vertebrobasilar dolichoectasia. *Neurology* 2008;70(1):66–72
- 2 Mangrum WI, Huston J III, Link MJ, et al. Enlarging vertebrobasilar nonsaccular intracranial aneurysms: frequency, predictors, and clinical outcome of growth. *J Neurosurg* 2005;102(1):72–79
- 3 Nakamura Y, Hirayama T, Ikeda K. Clinicoradiologic features of vertebrobasilar dolichoectasia in stroke patients. *J Stroke Cerebrovasc Dis* 2012;21(1):5–10
- 4 Yu YL, Moseley IF, Pullicino P, McDonald WI. The clinical picture of ectasia of the intracerebral arteries. *J Neurol Neurosurg Psychiatry* 1982;45(1):29–36

- 5 Kumral E, Kisabay A, Ataç C, Kaya C, Calli C. The mechanism of ischemic stroke in patients with dolichoectatic basilar artery. *Eur J Neurol* 2005;12(6):437–444
- 6 Steel JG, Thomas HA, Strollo PJ. Fusiform basilar aneurysm as a cause of embolic stroke. *Stroke* 1982;13(5):712–716
- 7 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(1):22–26
- 8 Echiverri HC, Rubino FA, Gupta SR, Gujrati M. Fusiform aneurysm of the vertebrobasilar arterial system. *Stroke* 1989;20(12):1741–1747
- 9 Aiba T, Nakazawa T. Non-communicating hydrocephalus due to megadolichobasilar artery—case report. *Neurol Med Chir (Tokyo)* 1995;35(2):104–106
- 10 Branco G, Goulão A, Ferro JM. MRI in aqueduct compression and obstructive hydrocephalus due to an ectatic basilar artery. *Neuroradiology* 1993;35(6):447–448
- 11 Ekblom K, Greitz T, Kugelberg E. Hydrocephalus due to ectasia of the basilar artery. *J Neurol Sci* 1969;8(3):465–477
- 12 Marinescu M, Remy A, Dufour H, Derome P, Cambon H. [A peculiar mechanism of hydrocephalus: the “water-hammering” effect]. *Neurochirurgie* 1998;44(2):117–120
- 13 Ricci G, Lenzi J, Esposito V. Hydrocephalus caused by dolichoectatic basilar artery. Case report. *J Neurosurg Sci* 2000;44(3):155–158
- 14 Siddiqui A, Chew NS, Miszkil K. Vertebrobasilar dolichoectasia: a rare cause of obstructive hydrocephalus: case report. *Br J Radiol* 2008;81(964):e123–e126
- 15 Smoker WRK, 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. *Am J Neuroradiol* 1986;7(1):61–72
- 16 Breig A, Ekblom K, Greitz T, Kugelberg E. Hydrocephalus due to elongated basilar artery. A new clinicoradiological syndrome. *Lancet* 1967;1:874–875
- 17 Rozario RA, Levine HL, Scott RM. Obstructive hydrocephalus secondary to an ectatic basilar artery surgical Neurology 1978;9(1):31–34
- 18 Healy JF, Well MV, HYPERLINK “https://www.ncbi.nlm.nih.gov/pubmed/?term=Rosenkrantz%20H%5BAuthor%5D&cauthor=true&cauthor\_uid=6976881” Rosenkrantz H. Computed tomography demonstration of enlarged ectatic basilar artery associated with obstruction of anterior third ventricle compression *Radiology* 1981;5(4):239–245
- 19 Branco G, Ginlao A, Ferro JM. MRI in aqueduct compression and obstructive HCP due to ectatic basilar artery. *Neuroradiology* 1993;35(6):447–448
- 20 Weber MA, Steiner T, Fiebich J. Ungewöhnliche Ursache einer akuten Bewusstseinsintrübung. *Radiologe* 2002;42:833–836
- 21 Thiex R, Mull M. Basilar megadolicho trunk causing obstructive hydrocephalus at the foramina of Monro. *Surgical Neurology* 2006;65(2):199–201
- 22 Kansal R, Mahore A, Dange N, Kukreja S. Dolichoectasia of vertebrobasilar arteries as a cause of hydrocephalus. *Journal of Neurosciences in Rural Practice* 2011;2(1):62–64

- 23 Seshadri R, Sadashiva N, Shukla D, Saini J, Pandey, P. Vertebrobasilar dolichoectasia presenting as symptomatic obstructive hydrocephalus: A case report with review of literature. *IJNS* 2012;1(2):165–168
- 24 Çelik Ö, Berkman ZM, Orakdöğen M, Ayan E, Somay H, Düzkalir HA. Obstructive hydrocephalus due to vertebrobasilar dolichoectasia: diagnostic and therapeutic considerations. *J Neurol Surg A Cent Eur Neurosurg.* 2013;74 Suppl 1: e4–e8
- 25 Zisimopoulou V, Ntouniadaki A, Aggelidakis P, Siatouni A, Gatzonis S, Tavernarakis A. Vertebrobasilar Dolichoectasia Induced Hydrocephalus: the Water-Hammer Effect. *Clin Pract* 2015;5(2):749
- 26 Ebrahimzadeh K, Bakhtevvari MH, Shafizad M, Rezaei O. Hydrocephalus as a rare complication of vertebrobasilar dolichoectasia: a case report and review of literature. *Surgical Neurology International* 2017;8:256