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Developmental venous anomaly causing trigeminal neuralgia

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Trigeminal neuralgia (TN) is a syndrome characterized by paroxysms of facial pain affecting mainly middle and old-aged patients[1,2]. Although TN's pathogenesis is not fully understood, there is increasing evidence that TN is caused by demyelination of trigeminal sensory fibres[1] mostly owing to an arterial neurovascular compression on the root entry zone (REZ) of the trigeminal nerve[1-5]. Veins, on the other hand, can be regarded as the only offending vessel in up to 13% of the patients[1,4,6].

Other unusual causes of TN include intrinsic brainstem lesions or even compression of the trigeminal nerve root by tumors, in which the compression can occur by the tumor itself, by an interposed blood vessel or by a distortion of the posterior fossa contents displacing the nerve root towards blood vessels or the skull base[1]. Nonetheless, in a small number of patients, the pathogenesis remains to be determined[1].

Although vein compression is usually accepted in TN's pathogenesis, either alone or in combination to arterial compression, developmental venous anomaly (DVA) causing TN is seldom reported with 17 cases described to date[2,3,5,7-15]. A patient with a classical longstanding right frontal division TN caused by a DVA is reported emphasizing the clinical features and management of such condition.

CASE

A 57-year-old man sought treatment for a longstanding classical right frontal division TN. The symptoms were first presented for seven years having benign evolution. The patient showed years of remittent symptoms while developing pain for just a few days. The neurological ex-

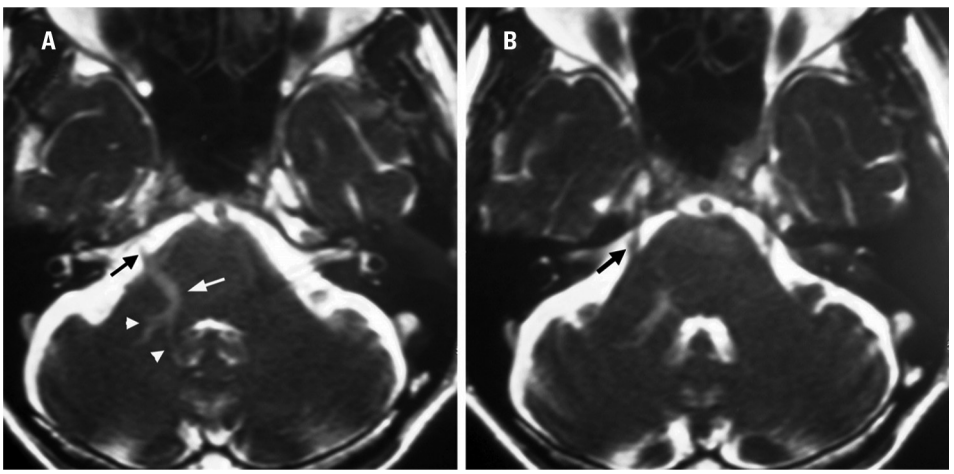

Fig 1. Sequential axial 3D CISS (three-dimensional constructive interference in steady state) MRI (1 mm width) reveals a large right transparenchymal vein (white arrow) and caput medusae (white arrow heads) [A] as well as the course of the trigeminal nerve (black arrow) and its relationships to the developmental venous anomaly (DVA) within the brachium pontis [A and B].

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NEURALGIA DO TRIGÉMIO CAUSADA POR ANGIOMA VENOSO

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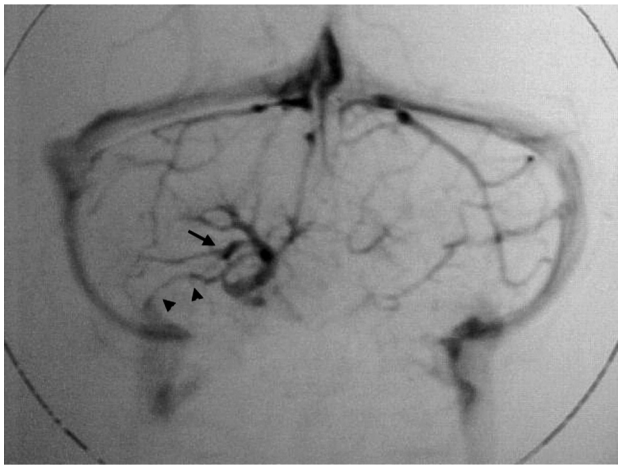

Fig 2. Digital subtraction angiography (DSA) (vertebral artery venous phase) in frontal projection showing a DVA on the right side draining into the petrosal venous system (black arrow) and jugular bulb (black arrow heads).

amination was unremarkable. Magnetic resonance imaging (MRI) of the brain revealed a large transparenchymal draining vein together with caput medusae involving the right brachium pontis thereby contacting the trigeminal REZ (Fig 1). Digital subtraction angiography (DSA) showed the typical angiographic features of DVA, namely the collection of dilated medullary veins draining into a single large draining vein. This vein was located in the cerebellopontine angle ultimately draining into the superior petrosal sinus and right jugular bulb (Fig 2). The symptoms were well controlled with a low dose of carbamazepine schema staying without medication during the remission period. The patient has been handled conservatively ever since. The Ethics Committee from the Hospital Universitário Pedro Ernesto approved this publication.

DISCUSSION

DVA, also known as venous angioma, is formally the most common type of intracranial vascular malformation[15]. The term DVA was initially postulated by Lasjaunias et al.[16] following a careful analysis of DVA's morphological and clinical details. Lasjaunias et al.[16] proposed that DVAs are anatomical variants because they consistently drain normal cerebral tissue within a normal arterial territory, are typically associated with absence of normal venous drainage pathways, demonstrate opacification in the same time as the normal veins at DSA and their surgical removal or ligature may lead to serious ischemic complications.

DVAs are usually incidental but have been controversially associated with cerebellar symptoms, headache, epilepsy, hydrocephalus, haemorrhage, and spontaneous thrombosis[3,7,8,11,13,15,16]. The true incidence of clinical symptoms remains unknown and it does not seem to be associated with a specific clinical presentation[14].

Apart from the clinical controversies[8,14], DVAs should be considered symptomatic when their location match with the patient's clinical presentation[15]. In this way, TN is possibly the only symptom directly attributable to DVA owing to contact or compression of the trigeminal REZ by the dilated vessel or even to irritation of the spinal trigeminal nucleus[2,3,5,7,8,11-14]. To the best of our knowledge, 17 cases of DVA causing TN have been reported to date[2,3,5,7-15]. Three cases were excluded from our analysis due to lack of information provided by the original authors[11,15] (Table).

When associated with a DVA, TN occurs in a younger population than usual (median age 44 years)[2,3,5]. It is worth mentioning that in all patients affected of TN from DVA compression a deep venous drainage was encountered, either predominantly to the superior petrosal sinus or incidentally to the vein of Galen, the jugular bulb or even to the occipital sinus. Like in ordinary TN[1,6], these patients commonly complain about pain for years (up to 47 years) before being considered for surgery (Table).

DVA management remains controversial[3,12]. Generally, conservative treatment for DVA has been advocated by many authors due to the unacceptable high morbidity and mortality rates related to its resection[15]. In patients affected of TN caused by DVA, conservative treatment has been performed rendering partial[2,12,14] or complete pain relief, as the presented case.

For patients with progressive symptoms or in life-threatening situations, the surgical treatment should be considered for posterior fossa DVA[16]. Surgery was performed in 10 of 15 patients reviewed for DVA causing TN[2,3,5,7-10,13] (Table). Microvascular decompression (MVD) was performed in all patients but one, in which a glycerol rhizotomy resulted in complete pain relief. DVA ligature was performed in 5 patients rendering complete pain relief in 4 patients[2,8,10] and death in one patient due to cerebellar infarction[13]. Besides to vein ligature, MVD with DVA reposition was accomplished in four patients[3,5,7,9], all of them reaching complete pain relief.

Currently, MVD is the surgical treatment of choice for TN owing to good long-term results together with low mortality rates (up to 1%)[1,2,6,17]. Considering that TN is not a lethal condition *per se*[17] and the unacceptable higher mortality rate (20%) following DVA ligature in comparison to reposition, we believe that DVA ligature for TN treatment should be no longer recommended. Alternatively to MVD, percutaneous procedures, namely radiofrequency thermorhizotomy and percutaneous balloon compression may also play an important role in TN management[1] providing good long-term relief of pain, while glycerol rhizotomy should be limited to selected cases due to higher pain recurrence rates[17].

Table 1. Summary of reported cases of trigeminal neuralgia caused by developmental venous anomaly (DVA).*

Author Year	Age (yr) Gender	Time of symptoms	Distribution of pain, side	Diagnostic work–up	DVA Location	Venous drainage	Treatment	Outcome	Associated VM
Pelz et al. 1983[7]	34 M	4 yr	V2, R	CT, DSA	Brachium pontis	Deep (SPS, JB)	MVD, vein reposition (previous neurectomy)	Complete pain relief	–
Martin et al. 1984[8]	–	–	–	CT, DSA	Brachium pontis	Deep (SPS)	MVD, vein ligature	Complete pain relief	–
Isu et al. 1985[9]	35 M	–	–	CT, DSA	–	Deep (SPS)	MVD, vein and artery reposition (SCA)	Complete pain relief	–
Trost et al. 1987[10]	23 M	4 mo	V3, L	CT, DSA	Brachium pontis	Deep (SPS)	MVD, vein ligature	Complete pain relief, transient facial paresis, V3 hypoesthesia	–
Raveau et al. 1992[12]	45 F	15 mo	V2, L	CT, MRI, DSA	Cerebellar hemisphere, posterior extension to brachium pontis	Deep (OS)	Conservative	Partial pain relief	2 posterior fossa cavernomas
Mori et al. 1994[13]	38 F	1 yr	NR, R	CT, MRI, DSA	Cerebellar hemisphere	Deep (SPS)	MVD, vein ligature	Dead	–
Nagata et al. 1995[5]	35 M	6 yr	V3, L	CT, MRI, DSA	Brachium pontis	Deep (SPS)	MVD, vein and artery reposition (AICA)	Complete pain relief	–
Küker et al. 1997[14]	62 M	47 yr	V1 / V2, L	MRI, DSA	Cerebellar hemisphere, pons	Deep (VG)	Conservative (previous op)	–	–
Korinth et al. 2002[3]	35 F	13 yr	V2 / V3, L	MRI, DSA	Brachium pontis	Deep (SPS, VG)	MVD, reposition	Complete pain relief	–
Peterson et al., 2002[2]	58 M	35 yr	–	CT	Brachium pontis	Deep	MVD, artery reposition (SCA), vein ligature	Complete pain relief	–
	43 F	10 yr	–	MRI	Brachium pontis	Deep	Glycerol Rhz	Complete pain relief	–
	74 M	22 yr	–	MRI	Brachium pontis	Deep	Conservative	Partial pain relief	–
	70 F	24 yr	–	CT, MRI	Brachium pontis, posterior pons	Deep	MVD, artery reposition, vein ligature (2 previous Glycerol Rhz, 1 MVD – vein and artery)	Complete pain relief	–
	59 M	12 yr	–	CT	Brachium pontis, posterior pons	Deep	Conservative	Partial pain relief	–
Present study	57 M	7 yr	V1, R	MRI, DSA	Brachium pontis	Deep (SPS, JB)	Conservative	Complete pain relief	–

M: male; F: female; yr: years; mo: months; R: right; L: left; CT: computed tomography; MRI: magnetic resonance imaging ; DSA: digital subtraction angiography; SPS: superior petrosal sinus; JB: jugular bulb; OS: occipital sinus; VG: vein of Galen; MVD: microvascular decompression; SCA: superior cerebellar artery; AICA: anterior inferior cerebellar artery; Rhz: rhizotomy; VM: vascular malformations.

In conclusion, DVA is a rare cause of TN occurring in a younger population than usual. Preoperative diagnosis of DVA may contribute for the decision making concerning TN management. Conservative treatment is a reasonable option rendering satisfactory pain control due to benign evolution of the disease while maintaining the functional status. When indicated, the surgical treatment in these given patients should consider MVD of the trigeminal nerve with vein repositioning or percutaneous procedures rather than vein coagulation in order to avoid ischemic complications.

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