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Anatomoclinical features of trigeminal neuralgia caused by vertebrobasilar dolichoectasia

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SUMMARY

Vertebrobasilardolichoectasia (VBDE) is a rare cause of trigeminal neuralgia (TN). The size and tortuous course of the offending vessel poses a particular challenge when attempting a surgical treatment with microvascular decompression. There are few reports on the radiological and surgical anatomy encountered in this condition. The anatomical and radiological features of a surgical series of 7 patients with TN caused by VBDE and treated with microvascular decompression (MVD) are presented. Morphometric measurements and three-dimensional reconstructions obtained from preoperative magnetic resonance imaging were compared with microsurgical findings. Trigeminal nerve compression was found on the left side in six cases (86%). The neurovascular compression was caused by the basilar artery (BA) in 4 cases and the vertebral artery (VA) in three cases, with two cases showing a multiple compression involving other vessels. The BA showed a mean maximal diameter of 6,5 mm (5-9,1 mm), a mean lateral deviation of 19,3 mm (14,4- 22,1 mm) and a mean elevation of the basilar tip above the dorsum sellae of 10,5 mm (4,4-14,8 mm). MVD was successfully performed in all patients yielding a permanent pain relief in six of the patients. Preoperative assessment of the neurovascular relations within the cerebellopontine angle is paramount for the surgical planning in patients with TN caused by VBDE.

Key words: Vertebrobasilar dolichoectasia – Trigeminal neuralgia

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INTRODUCTION

Vertebrobasilar dolichoectasia (VBDE) is a vascular disorder characterized by a dilated, elongated and tortuous vertebrobasilar system (Sacks et al., 1969). The incidence varies from 0.06% to 5.8% according to several surgical series (El-Ghandour, 2010; Passero et al., 2008; Vasovic et al., 2012). A recent review of 7345 asymptomatic individuals in an adult Japanese population encountered a 1.3% rate of VBDE on brain magnetic resonance (Ikeda et al., 2010). It is considered an acquired condition that appears in relation to hypertension and atherosclerosis (Kirsch et al., 1996). VBDE is frequently an asymptomatic condition, but compressive symptoms and ischemic events due to thromboembolism have been reported. Trigeminal neuralgia (TN) and hemifacial spasm have been described as the most frequent clinical expressions of VBDE, while brainstem compression and obstructive hydrocephalus are considered extremely rare (Linskey et al., 1994; Miyazaki et al., 1987).

The anatomical and radiological features of a surgical series of patients presenting with TN caused by VBDE are discussed. Measurements of the abnormal dilation and elongation of the vertebrobasilar system, as well as the three-dimensional reconstruction of the neurovascular relations in the cerebello- pontine angle, highlight the importance of preoperative magnetic resonance imaging for surgical planning in these cases.

MATERIALS AND METHODS

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We present a series of 7 cases of TN caused by VBDE out of 129 patients (5.4%) who underwent a microvascular decompression (MVD) at our department during an 18-year period (1997-2015). Clinical features, radiological and surgical findings of seven patients (6 men and 1 woman) with TN caused by VBDE were retrospectively analyzed by reviewing their medical charts and surgical protocols. Radiological images and reports were collected for each patient. In the 6 most recent cases, whose preoperative magnetic resonance imaging (MRI) and angiographic computed tomography (angioCT) were available on electronic support as DICOM (Digital Imaging and Communication in Medicine) images, morphometric measurements of the vertebrobasilar system were obtained. All measurements were performed on sagittal T1-weighted and DRIVE sequences (driven equilibrium radio frequency reset pulse) generated with a 1.5 Achieva System (Philips Medical Systems, DA Best, Netehrlands). These included the diameter of the basilar artery (BA) at the level of the midpons, the elevation of the basilar tip above the dorsum sellae and the maximal lateral deviation from the clival midline. In order to illustrate the compression of the trigeminal nerve, three-dimensional reconstructions of neurovascular elements of the posterior fossa were created using the smartbrush tool by BrainLab (Feldkirchen, Germany) (Fig. 1).

All patients in this series complained of severe pain refractory to different antiepileptic drugs and were therefore offered a surgical treatment consisting in MVD. With the patient in a three-quarter prone position, a lateral suboccipital craniectomy was performed. A retrosigmoid approach to the cerebello-pontine angle was carried out and the cisternal segment of the trigeminal nerve from the root entry zone at the pons to the porus trigeminus was carefully inspected for vascular compressions under surgical microscope magnification. Arachnoidal adhesions were dissected with microsurgical technique and a MVD was performed by inserting pieces of Teflon® between the nerve and the compressing vessel.

The mean follow-up was 51 months (range 6-180 months).

RESULTS

Between January 1997 and July 2015, 129 patients presenting pharmacologically refractory classical TN underwent surgical treatment with MVD. On the preoperative MRI study, 7 of these patients (5.4%) showed a VBDE, consisting in a tortuous and dilated vertebrobasilar system that invaded the cerebello-pontine angle causing a

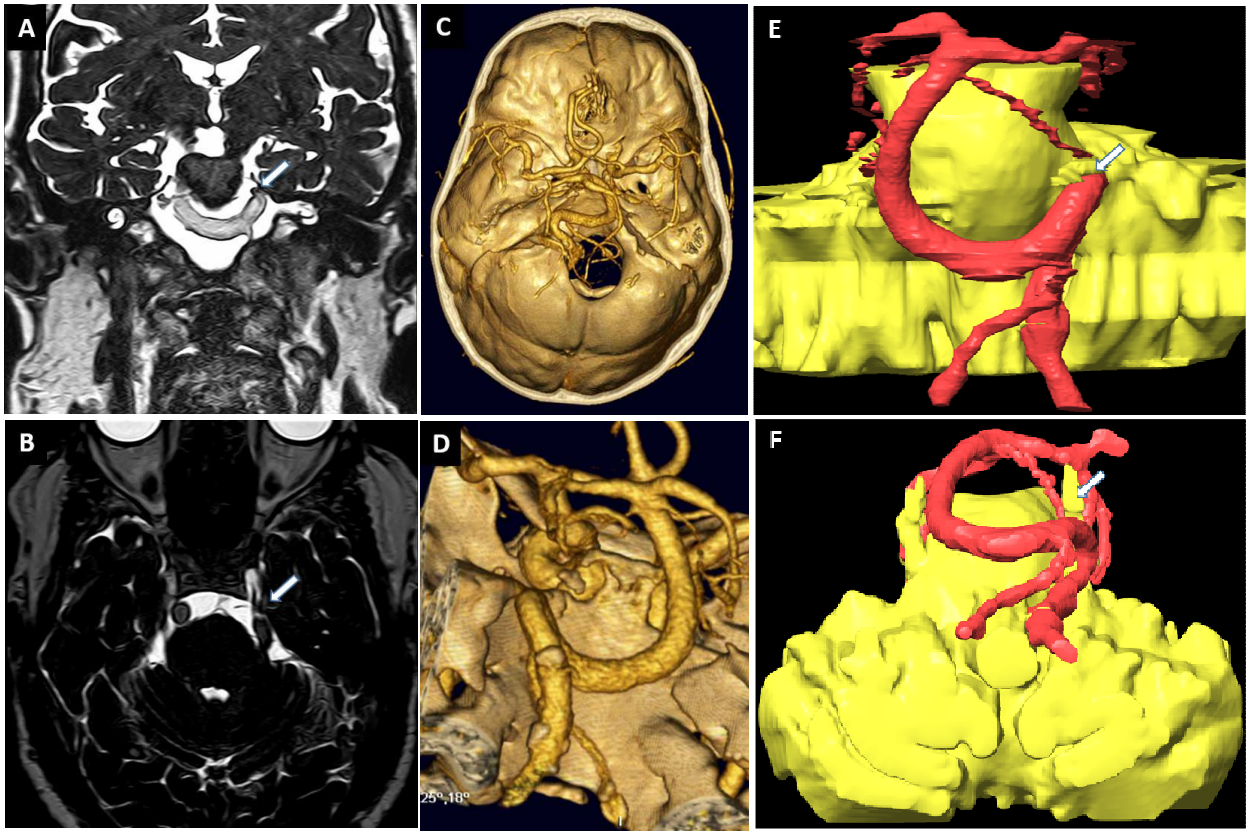

Fig. 1. (A,B) MRI DRIVE coronal and axial sequences showing a dolichoectatic vertebrobasilar system which causes compression of the left trigeminal nerve. **(C,D)** The angio-CT demonstrates a VBDE with a dominant left vertebral artery reaching up to the level of the petrous ridge**. (E,F)** Three-dimensional reconstructions illustrating the relation between the left trigeminal nerve (arrow) and the left vertebral artery.

compression of the trigeminal nerve. The median age was 67 years and patients had been diagnosed of trigeminal neuralgia for a mean duration of 8 years. Six of the patients (86%) had a left-sided facial pain. The pain showed the typical features of classic TN with daily paroxysms of shooting hemifacial pain radiated to one or more trigeminal territories without signs of motor or sensitive deficit. Pain affected the territory of the second trigeminal division in 6 patients, the first division in 4 patients and the third division in 3 patients. There was no case of ipsilateral hemifacial spasm and 6 of the patients had a history of arterial hypertension.

The existence of a VBDE was diagnosed on MRI in all cases. The vertebrobasilar system showed a significant asymmetry of the diameter of the vertebral arteries in 5 patients. There was a dominance of the vertebral artery (VA) ipsilateral to the TN in two cases and of the contralateral VA

Table 1. Clinical and morphometric features

Case	Age/sex	Side	Pain distribution	Compressing Vessel	Maximal diameter BA (mm)	Maximal lateral deviation (mm)	Elevation above dorsum sellae (mm)
1	69/W	Left	V123	VA	5	-	-
2	58/M	Right	V2	BA	5,8	21,2	4,4
3	72/M	Left	V3	BA	5,7	19,1	9,1
4	57/M	Left	V2-3	VA	8,7	22,1	14,8
5	78/W	Left	V1-2	VA+AICA	4,5	17,2	10,2
6	74/M	Left	V1-2	BA	9,1	21,7	14,8
7	62/M	Left	V1-2	BA+SCA	6,4	14,4	9,4

BA: basilar artery; VA: vertebral artery

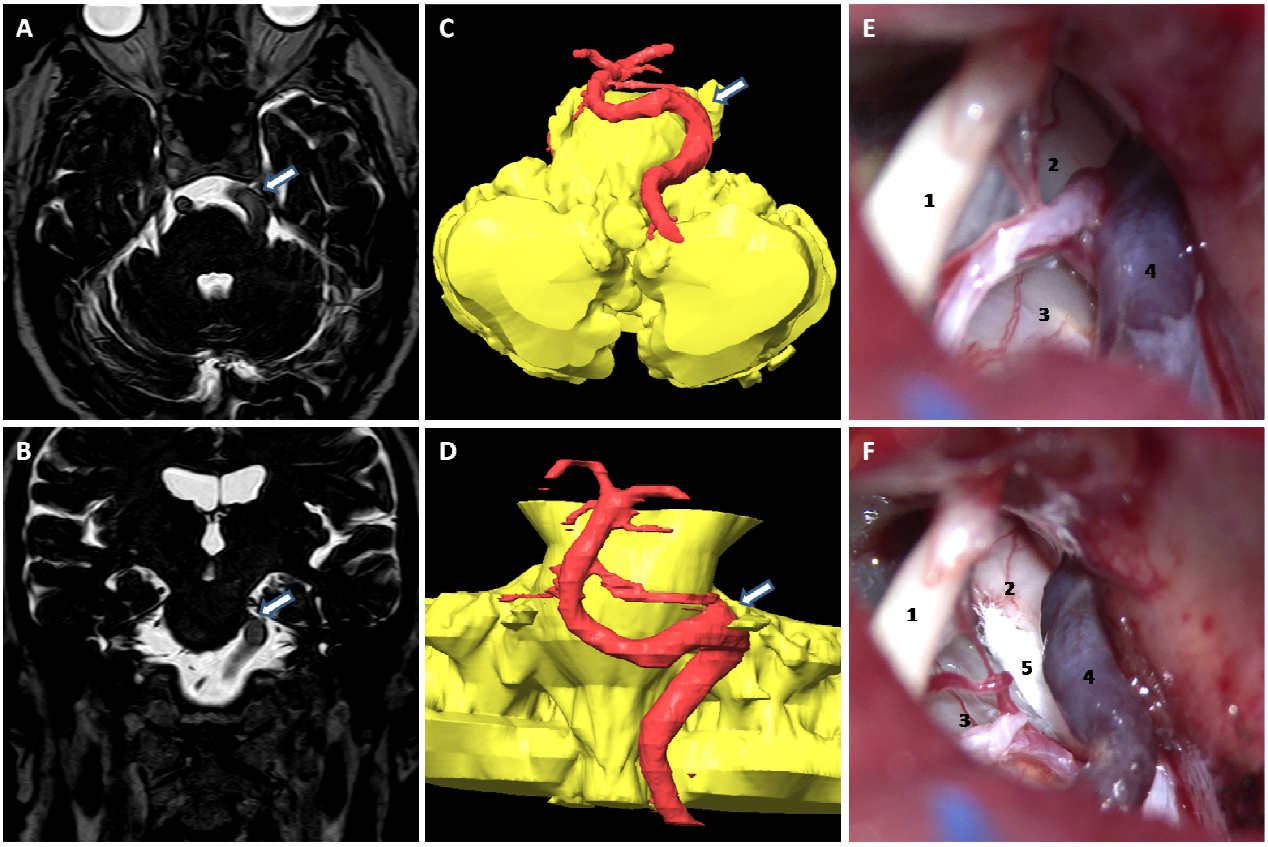

Fig. 2. (A,B) MRI DRIVE coronal and axial sequences depicting a tortuous and dilated basilar artery occupying the left cerebello-pontine angle and compressing the caudal aspect of the trigeminal nerve (arrow). **(C,D)** Three-dimensional reconstructions showing how the basilar artery arising from a dominant left vertebral artery impinges on the left trigeminal nerve. **(E)** Microsurgical image of the left cerebello-pontine angle as seen through a retrosigmoid approach demonstrating a compression of the left trigeminal nerve (2) by the basilar artery (3) and its close relation to the seventh and eighth cranial nerve complex (1) as well as the superior petrosal vein (4). **(F)** Microsurgical view after microvascular decompression with a Teflon pad (5).

in 3 cases. In all patients the vertebrobasilar system showed a tortuous course that exceeded the lateral limits of the clivus and created a loop in the cerebello-pontine cistern where it contacted and compressed the cisternal segment of the trigeminal nerve (Fig. 2 A-D). The compression of the trigeminal nerve was caused by the ipsilateral VA in three cases and by the BA in the remaining four cases. Measurements from axial a coronal DRIVE sequences showed a mean maximal diameter of the basilar artery of 6.5 mm (5-9.1 mm) and a mean lateral deviation of 19.3 mm (14.4-22.1 mm). The mean elevation of the basilar tip above the dorsum sellae was 10.5 mm (4.4-14.8 mm) on sagittal T1-weighted sequences (Table 1).

Microsurgical exploration of the cerebello-pontine angle cistern confirmed the existence of a compression by a VBDE. In two cases a combined compression was found, caused by the VA and the anterior inferior cerebellar artery (AICA) in one case, and by the BA and by the superior cerebellar artery (SCA) in the other case. In all seven patients the trigeminal nerve could be dissected away from the artery and decompressed with interposition of Teflon®. In one case the vascular contact was considered only mild, and a partial sensory rhizotomy was added (Fig. 2 E-F).

All patients experienced immediate postoperative pain relief. At last follow-up six of the patients (85.7%) remained pain-free and without medication. One patient had a recurrence of symptoms after 7 years and required further treatment with percutaneous balloon compression. There were no mortality or permanent neurological deficits. One patient developed a postoperative cerebrospinal fluid fistula that was satisfactorily handled with a transient lumbar drain.

DISCUSSION

The course of the BA lies in the midline or paramedian but medial to the margin of the sphenoid clivus or dorsum sellae in 98% of the people (Smoker et al., 1986). The BA divides into both posterior cerebral arteries in the interpeduncular fossa at the level of the superior aspect of the dorsum sellae (Yu et al., 1982). The average diameter of the BA based on autopsy findings and on angiographic measurements lies between 3.1 and 3.6mm (Rai et al., 2013; Wollschlaeger et al., 1967). VBDE has been defined as an abnormal dilation and elongation of the vertebrobasilar system with a maximal basilar diameter over 4.5 mm and a course exceeding laterally beyond the limits of the clivus or the dorsum sellae or cranially above the suprasellar cistern (Gautier et al., 1988; Smoker et al., 1986; Ubogu et al., 2004). The mechanism of development of VBDE is still unclear. Some series have described a loss of elastin in the elastic layer of the BA (Thiex et al., 2006). Together with the occurrence of VBDE in younger patients, this suggests a congenital vasculopathy. But the association of VBDE with hypertension and atherosclerotic changes, as well as the significant prevalence of other cardiovascular risk factors such as smoking, obesity, dyslipidemia and diabetes mellitus in individuals with VBDE, point at a degenerative process (Ikeda et al., 2010; Nishizaki et al., 1986). A combined hypothesis proposes an unequal development of the longitudinal neural arteries, which are the embryological precursors of the BA. The resulting asymmetric flow in the vertebrobasilar system would cause over time degeneration of the elastic layer, atherosclerosis and a growing elongation and tortuosity of the vessel (Vasovic et al., 2012).

The natural history of VBDE is widely unknown, but it is suspected that most individuals are asymptomatic (Ikeda et al., 2010; Vasovic et al., 2012; Yuan et al., 2014). The most common clinical manifestation of this condition is ischemic stroke, which was observed in 36% of the cases of a prospectively followed series of 156 patients. Cerebrovascular events are also de most frequent cause of recurrent symptoms occurring in up to 48% of the patients (Passero et al., 2008). Although far more uncommon, bleeding in form of a subarachnoid or intracerebral hemorrhage also occurred in this series. Compressive symptoms have been described for cranial nerves III to XII, as well as for the brainstem (Herpers et al., 1983; Passero et al., 2001; Vasovic et al., 2012; Yuan et al., 2014). By far the most frequently compressed structures are the fifth and seventh cranial nerves, which may cause TN and hemifacial spasm (El- Ghandour, 2010; Passero et al., 2008). Obstructive hydrocephalus is an extremely rare manifestation of VBDE and may be caused by an actual compression the third ventricle and the aqueduct of Silvius or by a hydrodynamic effect as a result of the pulsatility of the VBDE transmitted to the bottom of the third ventricle and Monro foramina generating a “water hammer” effect (Siddiqui et al., 2008; Thiex et al., 2006; Yuan et al., 2014).

The diagnosis of VBDE was initially based on CT scan and digital substraction angiography (Smoker et al., 1986). The introduction of MRI in the diagnostic protocol of trigeminal neuralgia to rule out painful trigeminal neuropathies caused by posterior fossa tumors, multiple sclerosis or vascular malformations enhanced the diagnosis of VBDE as a cause of neurovascular compression (Cruccu et al., 2008; Harsh et al., 1991; Kirsch et al., 1996). More recently the introduction of high resolution three-dimensional T2-weighted MRI sequences and three-dimensional MR angiography has provided a precise tool for the preoperative assessment of the neurovascular anatomy in the posterior fossa cisterns (Becker et al., 2008; Borges et al., 2010). Three dimensional T2-weighted MRI like the DRIVE sequences em-

ployed in our series show a detailed picture of vascular and neural structures which appear hypointense and are clearly outlined against the hyperintense signal of cerebrospinal fluid. These MRI sequences have proved a high sensibility ranging from 76% to 97% and specificity between 75 and 95% for the diagnosis of neurovascular compression of the trigeminal nerve in patients with TN (Anderson et al., 2006; Han-Bing et al., 2010; Leal et al., 2010; Maarbjerg et al., 2015; Vergani et al., 2011). Concerning VBDE, although the diameter of the dilated vertebrobasilar system makes the diagnosis straight forward, it is important to obtain an accurate view of the anatomical relationship between the tortuous vertebrobasilar system and the trigeminal nerve. First, because the compression may lie caudal, medial or cranial to the nerve (Yang et al., 2012). Second, because, as was seen in two of our cases, other vessels may be involved too in a multiple compression of the trigeminal nerve (Linskey et al., 1994). Third, because a simultaneous compression of the ipsilateral facial nerve causing hemifacial spasm has been described in 16 to 40% of the cases (El-Ghandour, 2010; Linskey et al., 1994; Ma et al., 2013). Fourth, because the size and firm position of the tortuous, dilated and often atherosclerotic vertebrobasilar system requires a precise knowledge of the spatial relations of vascular, neural, dural and bony elements in and around the cerebello-pontine angle to design the surgical strategy when attempting a MVD in a case of VBDE. Therefore the three-dimensional reconstruction employed in the present series may contribute to illustrate this particular anatomy.

Several authors have pointed out the technical difficulty of MVD in cases of VBDE, mainly because of the large caliber and rigidity of the partially calcified basilar artery (Banczerowski et al., 2014; Linskey et al., 1994; Lye, 1986). An increased risk of neurological complications has been described due to the need of extensive manipulation of posterior fossa neurovascular structures in order to achieve a satisfactory decompression (Linskey et al., 1994; Miyazaki et al., 1987). For these reasons, some advocate for alternative surgical treatments such as percutaneous procedures or radiosurgery in these cases. Nevertheless recent surgical series of MVD in TN caused by VBDE report pain outcomes and complication rates similar to those of MVD for TN due to other arterial compressions (El-Ghandour, 2010; Ma et al., 2013; Yang et al., 2012).

Conclusions

VBDE is a rare cause of TN that demands a precise knowledge of the neurovascular relations in the cerebello-pontine angle. Our experience supports the choice of MVD as the first line treatment for medically refractory TN caused by VBDE.

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