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Symptomatic obstructive hydrocephalus due to cerebrospinal fluid pathway obstruction from vertebrobasilar dolichoectasia: Case report and review of the literature

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***Abstract*---**Vertebrobasilar Dolichoectasia (VBD) is a rare vascular anomaly in which the vertebral/basilar artery is elongated, swollen, and tortuous. Symptoms can be ischemic or obstructive hydrocephalus although they are rarely found because it is usually asymptomatic. Therefore, it is necessary to establish a proper diagnosis to obtain appropriate management. A 62-year-old man had memory loss, imbalance walking, and urinary incontinence which then gradually decreased consciousness. Magnetic resonance imaging (MRI) revealed active hydrocephalus obstruction and suspicion of a fusiform type aneurysm in the basilar artery. Digital Subtraction Angiography (DSA) procedure was performed and the dolichoectasia of the basilar artery was obtained with non-communicant hydrocephalus which then improved after a Ventriculo-Peritoneal (VP) shunt was inserted. The information in this case report is essential for scholarly purposes and the participant gave written informed consent for publication.The gold standard imaging with DSA is needed to confirm the case of VBD. Information regarding the management of patients with VBD is scarce. Patients with manifestations of compression may undergo surgical evaluation with arterial repositioning or ventricular shunt placement. For patients with cerebrovascular complications, acute management should be based on the best care practices for patients with hemorrhagic or ischemic stroke. The use of

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anticoagulants is controversial and may increase the risk of hemorrhagic complications. Control of vascular risk factors and the use of antiplatelet and statin therapy are recommended.

***Keywords*---**Digital Subtraction Angiography, Obstructive Hydrocephalus, Vertebrobasilar Dolicoectasia

Introduction

Vertebrobasilar artery dolichoectasia/Vertebrobasilar Dolicoeectasia (VBD) is a rare vascular anatomical anomaly condition, characterized by elongated, swollen, and tortuous vertebral and basilar arteries accompanied by hemodynamic abnormalities in these arteries. Other known names include megadolichoectasia, fusiform aneurysm of the vertebrobasilar artery, and the tortuous vertebrobasilar system (Oishi et al., 2018; Prasad et al., 2021).

The prevalence of VBD is about 0.08-6.5% in the general population, whereas the prevalence increases by 3-17% in patients with stroke. The primary site of VBD was found in the basilar artery (51%), followed by the bilateral to basilar vertebral arteries (33%) and in both vertebral arteries (16%). Several known risk factors include hypertension (most often), old age, and male sex, it has also been reported in several studies that the incidence of VBD may be associated with congenital connective tissue disorders such as Marfan syndrome and Ehlers–Danlos syndrome (Conradie & Bonnet, 2021; Prasad et al., 2021).

Pathological findings of VBD represent an enlarged external diameter of the artery and thinning walls, with degeneration of the internal elastic lamina layer with multiple cell gaps, thinning of the tunica media due to reticular fiber deficiency, and smooth muscle atrophy. The etiology of these changes is not fully known. Some authors suspect long-standing systemic hypertension as one of the causes, in other studies suspect congenital vasculopathy as the cause (Del Brutto et al., 2017).

VBD is usually asymptomatic and less than 10% of patients have neurological symptoms. Symptoms that appear can be ischemic symptoms or obstructive hydrocephalus depending on the mechanism of disturbance in the surrounding structures. The diagnosis of VBD was started with the help of a Computed Tomography (CT) Scan in 1986 and then with Magnetic Resonance Imaging (MRI) in 1988. CT Scan is generally the first done although with low sensitivity to assess acute ischemia that can occur in VBD as well as sometimes have artifacts caused by bony structures around the brain, but the sensitivity to intracranial and extraction bleeding in the first 24 hours of onset is quite high up to more than 95%. The presence of widening of the vertebral arteries may be suspected as a dolicoectatic condition. On contrast administration, strong patent dolicoectatic vessels can be seen that sting the contrast. MRI examination is more sensitive to see small areas of ischemia in the cerebellum and brainstem, which are the predilection areas for blockage of the vertebrobasilar vessels. MRI angiography can detect occlusion and stenosis of blood vessels with high accuracy but is less

useful in determining the degree of stenosis. (Del Brutto et al., 2021; Prasad et al., 2021).

The average diameter of the basilar artery ranges from 1.5 to 4 mm. Some of the diagnostic criteria for VBD are (1) a blood vessel diameter of more than 4.5 mm along the vessel path with a vessel deviation of more than 10 mm from the initial position, (2) a basilar artery length of more than 29.5 mm and (3) a length of vertebral artery intracranial segment more than 23.5 mm (Lee et al., 2019).

With the advancement of medical development, currently, Digital Subtraction Angiography (DSA) examination is being carried out as the golden standard for establishing the diagnosis of VDB. The advantage of using DSA is that it can provide an overview of the shape of the dolichoic blood vessels originating from the aortic arch and the direction of these blood vessels to more distal branches, to provide a better picture of the morphology of the blood vessels and can see pathological conditions in the intraluminal more specifically (Brinjikji et al., 2017; Li et al., 2021).

VBD has often been associated with a poor prognosis and the outcome varies greatly from individual to individual. Life expectancy in the next 5 years is only around 54.2%, 39.5% at 10 years, and 23.5% at 15 years, and is better in asymptomatic patients. A study by Passero et al in a prospective study of 156 VBD patients showed a progressive increase in morbidity and mortality. Most VBD patients have cerebrovascular disorders as the most common cause of death (Umana et al., 2021; D. P. Zhang et al., 2020; X. Zhang et al., 2018).

In this case report, we report a patient with vertebrobasilar artery dolichoectasia who showed symptoms of hydrocephalus due to compression of the Sylvius aqueduct and experienced clinical improvement after insertion of a ventriculoperitoneal tube. Establishing the right diagnosis is one of the main modalities to get the appropriate treatment.

Method

The patients involved in this case report were patients who were hospitalized and underwent surgery at the Dr. Soetomo General Academic Hospital in Surabaya Indonesia. The patient underwent preoperative examination, investigation, and postoperative examination at the same hospital. Primary data was obtained from direct history taking to the patient and other secondary data came from medical records which contained information on supporting laboratories and supporting radiology. The patient has agreed and signed the informed consent that it will be used as an interesting case to be published for humanitarian purposes and the advancement of science. All patient identities are not disclosed in the publication to maintain patient privacy.

Discussion

Case Presentation

A 62-year-old man came to the polyclinic of Dr. Soetomo Regional General Hospital Surabaya with a history of symptoms of memory loss, imbalance while walking, and episodes of urinary incontinence over the previous 3 years. Complaints are felt to be getting worse with the emergence of a gradual decrease in consciousness. The patient is known to have a history of hypertension for the previous 2 years, controlled by the administration of amlodipine. On physical examination, vital signs were within normal limits and no additional abnormalities were found. Neurological examination in the patient was within normal limits with Glasgow Coma Scale (GCS) 456, cranial nerves were within normal limits, and there were no signs of limb weakness or impaired touch sensation.

Magnetic resonance imaging (MRI) was performed on the patient showing active obstructive hydrocephalus and suspicion of a fusiform aneurysm in the basilar artery. The axial T2 flair MRI sequence (Fig. 1, A) showed widening of the lateral and third ventricles, accompanied by periventricular edema, suggesting active hydrocephalus. The axial T2 MRI sequence (Figs 1, B) shows dilatation from the left vertebrobasilar artery to the level at the level of the 3rd ventricle. The dilated and tortuous basilar artery causes compression of the mesencephalon which results in obstruction of the Sylvius aqueduct causing obstructive hydrocephalus in this patient.

In this patient's case, dolichoectasia causes compression of the Cerebrospinal Fluid (CSF) flow from the 3rd ventricle so that the appearance of hydrocephalus that occurs is obstructive/non-communicating. The patient underwent an emergency VP shunt which resulted in decompression of the CSF in the ventricles and improvement of symptoms. On a non-contrast CT scan of the head, a postoperative evaluation of the VP shunt showed resolution of the hydrocephalus. Another finding that is seen is a fusiform dilatation that is easily visible from the basilar artery along with the elongation that occurs. The effects of compression on the brainstem are less obvious (figure 2).

Establishing a diagnosis through CT and MRI is not the golden standard for evaluating vascular abnormalities, so patients need to have a DSA procedure. The procedure was performed under local anesthesia and sterile procedures in the operating field, including puncture of the right common femoral artery and insertion of a 5F sheath. Cerebral angiography was performed with a Vert 5F catheter.

The findings on DSA are (1) Right Carotid Communis Artery injection: carotid bifurcation is normal; (2) Injection of Right Internal Communis Artery: Anterior Cerebral Artery, Middle Cerebral Artery and normal distal circulation;(3) Injection of Left Carotid Communis Artery: carotid bifurcation normal; (4) Injection of Left Internal Communis Artery: ACA, MCA, and distal circulation are normal; (5) Left Vertebral Artery injection: Injection in the Left Subclavian Artery, the catheter has difficulty accessing the Left Vertebral Artery. Basilar artery dolichoectasia is

seen; (6) Brachiocephalic injection into the Right Vertebral Artery: tortuous access to the RVA appears. The catheter cannot ascend to the RVA. The patient is stable, during and after the procedure no new neurologic deficits were found. It was concluded that there was basilar artery dolichoectasia. The DSA results can be seen in Figure 3. The shape of the VBD in this patient's case can be seen on the MRI and DSA which are shown in Figure 4.

In this case report, we report a patient with a rare case of obstructive hydrocephalus associated with vertebrobasilar dolichoectasia pressing on the aqueducts Sylvie. The image found on the head CT scan of the dilation of both lateral and 3rd ventricles indicates the level of blockage in this patient. The presence of a connection between the lateral and third ventricles indicates a patent foramen Monroe and the aqueducts Sylvie is considered as the site of obstruction. In this case, initially, symptoms appeared in the form of signs of Normal Pressure Hydrocephalus (NPH) in the form of memory loss, imbalance while walking, and episodes of urinary incontinence which were the result of basilar artery pulsation that interfered with CSF flow from the ventricles. 3. Unilateral VP Shunt installation in one of the lateral ventricles gives good results with reduced symptoms felt. Dolichoectasia cases are often associated with hypertension, advanced age, and male gender where all of these risk factors can be found in patients (Del Brutto et al., 2017; Wang et al., 2019).

From the histological and histochemical examination, it appears that in the case of VBD there is atherosclerotic degeneration of the vessel wall, loss of elastic fibers, and the presence of fibrosis. This condition is mostly associated with hypertension, which in this case was found in a patient who is believed to be responsible for the formation of VBD. As a result of high intraluminal pressure in cases of hypertension, elastic fibers are reduced. Another study states that hypertension causes ongoing stress on the walls of blood vessels so that they damage and cause the collagen and elastin connective tissue to stretch. The etiology of VDB is still unclear and is being debated, but several factors that have been mentioned are thought to play a role in the formation of VBD, including congenital, immune reactions and degenerative diseases such as hypertension and atherosclerotic plaque formation (Oishi et al., 2018; Wang et al., 2019).

Hydrocephalus is a rare complication of VBD. VBD itself is usually asymptomatic, less than 10% have symptoms of neurological deficits. The estimated risk of developing hydrocephalus in 5 years is only about 3.3%. In patients with symptomatic symptoms that appear quite typical of the NPH triad (urinary incontinence, dementia, and balance disorders when walking). In the literature, it is stated that this is due to the water hammer effect mechanism that can occur due to the development of VBD which has not yet given a mass effect, but pulsatile arteries that continue to the 3rd ventricle and lateral ventricle through the foramen of Monroe can interfere with the flow velocity of the CSF distally so that hydrocephalus appears due to CSF accumulation is proximal and the pressure tends not to be high/normal pressure. As the VBD progresses, the patient eventually develops a second complication which is quite rare, namely direct suppression of the CSF flow pathway due to mass effect. The location of the dilated basilar artery is also up to the floor level of the 3rd ventricle. By looking at the dilation of the ventricles in the proximal direction (lateral and third

ventricles), it can be identified that the level of blockage is in the aqueducts Sylvie, causing non-communicating/obstructive hydrocephalus. Increased Intracranial Pressure (ICP) due to CSF obstruction can be overcome in line with the reduction of symptoms after VP Shunt installation. In this case, there were no symptoms of thromboembolic ischemia (due to hemodynamic and hemostatic changes in tortuous blood vessels) or compression of the brainstem and cranial nerves which can also be a complication of VBD. Intracranial hemorrhage that can occur in VBD patients was not found in these patients (Ebrahimzadeh et al., 2018; Martinez-Nunez et al., 2021).

Information regarding the management of patients with VBD is still difficult to find. Patients presenting with compression may undergo surgical evaluation with either arterial repositioning or placement of a ventricular shunt. For patients with cerebrovascular complications, acute management should be based on best care practices for patients with hemorrhagic or ischemic stroke. Currently, the management of VBD is mostly conservative, although there is no evidence of a long-term prognosis of the efficacy of using oral antiplatelet or anticoagulants. In another study, it was stated that the use of antiplatelet therapy could worsen the condition by triggering the incidence of bleeding. The use of anticoagulants is controversial and may increase the risk of hemorrhagic complications (Zdravković et al., 2017).

This case report shows that even with advanced radiological imaging techniques, establishing the diagnosis of VBD remains a challenge in itself. Because clinically it is difficult to distinguish the incidence of NPH which often occurs in old age. Determination of the exact location of obstruction due to VBD is also obtained on a good radiological examination because it will affect the next treatment. The diagnostic criteria for VBD cannot be established with certainty through CT Scan or MRI imaging, therefore DSA needs to be done. This case shows the usefulness and role of DSA as a diagnostic tool and a determinant of further management (He et al., 2019)

Conclusion

Specific measures in as an effort to prevent acute exacerbations of dolichoectasia have not yet been found, therefore, in patients with VBD, prompt and appropriate monitoring and management are needed when early symptoms of complications such as hydrocephalus appear in this case. The prognosis of VBD patients who show symptoms is not so good. Several endovascular interventional treatments that have been carried out have not shown success.

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References

Brinjikji, W., Nasr, D. M., Flemming, K. D., Rouchaud, A., Cloft, H. J., Lanzino, G., & Kallmes, D. F. (2017). Clinical and imaging characteristics of diffuse intracranial dolichoectasia. *American Journal of Neuroradiology*, *38*(5), 915–922. https://doi.org/10.3174/ajnr.A5102

Conradie, J. M., & Bonnet, E. G. (2021). Dolichoectasia and Its Diagnostic Criteria: A Case Report and Literature Review. Dalam *Cureus* (Vol. 13, Nomor 1, hlm. e12516). https://doi.org/10.7759/cureus.12516

Del Brutto, V. J., Gutierrez, J., Goryawala, M. Z., Sacco, R. L., Rundek, T., & Romano, J. G. (2021). Prevalence and Clinical Correlates of Intracranial Dolichoectasia in Individuals With Ischemic Stroke. *Stroke*, *52*(7), 2311–2318. https://doi.org/10.1161/STROKEAHA.120.032225

Del Brutto, V. J., Ortiz, J. G., & Biller, J. (2017). Intracranial arterial dolichoectasia. *Frontiers in Neurology*, *8*(JUL), 12–14. https://doi.org/10.3389/fneur.2017.00344

Ebrahimzadeh, K., Bakhtevari, M. H., Shafizad1, M., & Rezaei, O. (2018). Hydrocephalus as a rare compilation of vertebrobasilar dolichoectasia: A case report and review of the literature. *Surgical Neurology International*, *9*(1). https://doi.org/10.4103/sni.sni

He, X., Duan, C., Zhang, J., Li, X., Zhang, X., & Li, Z. (2019). The safety and efficacy of using large woven stents to treat vertebrobasilar dolichoectasia. *Journal of Neurointerventional Surgery*, *11*(11), 1162–1166. https://doi.org/10.1136/neurintsurg-2019-014933

Lee, J. M., Park, J. S., & Koh, E. J. (2019). Severe vertebrobasilar dolichoectasia as a cause of obstructive hydrocephalus: A case report. *Medicine*, *98*(21), e15752. https://doi.org/10.1097/MD.0000000000015752

Li, B., Zhou, B., Zhang, M.-Z., Qin, R.-Q., & He, Y. (2021). Extensive intracranial arterial dolichoectasia involving distal branches of intracranial arteries: Two cases report and review of the literature. *The International Journal of Neuroscience*, *131*(11), 1133–1138. https://doi.org/10.1080/00207454.2020.1774577

Martinez-Nunez, A. E., Fynke, J. E., & Miller, D. J. (2021). Under pressure: Pressure-dependent pontine compression by a dolichoectasia basilar artery, a case report. *The Neuroradiology Journal*, *34*(6), 686–687. https://doi.org/10.1177/19714009211017785

Oishi, Y., Ueda, R., Kuranari, Y., & Imanishi, T. (2018). Vertebrobasilar Dolichoectasia Causing Symptomatic Hydrocephalus Due to Third Ventricular Compression. *Journal of Stroke and Cerebrovascular Diseases*, *27*(12), 3503–3504. https://doi.org/10.1016/j.jstrokecerebrovasdis.2018.08.021

Prasad, S. N., Singh, V., Selvamurugan, V., & Phadke, R. V. (2021). Vertebrobasilar dolichoectasia with typical radiological features. *BMJ Case Reports*, *14*(2). https://doi.org/10.1136/bcr-2020-239866

Umana, G. E., Alberio, N., Graziano, F., Fricia, M., Tomasi, S. O., Corbino, L., Nicoletti, G. F., Cicero, S., & Scalia, G. (2021). Vertebrobasilar Dolichoectasia, Hypoplastic Third Ventricle, and Related Biventricular Hydrocephalus: Case Report and Review of the Literature. *Journal of Neurological Surgery. Part A, Central European Neurosurgery*. https://doi.org/10.1055/s-0041-1725049

Wang, J., Jia, L., Yang, X., Jia, X., Liu, J., Liu, P., Miao, Z., Zhang, Y., Tian, Z., Wang, K., Wang, Z., Zhang, Y., & Lv, M. (2019). Outcomes in symptomatic

patients with vertebrobasilar dolichoectasia following endovascular treatment. *Frontiers in Neurology*, *10*(JUN). https://doi.org/10.3389/fneur.2019.00610

Zdravković, M., Kostov, M., Petrovic, S., & Zdravkovic, J. (2017). Dolichoectasia of Vertebrobasilar Arteries as a Cause of Hydrocephalus and Ischemic Cerebral Stroke. *American Journal of Forensic Medicine and Pathology*, *38*(4), 359–361. https://doi.org/10.1097/PAF.0000000000000348

Zhang, D. P., Yin, S., Zhang, H. L., Li, D., Song, B., & Liang, J. X. (2020). Association between Intracranial Arterial Dolichoectasia and Cerebral Small Vessel Disease and Its Underlying Mechanisms. *Journal of Stroke*, *22*(2), 173–184. https://doi.org/10.5853/jos.2019.02985

Zhang, X., Jing, J., Zhao, X., Liu, L., Meng, X., Wang, A., Pan, Y., Wang, D., Wang, Y., & Wang, Y. (2018). Prognosis of dolichoectasia in non-cardioembolic transient ischemic attack and minor stroke. *Neurological Research*, *40*(6), 452–458. https://doi.org/10.1080/01616412.2018.1451016

Figure Legends

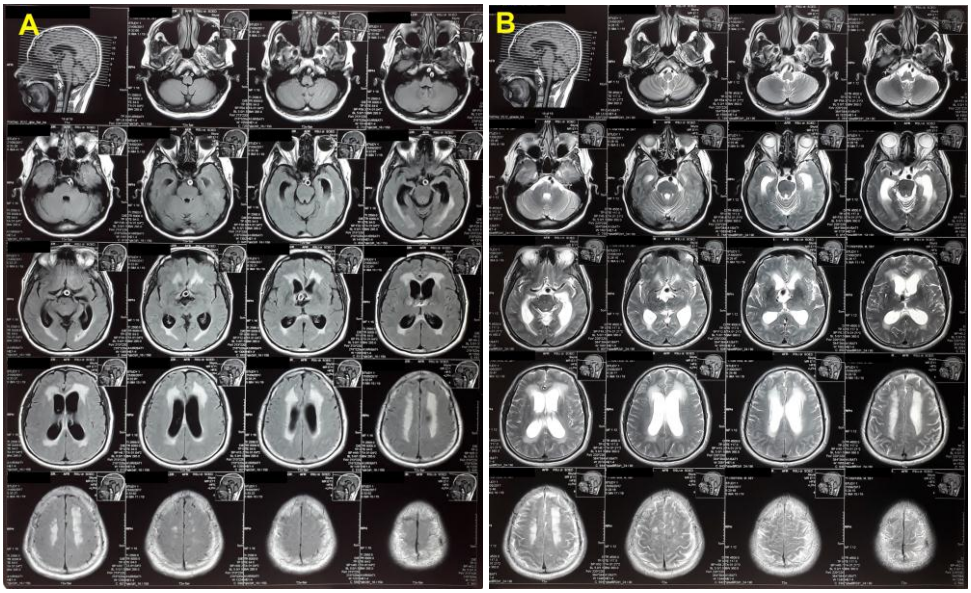

Figure 1. Axial head MRI sequences T2 Flair (A) and T2 (B)

Flair's T2 sequence showed periventricular edema as a sign of active hydrocephalus. CSF flow compression is seen in the T2 sequence.

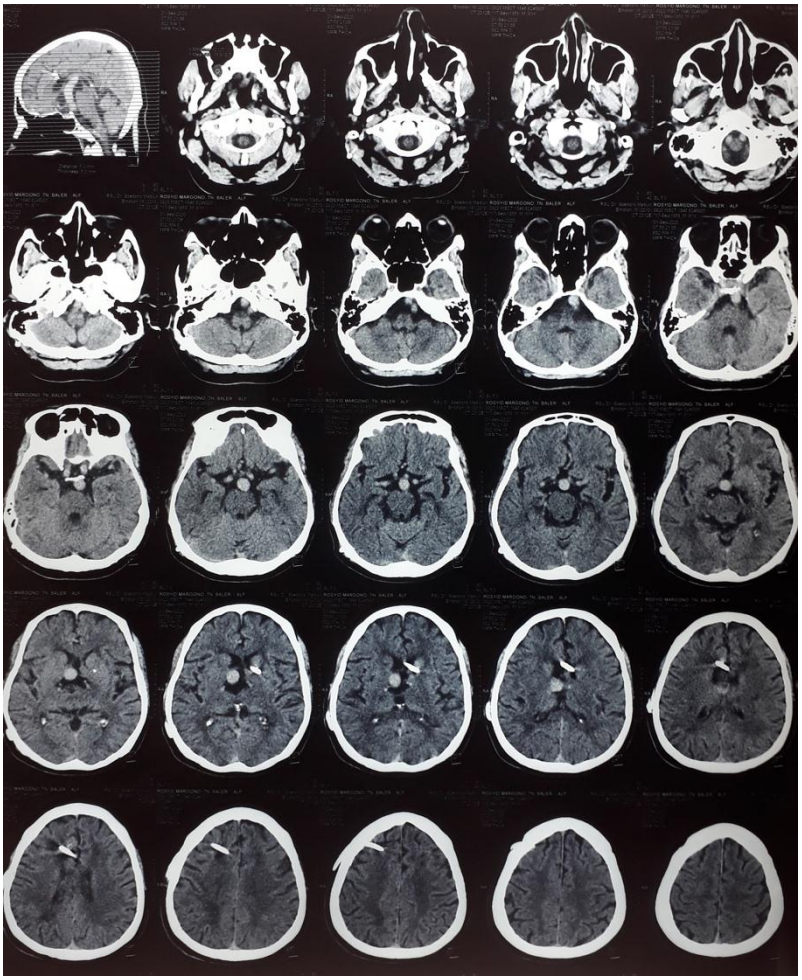

Figure 2. CT Scan of the axial head without contrast

CT scan of the head post VP shunt installation. There was a resolution of hydrocephalus with dolicoectatic images of the vertebrobasilar vessels.

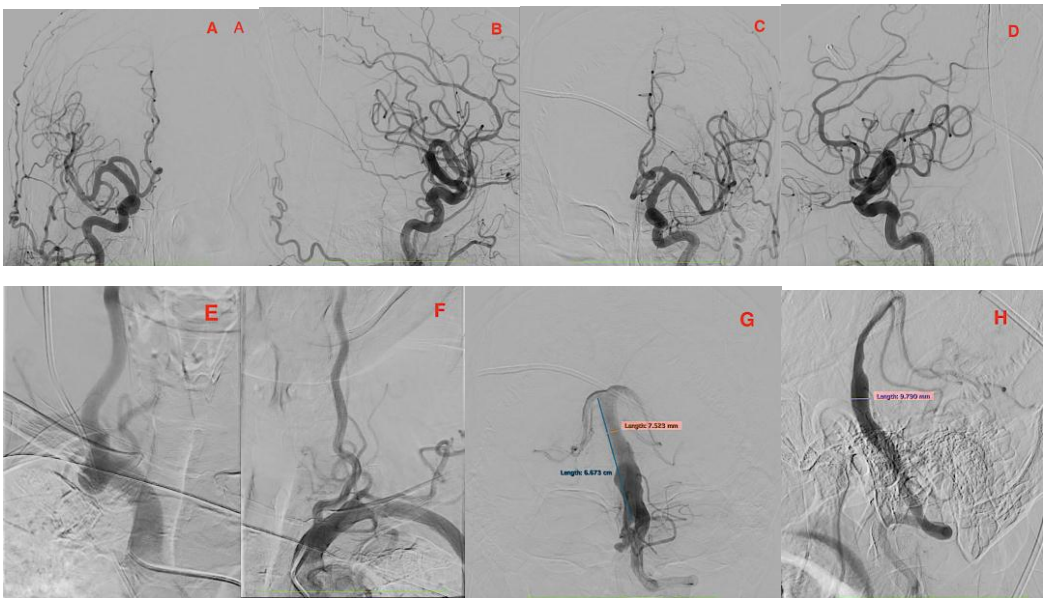

Figure 3. Findings on DSA

DSA in a 62-year-old man with suspected basilar fusiform aneurysm from MRI-MRA results. Injection of Right ICA AP (A), Lat (B) found normal ACA and MCA, without other vascular abnormalities; Left ICA injection AP (A), Lat (B) found ACA and MCA normal, without other vascular abnormalities.; RVA (E) and LVA (F) injections showed tortous access in VA, basilar dolichoectasia; The morphology of the basilar artery appeared AP (G) with a length of 66.7 mm – 7.52 mm in diameter and Lat (H) according to the VBD criteria.

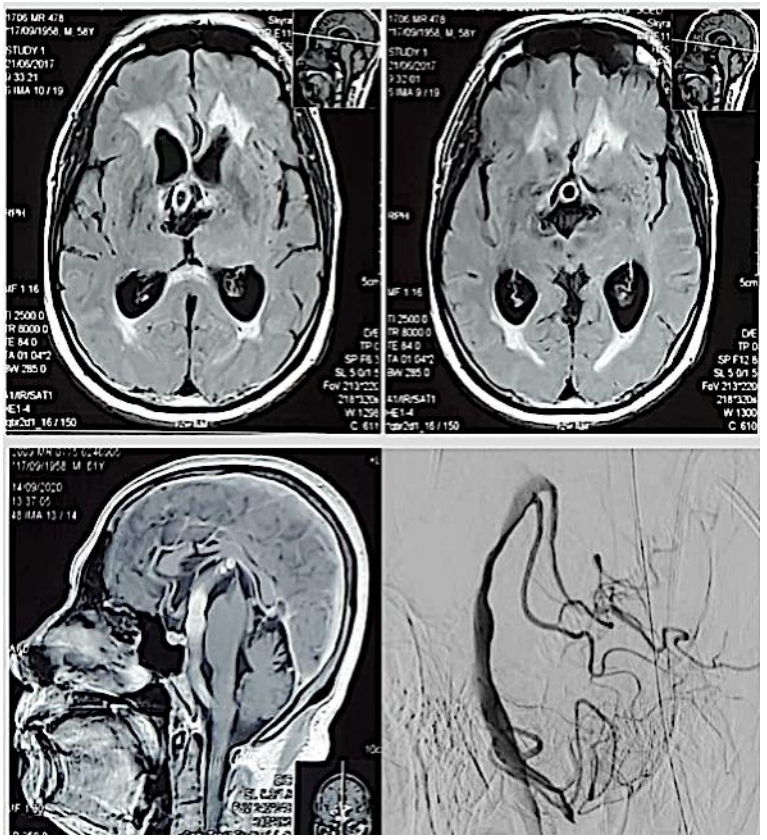

Figure 4. VBD findings on MRI and DSA.

Visible location and anatomical morphology that shows VBD in patients.