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Vestibulo-Cochlear Symptoms Revealing Vertebro-Basilar Dolicho-Ectasia

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Abstract | **Case Report**

Introduction: Vertebrobasilar dolichoectasia (VBD) are uncommon arteriopathies defined by increased length and caliber of one or more intracranial arteries. The clinical picture is polymorphic. They are most often associated with ischemic or hemorrhagic stroke, but this pathology can also cause symptoms of cranial nerve compression. ***Objective***: We report the case of a vertebro-basilar dolichoectasia collected in the radiology department of Ibn Tofail Hospital in Marrakech, revealed by vestibulo-cochlear symptoms and diagnosed by brain MRI. In the light of this observation, we review this uncommon pathology and the contribution of magnetic resonance imaging in its diagnosis. ***Case report:*** The patient was 68 years old and had a history of severe sleep apnea syndrome. He presented with rotatory vertigo, tinnitus and hypoacusis on the left side of the head for 3 months. The neurological examination was normal and the examination of the ENT sphere revealed bilateral sensorineural hearing loss and left hyporeflexia. A cerebral MRI was performed as part of the etiological workup, which revealed elongation and dilation of the vertebral arteries measuring 3.9 mm on the left and 5.2 mm on the right, with loops of which the left crossed the homolateral cochleovestibular nerve 7 mm from its emergence, responsible for a deviation of its path, and the glossopharyngeal nerve 4 mm from its emergence. It also showed an elongation and dilatation of the basilar trunk measuring 6.8 mm, presenting a loop lateralized to the left at the level of its origin (Smoker's Grade 2) with a bifurcation opposite the floor of the V3 (Smoker's Grade 2).Thus, the diagnosis of a DEVB responsible for a vascular-nervous conflict was retained. The indication for surgical management was given but was refused by the patient due to the deep location of the arteriopathy. ***Discussion and conclusion:*** DEVB is a rare and progressive condition with cranial nerve compression as a possible complication. Brain MRI is the diagnostic method of choice. The positive diagnosis of arterial dolichoectatic disease in general and of a vertebral mega-artery is not easily recognized by radiologists because of the frequency of normal variations in the length and diameter of the artery. Radiological exploration allows to identify the vascular-nervous conflict, to specify the degree of nerve compression, to foresee the surgical difficulties and the evolutionary follow-up of the disease.

Keywords: Vertebrobasilar dolichoectasia (VBD), intracranial arteries, hemorrhagic stroke, hyporeflexia.

INTRODUCTION

Vertebrobasilar dolichoectasia (VBD) are uncommon arteriopathies defined by increased length and caliber of one or more intracranial arteries [1].

The clinical picture is polymorphic. They are most often associated with ischemic or hemorrhagic stroke, but this pathology can also cause symptoms of cranial nerve compression [2].

In the majority of reported cases, the facial and trigeminal nerves are most likely to be involved. In contrast, abducens, trochlear, vestibular, glossopharyngeal, or vagal nerve involvement is rare [3].

We report the case of a vertebro-basilar dolichoectasia collected in the radiology department of Ibn Tofail Hospital in Marrakech, revealed by vestibulo-cochlear symptoms and diagnosed by brain MRI.

In the light of this observation, we review this uncommon pathology and the contribution of magnetic resonance imaging in its diagnosis.

CASE REPORT

The patient was 68 years old and had a history of severe sleep apnea syndrome. He presented with rotatory vertigo, tinnitus and hypoacusis on the left side of the head for 3 months.

The neurological examination was normal and the examination of the ENT sphere revealed bilateral sensorineural hearing loss and left hyporeflexia.

A cerebral MRI was performed as part of the etiological workup, which revealed elongation and dilation of the vertebral arteries measuring 3.9 mm on the left and 5.2 mm on the right, with loops of which the left crossed the homolateral cochleovestibular nerve 7 mm from its emergence (at the level of its REZ zone), responsible for a deviation of its path, and the glossopharyngeal nerve 4 mm from its emergence (exceeding the REZ zone) (Figure 1). It also showed an elongation and dilatation of the basilar trunk measuring 6.8 mm, presenting a loop lateralized to the left at the level of its origin (Smoker's Grade 2) with a bifurcation opposite the floor of the V3 (Smoker's Grade 2) (Figure 2 and Table 1).

Thus, the diagnosis of a DEVB responsible for a vascular-nervous conflict was retained. The indication for surgical management was given but was refused by the patient due to the deep location of the arteriopathy.

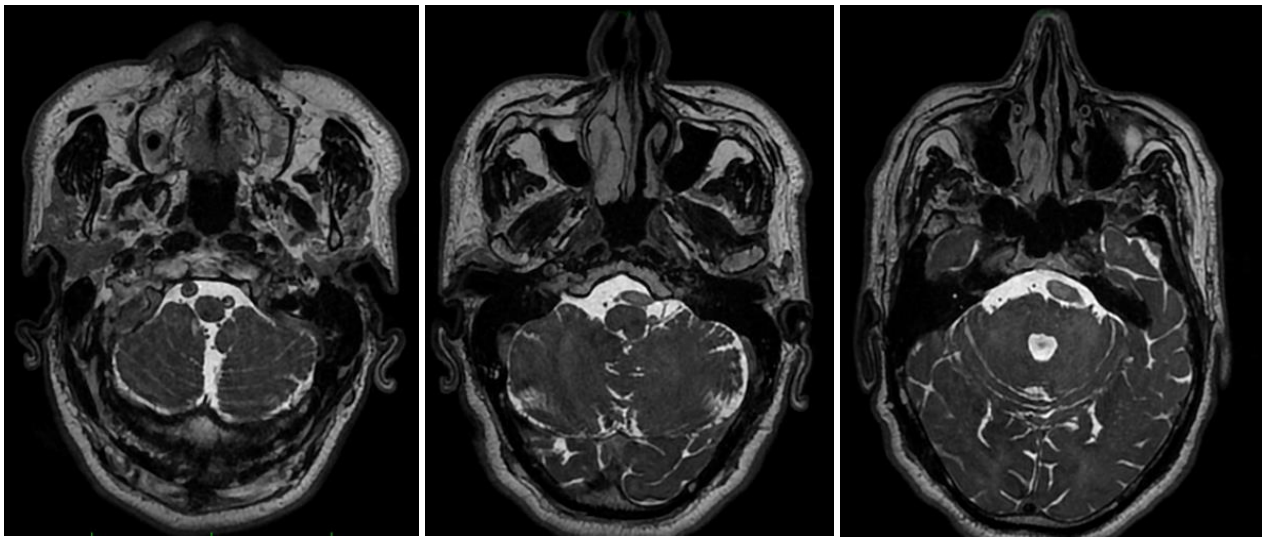

Figure 1: Brain MRI, T2 axial sequence: vertebral artery dilation measuring 3.9 mm on the left and 5.2 mm on the right (A); left vertebral artery crossing the cochleovestibular nerve at its REZ (B); dilated basilar trunk lateralized on the left (Smoker's Grade 2)

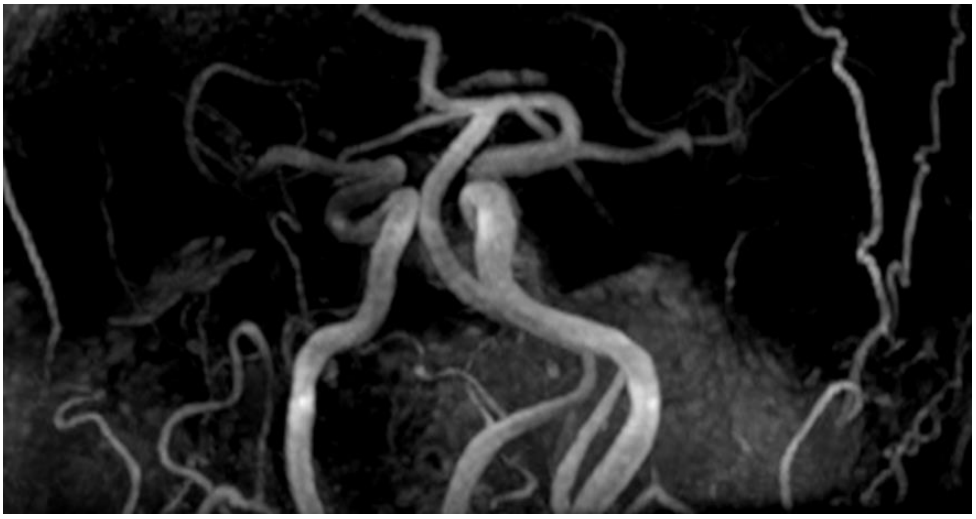

Figure 2: Intracranial magnetic resonance angiography (MRA): vertebrobasilar dolichoectasia with a basilar trunk diameter of 6.8 mm, Smoker grade 2 TB bifurcation height, and grade 2 TB lateralization

Table 1: Critères de Smoke (IRM) pour le diagnostic de la DEVB *IRM (imagerie par résonance magnétique); TB (tronc basilaire); V3 (3 ème ventricule)*

Critère	Grade	IRM
Diamètre du TB	0	1.9 – 4.5 mm
Score de 1 est anormal	1	> 4.5 mm
Latéralité	0	Le long de la ligne médiane
Score ≥2 est anormal	1	Médial au bord latéral du clivus ou du dorsum sellae
	2	Latéral au bord latéral du clivus ou du dorsum sellae
	3	Au niveau de l'angle ponto-cérébelleux
Hauteur de la bifurcation du TB	0	Au niveau ou en dessous du dorsum sellae
Score ≥2 est anormal	1	Au niveau de la citerne suprasellaire
	2	Au niveau du V3
	3	Refoulant et surélevant le V3

DISCUSSION

DEVB is an uncommon arteriopathy characterized by progressive ectasia, elongation and tortuous appearance of the vertebrobasilar system. Its prevalence is estimated at 0.05-18% [1]. Risk factors associated with the development of this entity include advanced age, male gender, smoking, hypertension, and a history of myocardial infarction [4].

To date, the mechanism contributing to DEVB has not been elucidated. The main pathophysiological hypothesis involved in DEVB may reflect aberrant vascular remodeling and arterial wall degradation caused by high blood pressure [4].

The clinical manifestations of DEVB, which is often asymptomatic, can vary considerably, ranging from exertional headache to fatal consequences such as ischemic or hemorrhagic stroke. It is considered a potentially serious condition that can cause dysphagia, dyspnea, quadriplegia, or hemiparesis due to the compression of the posterior cerebral fossa [5].

Compression of the lower brainstem can trigger vestibulo-cochlear symptoms such as deafness, tinnitus, and vertigo. Few cases of DEVB involving the cochleovestibular nerve have been reported [6]. The trigeminal and facial nerves are more likely to be commonly involved; abducens nerve pain resulting from VBD-related compression is relatively rare. Of note, multiple cranial nerve involvement in patients with VBD is rarely reported [7].

The positive diagnosis of arterial dolichoectatic disease in general and of a vertebral mega-artery, is not readily recognized by radiologists given the frequency of normal variations in arterial length and diameter. With the advent of computed tomography (CT), cerebral magnetic resonance imaging (MRI), and vascular imaging, vertebral mega-artery can be diagnosed noninvasively. Radiological exploration should include 3D-T1 MRI sequences without and with gadolinium injection, 3D-T2-high resolution and MRA-TOF [8]. This exploration makes it possible to identify the vascular-nervous impingement, to specify the degree of compression of the nerve, to foresee the surgical difficulties and the evolutionary follow-up of the disease [8]. The 3D-T1 and 3D-T2-high resolution sequences give a good contrast between cerebrospinal fluid and vascular-nervous structures.

Smoker *et al*., [9] were the first to establish criteria for the diagnosis of DEVB: the basilar trunk is considered dilated if its diameter exceeds 4.5 mm, and it is considered elongated if it lies next to the lateral border of the clivus or dorsum sellae. Thus, the Smoker criteria use three quantitative measures of basilar trunk morphology to diagnose dolichoectasia: laterality, bifurcation height, and basilar trunk diameter. Application of Smoker's criteria allows the diagnosis of DEVB. Elongation is indicated by abnormal laterality or height, ectasia is indicated by abnormal diameter, and dolichoectasia is indicated by the presence of both elongation and ectasia (Table 1).

Vasculo-nerve impingement is only responsible for cranial pair neuralgia when the compression is exerted on the nerve root and more precisely at the level of the Root Entry/exit zone (REZ) [10]. This zone corresponds to a region of hyper-reactivity and hyperexcitability, anatomically located at the transition zone between central and peripheral myelin [11].

Currently, there is no effective treatment for DEVB itself because of the location and pathway of the anomaly and the large main and perforating arteries that supply the brainstem [2]. In cases of symptomatic

neurovascular impingement, there is no specific treatment. Some studies have reported that microvascular decompression has successfully treated vascular compression of the oculomotor, trochlear, trigeminal, abducens and facial nerves [12]. However, most reported cases were treated conservatively, including medical treatment of symptoms, because the surgical risk outweighed the benefit [13].

CONCLUSION

DEVB is a rare and progressive condition with cranial nerve compression as a possible complication. Brain MRI is the diagnostic method of choice.

The positive diagnosis of arterial dolichoectatic disease in general and of a vertebral mega-artery is not easily recognized by radiologists because of the frequency of normal variations in the length and diameter of the artery. Radiological exploration allows to identify the vascular-nervous conflict, to specify the degree of nerve compression, to foresee the surgical difficulties and the evolutionary follow-up of the disease.

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