

Vertebrobasilar Dolichoectasia: Clinical Neuroimaging Correlation

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ABSTRACT:

BACKGROUND:

Vertebro-basilar dolichoectasia (VBD) is an uncommon vasculopathy of unclear origin affecting the arterial wall of vertebral artery (VA) and basilar artery (BA). A variety of clinical syndromes and neuroimaging features have been associated with VBD.

OBJECTIVE:

This cross sectional study is conducted to shed a light on the neuroimaging signs of VBD and explore the association between the clinical presentation and imaging parameters of VBD in patients clinically suspected to have VBD with neuroimaging diagnosis.

MATERIAL AND METHODS:

This randomized cross sectional study was performed in Middle Euphrates Neuroscience Centre in Annajaf city between April 2012 to December 2012, A 34 patients with neuroimaging diagnosis of VBD (18 patients by MRI and 16 patients by MDCT) were included, VBD were assessed by studying the basilar artery width (BAW), bending length (BL) and level of basilar artery bifurcation according to Dan Deng et al criteria. The findings were correlated with patient clinical presentation.

RESULTS:

Thirty four patients, (24 males & 10 females) with age range between 25-81 years and mean of 57.9 year were included in this study. Among all presenting symptoms, only headache showed statistically significant association (p value <0.05) with the width of the basilar artery. There was higher prevalence of single cranial nerve involvement (SCNI) in patients with more tortuous basilar artery (55.5% of those with BL more or equal to 10 mm versus 16% of those less than 10 mm) with statistically significant association (p value <0.05), suprasellar extension was seen in 26.4% of cases and showed significant association with headache and visual symptoms.

CONCLUSION:

VBD is an important clinical entity and could be associated with or responsible for many serious clinical presentations, special attention is needed for VBD diagnosis and its parameter during evaluation of brain imaging especially in patients with un-explained or vague presentation.

KEYWORDS: vertebrobasilar, dolichoectasia.

INTRODUCTION:

As we know vertebrobasilar circulation includes two vertebral arteries VA, the basilar artery (which is formed by the union of the two vertebral arteries), and their branches and supplies the

medulla, pons, mesencephalon, and cerebellum. The two vertebral arteries join at the caudal border of the pons to form the single basilar artery⁽¹⁾.

The term "dolichoectasia" derives from the Greek words: dolichos, meaning elongation, and ectasia, meaning dilatation. A megadolichobasilar anomaly and giant fusiform aneurysm, or giant serpentine aneurysm, or the "arteriaesigmoïdes" and "dilatative" arteriopathy are other terms used to define highly tortuous dilated arteries, which are primarily found in the vertebrobasilar system⁽¹⁻²⁾. Dilatation and elongation of the vertebrobasilar arterial system has long been a recognized clinical

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entity⁽³⁾. The incidence of VBD ranges from 0.06 to 5.8% in the general population⁽⁴⁾. VBD parameters BL (basilar artery length) and BAW (basilar artery width) were classified according to Dan Deng et al criteria⁽⁵⁾. The height of the BA bifurcation and the position of the BA were classified according to Smoker et al.⁽⁶⁾. The clinical presentations of VBD are varied and is usually attributable to the mass effect of the dilated vessels and possibly to associated flow turbulence and can include either isolated or combined symptoms of the brainstem/cranial nerve dysfunction, cervicomedullary junction compression motor deficits, cerebellar dysfunction or central sleep apnea⁽⁷⁾, or ischemic stroke. Single Cranial nerve palsies (SCNI)⁽⁸⁻¹⁰⁾, hydrocephalus, trigeminal neuralgia⁽¹¹⁾, hemifacial spasm⁽¹²⁾, nystagmus, chiasmal compression, ataxia, and dementia all have been reported an association with VBD⁽¹³⁾. Acquired and congenital causes have been recognized as an etiological elements in different studies^(4, 8, 14). In 2008, Tiltic M et al, not only considered VBD as a stroke risk factor, but also as a cause of compression on nerves surrounding brainstem and turbulence-related tinnitus & vertigo⁽¹⁵⁾. In 2010, Clinical- radiologic analyses done by Nakamura Y et al showed increased prevalence of pontine infarcts in patients with VBD⁽¹⁶⁾, while hemi facial spasm and ipsilateral trigeminal neuralgia were reported as a result of facial and trigeminal nerves compression by VBD⁽¹⁷⁾.

VBD is not readily recognized by neurologists and radiologists as a result of normally wide variation in the caliber and tortuosity of the VA and BA seen in healthy individuals. This study has two folded aims, firstly to shed a light on the radiological signs of this vasculopathy and secondly to explore the association between the clinical presentation and various imaging parameters of VBD in patients clinically suspected and radiologically proved to have VBD.

MATERIAL AND METHOD:

This cross sectional observational study was carried out between April 2012 and December 2012 in the Middle Euphrates Neuroscience Centre, Al-Najaf, Iraq. Thirty four patients were submitted to evaluation by this study (24 males and 10 females). All patients had complete neurological evaluation with radiological diagnosis of VBD.

VBD parameters BL and BAW were classified according to Dan Deng et al criteria⁽⁵⁾. The height of the BA bifurcation and the position of the BA were classified according to Smoker et al.⁽⁶⁾. The ectasia was diagnosed when male patient has BA diameter larger than 4.2 mm, a left VA larger than 3.7 mm, or a right VA larger than 3.5 mm while female patient needs to exhibit a BA diameter of larger than 4.0 mm, a LVA larger than 3.4 mm, or a RVA larger than 3.1 mm (figure 1).

Tortuosity and elongation can be established when BA length is greater than 29.5 mm (figure 2) or bending length (equal to the distance between most lateral portion of BA perpendicular to a straight line joining the BA origin to its bifurcation on magnetic resonance angiogram(MRA) exceeds 10 mm.

Because of elongation, BA tends to bifurcate higher than normal. The height of the BA bifurcation was classified according to Smoker et al into 4 grades⁽⁶⁾, which include:

- Grade 0, the bifurcation is at or below the dorsum sellae.
- Grade 1, the bifurcation is within the suprasellar cistern.

Grade 2, the bifurcation is at the level of the third-ventricle floor.

Grade 3, the bifurcation is indenting and elevating the third-ventricle floor. of the total 34 patient, 18 were studied by magnetic resonance imaging (MRI) using 1.5 Tesla whole body magnet scanners (PHILIPS ACHIEVA 2010) with 3-dimensional (3D) images and field of view (FOV) of 200 mm enhanced by maximum intensity projection (MIP) techniques for intracranial MR-arteriography (MRA). MRI in the axial, coronal, and sagittal planes were obtained using standard spin echo longitudinal relaxation time-weighted sequence (T1), turbo spin (TS) transverse relaxation time-weighted sequence (T2), and fluid-attenuated inversion recovery (FLAIR) techniques, without intravenous gadolinium. Arterial dimensions were subsequently measured directly on the workstation monitor using measurement tools. The remaining 16 patients were studied by multidetector computerized tomography (MDCT), 64 slices, (Philips Brilliance 2010), 15 without and one with intravenous iodinated contrast material (iohexol).

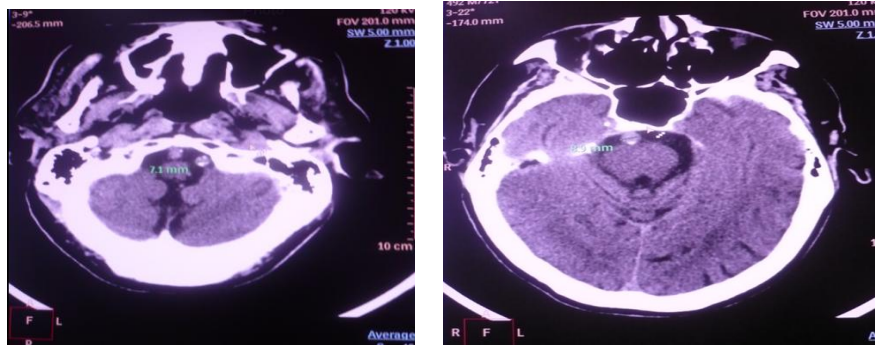


Figure 1: Axial images of non-enhanced computerized tomography scan of the brain, showing gross dilation (8.9 mm) of basilar (A) and vertebral (B) arteries(7.1mm).

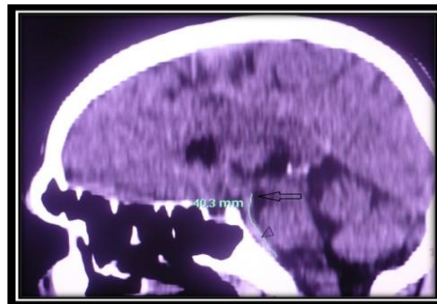


Figure 2: A multidetector computerized tomographic sagittal image of brain in 42 years old patient showing elongated basilar artery (arrow head) measuring 40.3 mm and reaching suprasellar cistern (long arrow).

Patient Selection criteria was included, if he/she was aged 18 years or older (on the date of the radiological examination) and had a radiologic signs of dolichoectasia affecting the vertebrobasilar system. Patients were referred by expert neurologists, where the clinical pictures were highly suggestive of VBD, while patients referred to MRI and CT units for another cause but found to have radiological signs of VBD, they had to be seen retrospectively by the neurologist to evaluate their clinical symptoms. Patients with intracranial pathology other than VBD and trauma cases were excluded from the study except those with recent ischemic changes exclusively involving the posterior circulation territories or those with mild to moderate physiological cerebral atrophy associated with aging process.

Measurements of basilar artery

Vertebral artery (VA) dominance and bending were evaluated firstly. VA was determined as right or left sided or equivalent. A line was drawn between

origin of the basilar artery to its bifurcation for reference to decide the side of BA bending (figures 3 A and B).

When there is no union of vertebral arteries, in a case such as aplasia of basilar artery, side of bending was based on an imaginary line. If the basilar artery ran in an S-shape over the standard line, a decision had to be made according to which side bending was dominant.

Bending of BA (tortuosity) was decided as right or left sided or no bending. A straight line was drawn between points of union to the point of bifurcation of the basilar artery and considered as standard line. Another horizontal line was then drawn vertical to the standard line from the point of the greatest bending of the basilar artery; the length of this line was defined as bending length (BL). For statistical requirement of this study, measurement of BA diameter was done at level of midpons, determined by anatomical landmarks of the fourth ventricle and middle cerebellar peduncles levels (figures 4, 5).

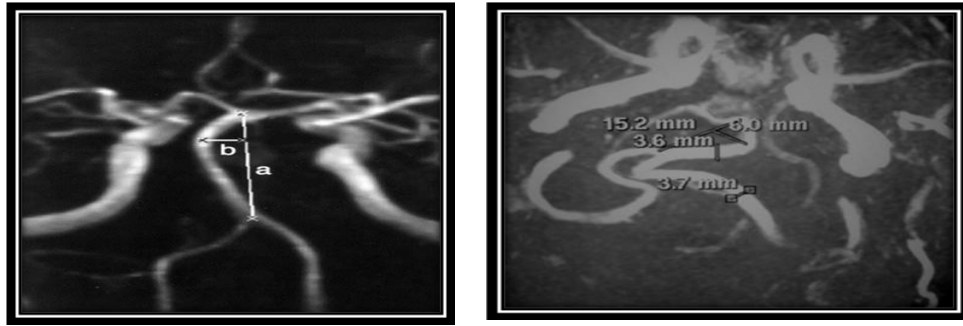


Figure 3: Images of 3-dimensional time-of-flight (TOF) magnetic resonance arteriography (MRA) of two different patients (A) and (B) showing measurement of bending length (b) perpendicularly from midpoint of basilar artery width to the standard line (a).

To confirm diagnosis of VBD, measurement of BA length (from its origin to its bifurcation) was performed using freehand calipers for tortuous course of the artery directly on 3-dimensional TOF MRA and indirectly on reconstructed images in usual MRI images. For CT scans, sectional modification of axial and coronal sections was created to obtain full length of BA curvature (figure 6). To evaluate the relationship between radiological signs of dolichoectatic patients and

their clinical symptoms, we divided the measurements of each parameter (BAW, BL) into two groups and studied the prevalence of clinical symptoms in each group separately. The relationship between the different VBD parameters (BAW, BL and suprasellar extension) and clinical presentations (headache, dizziness, ataxia, tinnitus, cranial nerve involvement, brainstem ischemia symptoms) was statistically analyzed using chi-square and Fischer exact test. The association was considered significant if the p-value was less than 0.05.

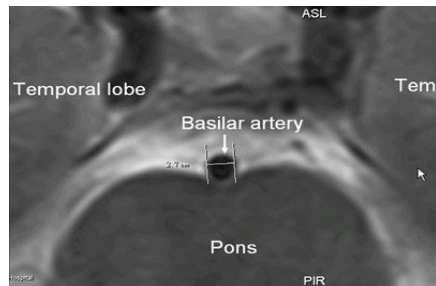


Figure 4: An axial magnetic resonance (MR) image at midpons level showing the measurement of basilar artery width (BW).



Figure 5: An axial computerized tomographic image of the brain in a female showing the ectatic basilar artery (arrow) measuring 5.2 mm at mid-pons level where fourth ventricle (arrowhead) and middle cerebellar peduncle (star).

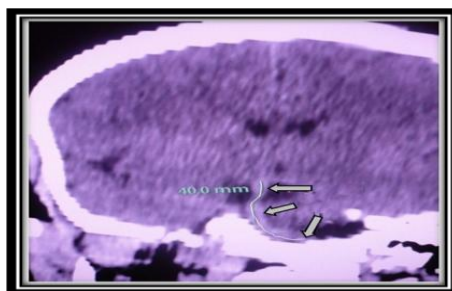


Figure 6: A coronal brain computerized tomographic (CT) image, reformatted along the tortuous course of basilar artery (arrows) to measure the full length of basilar artery using freehand calipers (here it is 40 mm long).

RESULTS:

- Age and gender distribution. VBD was seen more in males than females (24 versus 10, with a ratio of 2.4:1). Patient's age ranged between 25-81 years with mean age of 57.9 year.
- Tortuosity and dominance. Twenty cases of the 34 had BA tortuosity on the right (58%) and 14 cases on the left (42%). VA dominance on the left was found in 20 cases (58%) and on right 12 case (35%), 2 cases (5.8%) were equivalent. The curvature of the basilar

artery (tortuosity) was in opposite direction of the dominance of VA.

- Dolichoectasia parameters in relation to symptoms.

1) Basilar artery width

There was a statistically significant association (p value <0.05) between BAW and headache while no significant association was found with other symptoms as shown in table 1, those with BAW ranging from 4-4.9 mm and those with BAW of ≥5 mm.

Table 1: Clinical presentations according to the BAW.

BAW (mm) N=34	Vertigo of unknown origin	Central vertigo with ataxia (BSI)	Dizziness	Headache	SCNI (II, III, V, VII)	Tinnitus
4-4.9 mm N=18	3(16.6%)	0	3 (16.6%)	1 (0.05%)	7(38.7%)	3(16.6%)
≥ 5mm N=16	3(18.7%)	3(18.7%)	1 (0.06%)	7 (43%)	2(12%)	5(31.3%)
p-value	0.874	0.093	0.604	0.01	0.125	0.428

BSI=Brainstem ischemia. SCNI =single cranial nerve involvement (CNII, CNIII, CNV, CNVII)

2) Bending length (BL).

Statistically, there was a significant association (P-value < 0.05) between BL and only SCNI in the dolichoectatic patients, otherwise no

significant association was seen with other clinical symptoms in relation to BL as shown in table 1, those with BL<10 mm and those with BL equal or > 10 mm.

Table 2: Prevalence of clinical symptoms in relation to BL.

Basilar length (BL)(mm) N=34	Vertigo of unknown origin	Central vertigo with ataxia	Dizziness	Headache	SCNI (II,III,V, VII)	Tinnitus
<10mm N=25	5(20%)	1(4%)	4(16%)	5(20%)	4(16%)	7(28%)
≥10mm N=9	1(11%)	2(22%)	0	3(33.3%)	5(55.5%)	1(11%)
p-value	1	0.164	0.553	0.648	0.034	0.403

BSI=Brainstem ischemia. SCNI =single cranial nerve involvement (CNII, CNIII, CNV, CNVII).

3) Suprasellar extension.

Of the total 34 cases, 9 (26.4%) had a bifurcation of basilar artery at suprasellar cistern (grade 1), while the rest (73.6%) were grade 0. The prevalence of clinical symptoms in those 9 patients with suprasellar extension is shown in table 3. All three cases of visual involvement showed suprasellar

bifurcation of the basilar artery and 5 out of 8 cases of headache (62 %) had this bifurcation. Statistically significant association (p value <0.05) was seen between presence of suprasellar extension on one hand and headache and visual symptoms on other hand (table 3).

Table 3: Clinical features in relation to suprasellar BA extension.

Basilar artery extension N=34	Vertigo of U.O	Central vertigo with ataxia (BSI)	Headache	Visual involvement	Tinnitus	SCNI (II,III,V, VII)	Dizziness
suprasellar extension N=9	1 (11%)	2 (22%)	5 (55.5%)	3 (33.3%)	1 (11%)	1 (11%)	1 (11%)
No suprasellar extension N=25	5 (20%)	1 (4%)	3 (12%)	0	7 (28)	8 (32%)	3 (12%)
p-value	1	0.164	0.017	0.014	0.403	0.386	1

BSI =brainstem ischemia, U.O=unknown etiology

DISCUSSION:

In this observational study, we found that BA tortuosity is right-sided (58%) more frequently than left-sided (42%), and the BA curvature was in opposite side to the dominant VA in 85.2%. This finding was comparable to Hong et al⁽¹⁸⁾ result (83.5%). Hong et al attributed this directional relationship to the unique origin of BA from the junction of the two vertebral arteries, unlike most systemic arteries as BA is the only large artery in which two arterial flows merge. In addition, in angiographic and post-mortem studies, the left VA is often larger than the right VA while diameters of the VAs are of equal size in only 6–26% of patients⁽⁸⁾. Therefore, it could be postulated that the unequally mechanical forces resulting from asymmetric VA flow might lead to a morphological

deformation in the vertebrobasilar arterial system (lateral displacement or elongation of the BA, and hypoplasia of the VA). These deformations might asymmetrically induce the development of infarcts in the areas before or after the vertebrobasilar junction⁽¹⁸⁾.

VBD may be asymptomatic or symptomatic depending on the degree of ectasia and elongation of the BA and VA. The symptomatic manifestations include posterior circulation infarcts, compression of cranial nerves or brainstem⁽¹⁹⁾. In our study, all patients were symptomatic with the reported symptoms of vertigo, tinnitus, headache, dizziness, Brain stem ischemia (BSI as central vertigo, ataxia, dysarthria, dysphagia, and weakness), and SCNI (Otic nerve, Oculomotor, Trigeminal or and Facial nerves symptoms).

The significant association between the BAW and headache in dolichoectasia patients in this study may be a result of the pathological changes of brainstem due to the VBD-induced chronic compression or ischemia. Although other symptoms showed no statistically significant association with BAW, all BSI (three cases only of total patients), were seen in patients with BAW >7 mm which might give an impression that gross dilatation of basilar artery diameter can cause compression and possibly ischemia to the brainstem.

Basilar length BL gives an idea about the arterial deviation from the midline (curvature of the artery), so more the distance (large BL values) more the tortuosity of the artery. In this study, there was significant association only with SCNI, which might be explained on the base that according to the scoring of lateral displacement of the BA by Smoker et al ⁽⁶⁾, in which, grade III BA lays in the cerebellopontine (CP) angle cistern, the more BL value, the more BA liable to fall in grade 3 hence CP involvement, where compression of the nerve roots is a possibility producing clinical symptoms like trigeminal neuralgia. Direct compression by VBD was reported to be an uncommon cause for trigeminal neuralgia with estimated incidence ranging from 0.9% to 5.7% ⁽²⁰⁾. Vascular compression usually occurs at or near the root entry zone (REZ) of the trigeminal nerve ^(21, 22). Hamlyn ⁽²¹⁾ observed that 42 out of 46 patients who underwent posterior fossa surgery for treatment of trigeminal neuralgia had a vessel in contact with the nerve. Of those, 28 had a vessel in contact at the REZ, 12 had a vessel in contact lateral to the REZ, and 2 had a vessel in contact both at the REZ as well as lateral to it. Sindou et al ⁽²²⁾ observed the presence of a contacting vessel in 97% of 579 patients with idiopathic trigeminal neuralgia.

Similarly, unilateral facial nerve involvement in our patients can be attributed to the compression of the facial nerve at the REZ in the CP region which was considered to be responsible for the unilateral hemifacial spasm ^(23, 24).

Regarding level of BA bifurcation, our results of only 26.4 % of VBD patients having grade 1 (suprasellar extension) and remaining (73.6%) in grade 0, were different from that of Dan Deng et al study ⁽⁵⁾ where majority (69 %) had BA bifurcation within the suprasellar cistern (grade 1), the small sample in our might be responsible for this difference.

The significant association between headache and suprasellar extension of BA observed in our study could be consequent to the inconstant indentation of the third ventricle that causes intermittent episode of hydrocephalus presented clinically as attack of severe headache. However, no patient showed hydrocephalus at time of imaging in our study, possibly due to its intermittent inconstant nature.

The association of BA ectasia with hydrocephalus was reported by several authors and was attributed to impairment of outward CSF flow from the foramen of Monro, caused by the dilated artery pulsating against the third ventricle, producing a "water-hammer" effect or obstructive hydrocephalus ^(25, 26). Furthermore, there was also a significant association between visual disturbance and suprasellar extension of BA that is possibly secondary to the direct indentation of optic tracts or to the presumed intermittent hydrocephalus and increase in intracranial pressure may influence indirectly on optic nerves, that is supported by association of visual involvement with intermittent severe headache.

CONCLUSION:

When clinical features of VBD are considered, the increment in the basilar artery diameter was more related to headache; while the more tortuous the artery associated with higher prevalence of cranial nerves involvement. Furthermore, patients with VBD presented with visual involvement and headache are tending to have suprasellar extension of BA. Special attention needs to be paid by radiologists for important parameters of VBD during evaluation of brain imaging, particularly in patients with un-explained or vague presentation.

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