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BRAINSTEM COMPRESSION SYNDROME CAUSED BY VERTEBROBASILAR DOLICHOECTASIA

Microvascular repositioning technique

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Vertebrobasilar dolichoectasia (VBD) is an anatomic variant that consists of enlargement and dilatation, often associated with a tortuous and elongated vessel[1]. The anomaly is probably due to a marked thinning of the internal elastica lamina and media, most likely as a consequence of prolonged systemic arterial hypertension[2]. It accounts for approximately 3 to 5% of all cerebellopontine mass lesions. A variety of clinical syndromes have been related due to pulsatile compression by the aberrant vessel: cerebellar dysfunction, hydrocephalus, ischemic stroke, transient or permanent motor deficits, central sleep apnea, trigeminal neuralgia, as well as brain stem compression syndrome[1,3,4]. Microvascular decompression surgery was introduced in the 1960s and was initially used to treat trigeminal neuralgia, hemifacial spasm and glossopharyngeal neuralgia[5]. Lately, it was used to treat brainstem dysfunction caused by an ectatic vessel[1]. Nowadays, microvascular decompression with repositioning of the ectatic vessel is a new technique that has been used successfully.

The purpose of this study is to report and discuss a rare case of brain stem compression syndrome caused by vertebrobasilar dolichoectasia successfully treated with microvascular decompression repositioning technique and documented by computed tomography angiography (CTA) and magnetic resonance imaging (MRI).

CASE

A 60-years old man with a past medical history of diabete mellitus type 2 sought neurological treatment after experiencing mild progressive disartria for eight months. He did not have other complaints.

The patient's neurologial examination revealed, besides the speech abnormality, left side pyramidal syndrome with hiperreflexia and Babinski´s sign. All the other aspects of the neurological examination were intact.

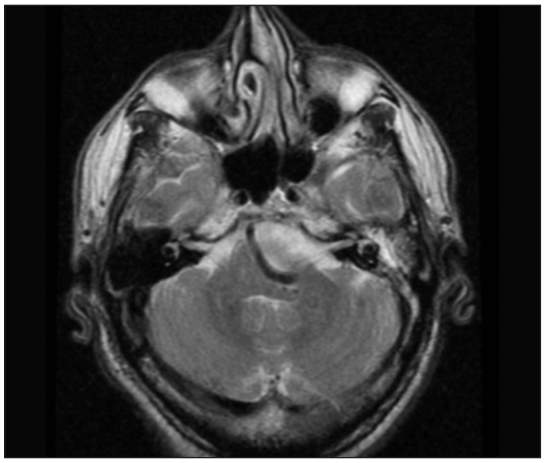

Fig 1. MRI (Flair - Axial) showed an elongated and tortuous vertebrobasilar artery that crossed the ventral aspect of the medulla oblongata causing mechanical compression at the left side.

Imaging investigation

MRI and CTA showed an elongated and tortuous vertebrobasilar artery that crossed the ventral aspect of the medulla oblongata causing mechanial compression at the left side (Figs 1 and 2).

Surgical technique

The patient was placed in the prone oblique (*park bench*) position, and a left far lateral suboccipitoretromastoidea approach was performed with left vertebral artery exposure. The dura was opened, and cerebrospinal fluid was released at the cisterna magna to provide a capacious working environment.

Arachnoid dissection revealed a large vascular structure, identified as the basilar dolichoectatic artery, dislocating and compressing the brain stem (medulla oblongata) in its left ventral region. As soon as the neurovascular conflicting area was

SÍNDROME COMPRESSIVA BULBAR CAUSADA POR DOLICOECTASIA VERTEBROBASILAR: TÉCNICA DE REPOSICIONAMENTO MICROVASCULAR

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identified, microvascular decompression repositioning technique, adapted from Yoshimoto et al, was performed and the tortuous dolichoectatic vertebrobasilar artery was pulled toward and fixed with unabsorbable nylon on the nearby dura mater, achieving a very satisfactory decompression result (Fig 3)[6].

Postoperative course

The patient had an excellent postoperative course. Seven days after the surgery, he was discharged with a discrete motor speech abnormality and a persistent Babinski´s sign on left side.

At a 6-month follow-up examination, the patient had recovered all neurological functions and had no complaints.

DISCUSSION

Vertebrobasilar dolichoectasia is a potentially severe condition that may cause severe disability due to ischemic or compressive dysfunction in the posterior fossa. It is an uncommon entity that afects less than 0.05% of the population, and accounts for aproximately 3 to 5% of all cerebellopontine angle mass lesions. Small cases series observed that the survival rate in VBD after 3 years follow-up was found to be 60%[1]. The epidemiological data about VBD are limited. Ubogu et al. reviewed 1440 magnetic resonance angiography (MRA) between 1995 and 1997, and found a VBD prevalence of 4.4% in their series. The same author also found that VBD is more common in women, 32 of 45 cases, and equally distributed between whites and african-american. Among their sample, 42% of the patients had incidental VBD findings on MRA performed on varius indications other than posterior circulation dysfunction[1]. Resta et al. observed 132 patients with dolichoectatic vertebrobasilar artery among 2256 angiographies, and only 7.73% have had neurological symptoms[7]. Necropsies series showed that 30% of the patients with the abnormality had symptoms, which lead us to think that the real incidence of vertebral ectasia and the frequency by which this anatomic feature induces neurological symptoms are still uncertain[8,9].

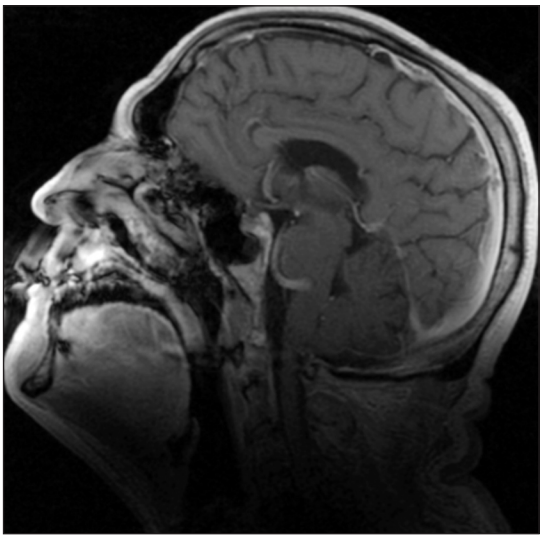

Fig 2. MRI (T1 Gadolinium - Sagittal) showed an elongated and tortuous vertebrobasilar artery that crossed the ventral aspect of the medulla oblongata causing mechanical compression at the left side.

Some authors suggest that VBD may be a congenital vasculopathy of the elastic layer of the arterial wall, and may be cause of posterior circulation dysfunction independent of atherosclerosis disease affecting the intimal layer[1,3,10]. Others suggest a hypothesis that try to explain

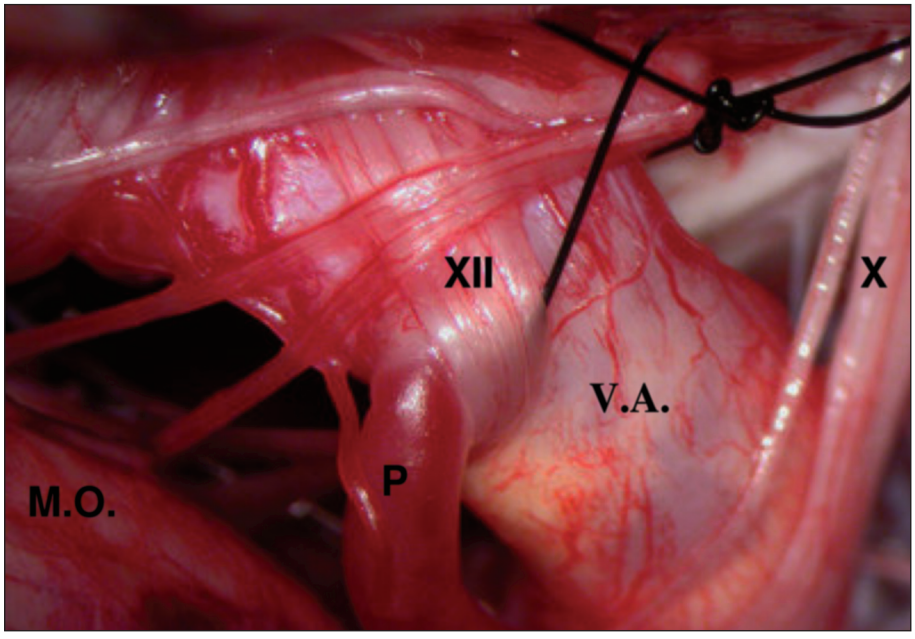

Fig 3. Left far lateral suboccipitoretromastoidea approach: transoperative photography showing the microvascular repositioning technique. M.O., medulla oblongata; P, posterior inferior cerebellar artery; V.A., Vertebral artery; XII, hypoglossal nerve; X, vagus nerve.

the posterior circulation ischaemia by the induced atherosclerosis in dolichoectatic arteries at the regions of maximum angulation[10-12]. Some still believe in the correlation between prolonged systemic arterial hypertension and the marked thinning of the internal elastic lamina and media of the abnormal vessel[2].

Padget et al., in 1954, proposed a developmental origin as a consequence of the zig zag pattern that the vertebral artery assumes during its development, to explain the artery tortuousity, although degenerative changes in the vessel might be considered[13]. Ubogu et al. suggest that VBD is part of a more widespread vasculopathy[1]. These author also studied the radiological features of the VBD and demonstrated that the major VBD location is the basilar artery alone (40%), followed by bilateral vertebral arteries, 22% basilar artery and both vertebral arteries, 16%. Basilar artery and single vertebral artery presentation, as demonstrated in the present case, account for only 4% of VBD location.

A cohort study define VBD as a basilar artery or vertebral artery diameter >4.5 mm or deviation of any portion of them higher than 10 mm from the shortest expected course, or basilar lenght >29.5 mm or intracranial vertebral artery lenght >23.5 mm[1]. Smoker et al. judged a basilar artery to be elongated if, at any point along its course, it lay lateral to the margin of the clivus or dorsum sellae or bifurcated above the plane of the suprasellar cistern[14].

Since there are no well established imaging criteria for VBD, some authors suggest that the clinical diagnosis of symptomatic VBD should be made based on posterior circulation dysfunction in the absence of significant stenotic or occlusive disease of the posterior circulation with an ectatic and tortuous vessel present on angiography, and no other potential cause for the symptoms[1]. The usefulness of neuradiological methods such as computed tomography and MRI are still controversial. MRI sensivity and specificity to diagnose VBD are unknown[15]. Some authors emphasized that good results depend on the correct MRI projection (oblique sagittal) and the appropriate gradient echo sequences. However, this is an acceptable method for diagnose and reduces morbidity of an invasive procedure.

The VBD anomaly can cause two kinds of symptoms: those resulting from the compression of structures adjacent to the aberrant vessel, such as the brainstem and cranial nerves roots compression and those resulting of ischemic events[1,9]. Cerebellar dysfunction, hydrocephalus, trigeminal neuralgia, ischemic stroke and medulla oblongata compression are some of these syndromes that have been associated with the pulsatile compression produced by the ectatic vessel. Levine et al. previously reviewed literature and showed that in 128 patients, 58% experienced cranial nerve compression, most of them complaining about facial spasm and trigeminal neuralgia, 48% experienced vertebrobasilar insufficiency or stroke or both, 31% had hydrocephalus and only 24% had symptoms and signs from brain-stem compression[16].

Kim et al., in 1985, were the first to report brainstem dysfunction caused by vascular compression[17]. Experimental studies have shown that rapid mechanical compression of the brainstem induces immediate damage, and this is directly proportional to the degree of compression. In patients with VBD, the compression has a slowly progression, so the brainstem can functionally tolerate severe distortion without overt clinical manifestations, wich explain why results in necropsy series and angiographies analysis are so different[18]. These conclusions, however, do not exclude the possibility of more subtle impairment of neural structures. Passero et al. showed that the most frequent subclinical abnormalities were the prolongation of blink reflex components and changes of the motor evoked potentials of upper and lower limbs. The authors suggest that functional testing, such as brainstem auditory evoked potencials (BAEPs), blink reflex (BR) and motor evoked potencials may be useful for long term monitoring of patients with VBD and may help in the decision making process prior to the surgical approach for relief of subjective symptoms[19].

Besides pyramidal tract signs, brain stem compression by vascular structures may also cause vertigo, dysphagia, sialorrhea, velar paresis, imbalance, tinnitus and others. Janneta et al., in experimental studies in 1985, induced hypertension values with a balloon model and found out the relationship between pulsatile brain stem compression as a possible cause of essential neurogenic hypertension[20]. In some rare circumstances, compression of the medulla oblongata by an aberrant vessel may also cause sleep disordered breathing such as central or obstructive apnea[3].

Microvascular decompression (MVD) was introduced in the late 1960s for treatment of cranial nerve disorders such as trigeminal neuralgia and hemifacial spasm, but it was Jannetta et al. that fifteen years later largely popularized it. Limited exposure, deep working area, and the potencial severity of adverse outcomes that can result from postoperative infarction and swelling are all characteristics of operations in the posterior fossa[21]. The goal of MVD for treatment of these disorders is complete alleviation of the symptoms with minimization of those surgical complications[22]. Microvascular decompression has become the appropriate operative technique to relieve simple neurovascular compression syndromes. It is a safe and highly effective procedure and is usually obtained by

interposing Teflon felt or padding or other synthetic implants between the offending vessel and the nerves[23].

Brainstem compression caused by VBD can not be treated using an ordinary technique of microvascular decompression. In the surgical strategy of this condition, it is not sufficient to achieve decompression only by insertion of prosthesis between the vessel and the brainstem, because the size and stiffness of the dilated artery makes standard microsurgery dificult. Moreover, the overall size of the ectatic vessel and its increased pulsatility do not allow an effective separating action by the prosthesis[3]. Besides that, an increased neural compression may be expected with the placement of prosthesis in the conflict area in such condition. Therefore, alternative surgical techniques should be considered.

Different techniques have been described by some authors in the literature. Ogawa et al. reported the use of a synthetic vascular graft sutured to the clival dura to move the vessel away from cranial nerves[24]. A silicone sling sutured to the petrous dura to reposition the basilar artery away from the trigeminal nerve was used by Stone et al.[25]. A vascular clip graft or a Silastic ring around the trigeminal nerve to isolate it from the offending vessel was reported by Laws et al. and Yoshimoto et al., respectively[6,26]. A silastic rubber sling stitched to the dura was used by Rawlinson and Coakham in patients with hemifacial spasm[27]. Fukushima and Kondo reported the use of teflon slings and pieces of Surgicel to mobilize the offending vessel and promote fibrous adhesions of the Teflon to the dura[28,29] Recently, alternative technique for neurovascular decompression of the brain stem have been reported, however its long-term success rate is still unknown and other studies are needed in order to prove its efficacy.

In the present case, we used a very simple microsurgical repositioning technique: the tortuous dolichoectatic vertebrobasilar artery was pulled toward and fixed with unabsorbable nylon on the nearby dura-mater, achieving a very satisfactory brain stem decompression result. The follow-up period in this case is not so long, but the excellent postoperative outcome obtained until today make us believe that the cure in this patient is very likely to be achieved.

In conclusion, vertebrobasilar dolichoectasia is a very rare cause of brain stem compression and a satisfactory outcome can be obtained with microvascular repositioning technique.

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