Developmental Venous Anomaly Responsible for Hemifacial Spasm

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SUMMARY – *Hemifacial spasm (HFS) is a facial movement disorder characterized by involuntary, unilateral and intermittent contractions of the facial muscles. It is one of the syndromes related to neurovascular conflict, first described by Jannetta et al. in 1979. Typically, HFS is due to pulsatile compression by the anterior inferior cerebellar artery. We describe a rare case of left developmental venous anomaly in a 59-year-old man referred to us with a six-month history of left-sided HFS. We performed an MR study of the brain and cerebellopontine angles, which demonstrated a compression of the ipsilateral facial nerve by the developmental venous anomaly.*

Introduction

Hemifacial spasm (HFS) is a neuromuscular disorder characterized by frequent and involuntary contractions of the facial muscles causing physical discomfort and embarrassment, and impairing quality life.

The major etiology of hemifacial spasm is neurovascular conflict that is the pulsatile compression of a vascular structure within a few millimetres from the origin of the facial nerve, name “root entry/exit zone” (REZ)¹. Persistence of the phenomenon leads to demyelination of the nerve. The REZ is near the Obersteiner–Redlich zone, a transition area 2-3 millimetres long on the nerve root, between central and peripheral axonal myelination, that is between oligodendrocytes cells and Schwann cells. It is situated at the nerve’s detachment from the pons, within the most proximal few millimetres of the nerve entering/exiting the brainstem; this zone is the most susceptible to vascular compression².

The facial nerve emerges to the brainstem surface (RExP) from the pontomedullary sulcus at the upper edge of the supraolivary fossa and strongly adheres to the surface of the pons for 8-10 mm (AS) before separating from the brainstem (RDP). The Obersteiner-Redlich

zone (TZ) is situated at the RDP and extends ~ 2 mm further distally.

The more proximal segments of the facial nerve root, with oligodendrocyte-derived myelin, may also be susceptible to the pathophysiological influences of neurovascular compression causing HFS³.

Campos-Benitez and Kaufmann described the facial root exit zone (REZ) as an area of ~ 10 mm extending from the RExP to the TZ¹. According to their results the incidence of vascular compression of the facial nerve is caused by the anterior inferior cerebellar artery (43%), posterior inferior cerebellar artery (31%), vertebral artery (23%), large vein (3%), and multiple compressing vessels (38%)². The affected locations are at the emergence of the facial nerve to the brainstem surface (RExP) in 10%, between the facial nerve and the surface of the pons (AS) in 64%, where the nerve separates from the brainstem (RDP) in 22%, and its distal portion, where the nerve has no contact with brainstem (CP) in 3%⁴.

HFS caused by developmental venous anomaly (DVA) is a very rare occurrence. DVA is the most frequently found cerebral vascular malformation constituting approximately 60% of all vascular lesions⁵.

The main etiology suggested for this entity is

Figure 1 Axial FSE T2 at the posterior cranial fossa demonstrates the tortuous course of the fourth segment of the vertebral artery with an abnormal vessel originating on the left cerebellar hemisphere and becoming extra-axial.

Figure 2 Coronal FLAIR demonstrates a hyperintense lesion on the left paramedian cerebellum with some hyperintense radial prolongations consistent with a caput medusae appearance.

an embryologic accident that results in either arrested formation or thrombosis of the developing venous drainage of the specific region⁵. This leads to a compensatory mechanism in which embryologic medullary venules persist and cluster in a large draining vein.

Clinical Case

A 59-year-old man presented with a six-month history of left-sided mild and quite intermittent hemifacial spasm initially involving the orbicularis oculi muscle and then gradually progressing in severity and frequency and spreading downward to other muscles innervated by the ipsilateral facial nerve. Finally the muscles were involved with impairment of facial expression structures, as well as the peri-orbital region and the ipsilateral oral angle. There were no associated sensory deficits, pain, weakness or other neurological manifestations including ocular motor palsy and hearing disturbance. The spasms were spontaneous and frequent and were exacerbated by voluntary facial movements, psychological stress, fatigue, anxiety or head movements. On the basis of a pathophysiologic explanation of hemifacial spasm, according to which an abnormal cross-transmission circuit between demyelinated

facial nerve fibers and the sympathetic nerve fibers near the offending vessel wall, causes a facial muscle response, while the offending vessel wall is electrically stimulated. The presence of the stapedial reflexes both during the acute phase and during the comfort phases confirmed this hypothesis. The reflex morphology is characterized by decreased amplitude and increased impedance. Electrical stimulation of one branch of the facial nerve on the affected side elicited a delayed response from the muscles supplied by other branches. This is called an “abnormal muscle response” and has been documented only in patients with hemifacial spasm, so it is used for diagnosis and intraoperative monitoring during microvascular decompression surgery. To confirm the diagnosis we performed an MR study with 1.5 T unit (GE Signa excite HD). In addition to standard brain study (FSE T2-w, SE T1-w, FLAIR, EPI, 3DT1-FSPGR after contrast administration), we also performed 3D-FIESTA and 3D-TOF sequences. The T2-w images demonstrated a flow void located along the left surface of medulla oblongata (Figure 1). A small oval signal alteration was also noted in the left cerebellar peduncle, hyperintense in T2-w and FLAIR (Figure 2) and hypointense in T2*-w, compatible with capillary telangiectasia (Figure 3). No associated parenchymal abnormalities were

Figure 3 Axial GE T2* showed a small hypointense lesion in the left cerebellar peduncle consistent with capillary telangiectasia.

Figure 5 Axial 3D-FIESTA at the same level as Figure 4.

Figure 4 Axial enhanced T1 3D-FSPGR shows the typical appearance of DVA with "caput medusae" associated with a dilated trunk with oblique course from the left cerebellum white matter to the sigmoid sinus.

Figure 6 Axial 3D-FIESTA shows a compression by the dilated trunk of DVA (arrowhead) of the left ventrolateral surface of the medulla oblongata with displacement of IX, X and XI nerves (arrow).

Figure 7 Axial 3D-FIESTA at the level of the left VII (large arrow) and VIII REZ (small arrow) shows the passage of the dilated vessel directed to the petrosal sinus through the two cranial nerves (transparent arrowhead).

Figure 8 Parasagittal post-processing 3D-FIESTA best shows the course of the abnormal vessel (arrowhead) between the VII (large arrows) and VII (small arrows) REZs.

Figure 9 Axial 3D-FIESTA at the level of left petrosal sinus (arrowhead) running over the cisternal portion of the V nerve, immediately before entering Meckel's cave.

Figure 10 Coronal MPR of 3D-FIESTA shows the end of the abnormal vessel on the left petrosal sinus (long arrow), without compression on the V nerve (short arrow).

found. The 3D-T1 FSPGR after contrast media administration and the 3D-FIESTA showed the nature of the vascular lesion more accurately with typical “caput medusa” appearance associated with a large vessel becoming extra-parenchymal: findings most consistent for DVA (Figures 4 and 5). By analysing 3D-FIESTA sequences we noted that the large vessels were directed downward toward the left sigmoid sinus and at the same level it was possible to highlight other smaller enlarged vessels moving upward to reach the left upper petrosal sinus. Before reaching the sigmoid sinus, the first vessel caused compression at the sulcus of the mixed nerves (Figure 6). The second vessel went back along the edge of the ventrolateral brainstem, up to reach the petrosal sinus; along its course the vessel passed between the REZ of VII and VIII cranial nerves causing longitudinal compression on the left VII cranial nerve (Figures 7 and 8). Finally this vessel reached the petrosal sinus making a loop around the cisternal portion of the left trigeminal nerve (Figures 9 and 10). In agreement with the patient's symptoms, a diagnosis of neurovascular conflict was established at the level of VII cranial nerve responsible for HFS. For a higher diagnostic evaluation the patient was offered angiographic study of the brain but he declined this procedure. In agreement with the patient, surgery was refused and only a medical treatment given administering 600 mg/day carbamazepine in a divided dose. Carbamazepine, an anticonvulsant, is used to manage severe muscle spasms and provide analgesia and mild sedation.

Discussion

Hemifacial spasm is a facial movement disorder characterized by involuntary, unilateral and intermittent twitching of muscles innervated by the facial nerve⁶. The same symptoms are common to many disorders such as tumours, arachnoid cysts, facial myokymia, blepharospasm and facial tic. Ipsilateral neurovascular compression is one of the commonest causes of hemifacial spasm^{7,8}.

Arterial compression of the nerve, described as the most common factor rather than venous compression, occurs upon the root exit zone of the facial nerve^{1,9}. The limits of this zone have never been clearly defined. Jannetta et al. identified the localization of neurovascular compression as the most proximal portion of the affected nerve in hemifacial spasm. They emphasized that these cranial nerves are most susceptible to pulsatile vascular contact at their root entry/exit zone, and therefore this is the area of interest in performing microvascular decompression treatment¹⁰.

Neurovascular compression can be localized in the following four anatomical portions of the facial nerve: root exit point (REXP), attached segment (AS), root detachment (RDP), the point corresponding to the transitional zone, and the distal cisternal portion (CP)⁴.

The severity of compression has been defined as follows: mild if there is contact without nerve indentation; moderate if facial nerve indentation is present; and severe in the case of an eventual deviation of the nerve course¹¹.

Vascular compression of the facial nerve root

entry zone (REZ) is due mainly to vessels, usually arteries, generally a branch of the anterior inferior cerebellar artery (AICA), posterior inferior cerebellar artery (PICA), or vertebral artery. Like Campos-Benitez and Kaufmann, Chung et al. evaluated the percentage distribution of aetiologies due to neurovascular compression: 45.9% AICA anomalies, 34.8% PICA anomalies, 12.5% vertebral artery anomalies and 6.8 % multiple vessel anomalies. Instead, Sindou reported multiple vessel anomalies in 40% of cases suggesting that unsatisfactory results of neurosurgery could be ensue from non-recognition of all these multiple anomalies (because of their rarity, as in our case)^{12,13}. Infratentorial intra-axial intrinsic lesions can cause HFS: for example gliomas, gangliomas or vascular anomalies through compression of the facial nucleus or nerve fibers, instead of hamartoma or neuroectodermal tumours which behave as intralesional epileptogenesis¹³.

The literature describes the frequency of site of onset: the orbicularis oculi muscle in 90%, the cheek in 11% and the perioral region in 10% of cases. Over months to years, the spasms spread gradually to other muscles innervated by the ipsilateral facial nerve. Tonic spasm is generally accompanied by twitching and synkinesis¹⁴. Our patient's symptoms followed the same chronological course, from the upper zone of the face downwards. In recent years improvements to MRI techniques have led to more frequent identification of vascular compression at the root exit zone of the facial nerve, even though a normal vascular MRI pattern does not rule out compression¹⁵.

In our case the infratentorial DVA was the cause of HFS. The term DVA was coined by Lasjaunias and now is widely used as a synonym for venous angioma, cerebral venous malformation or cerebral venous medullary malformation⁶. DVAs are congenital benign anatomic variations with angiogenically mature venous walls that lack arterial or capillary elements: it is hypothesized that they result from a focal arrest of venous development and retention of primitive medullary veins that drain into a single enlarged transcortical or subependymal collector vein which plays a compensatory role in relation to the absence of a normal cortical venous network¹⁶. These venous channels coalesce into a normal venous outflow tract and a focal stenosis may occasionally be noted as it enters the adjacent dural sinus predisposing to venous hypertension or thrombosis, or both. However the haemorrhagic risk for DVA not

associated with vascular malformations is low, but if it drains into the petrosal sinus, the risk increases in relation to the major pressure gradient¹⁷. When acute haemorrhage is observed with a DVA, it is likely to be associated with a cavernous malformation (CM). Most patients have no symptoms and frequently DVAs are found incidentally during a neuroimaging investigation. In symptomatic patients the most common symptom is a headache or seizure, especially when associated with a CM or hippocampal sclerosis. Other rare symptoms are linked to direct compression of cranial nerves, such as the trigeminal, facial and/or cochleovestibular nerves. The literature also reports cases of hydrocephalus by compression of the aqueducts mesencephali¹⁸. In our patient the DVA did not reveal haemorrhage, thrombosis and/or parenchymal abnormalities. A more recent investigation reported alterations in up to 65% of DVAs, like regional atrophy, the most frequent (30%), white matter lesions (28%) and dystrophic calcification¹⁹.

In our patient the standard MR study demonstrated an enlarged infratentorial DVA and according to the patient's symptomatology and MR findings, we also performed a detailed study of the cerebellopontine angle (CPA), comprising an axial 3D-TOF sequence for the evaluation of intracranial vessels and 3D-FIESTA or CISS (also known as a sequence to free precession of the steady state) for evaluation of the neurovascular relationship, especially at CPA level. These sequences are frequently used for the evaluation of patients with neurovascular compression. In our patient the DVA have an oblique cranio-caudal course from the caput medusa on the left cerebellar white matter to the sigmoidal sinus, causing compression at the level of the sulcus of mixed nerves (IX, X and XI cranial nerves). In addition, an enlarged vessel originated from the caudal portion of the DVA running back along the edge of the ventrolateral brainstem up to reach the petrosal sinus; along its course the vessel determined a longitudinal compression with REZ of the left VII cranial nerve. Therefore in our case the neurovascular conflict and the related syndrome were sustained by DVA.

A typical MR and X-ray finding of DVA is the radially arranged, anomalous veins (caput medusa appearance) that cover a centrally located dilated trunk, which drains into a dural sinus. Unenhanced T1-w and T2-w sequences may demonstrate flow voids and phase-shift artifacts. After contrast media administration

Figure 11 Axial and sagittal MIP of a T1-enhanced 3D FSPGR sequence shows the typical “caput medusa” appearance (arrow-heads), the dilated trunk (large arrow) and the abnormal vessel reaching the left petrosal sinus (small arrow).

enhancement of the medullary veins and venous collector is observed. The cluster of veins in DVAs has a spoke-wheel appearance referred to as “caput medusa”: enhanced T1-w FSPGR is the sequence of choice for depiction of DVAs. On X-ray, the caput medusa is visualized during the early to middle venous phase²⁰. X-ray is no longer performed at our centre for the diagnosis of DVA because MRI allows a pseudo-angiographic evaluation of these abnormalities, especially with enhanced three-dimensional sequences (Figure 11). The surrounding parenchyma does not normally show signal alterations, but signal changes like hyperintense signal on T1-w or hypointense signal in T2*-w, could suggest a bleeding lesion. Modern imaging will define the exact localization (intra- or extra-axial) to clear up the irritating symptomatology and guide the choice of appropriate treatment (microvascular decompression or wait and see)¹⁰.

Medical treatment of this condition is recommended in patients with mild-moderate symptoms. Surgical management of HFS is reserved

for patients with severe symptoms or patients no longer responsive to medical therapy.

Surgery generally has an excellent outcome, but botulinum toxin may work even in the case of large vessel anomalies, even though the results are not definitive²¹. Microvascular decompression, first described by Jannetta et al., is an effective treatment option for HFS caused by such vascular compressions. In cases of hemifacial spasm three varieties of neurovascular compression are described: veins, elongated VAs, and rostrally situated vessels. Venous contacts are not routinely treated in cases of HFS instead of typical neurovascular compression by an artery (1). It is important to remember that vein preservation is a key point for the treatment of hemifacial spasm and prevention of any possibility of venous infarction; the vein must be carefully dissected away from the exit zone of the facial nerve. Small pieces of shredded Teflon are placed between the nerve and the vessel¹⁰. The lesion can be managed conservatively in asymptomatic or mildly symptomatic patients²².

In conclusion this article describes a rare case of HFS caused by infratentorial DVA. It is important that, in these cases, a thorough work-up is always performed for definitive diagnosis

and appropriate management. Careful reading of MR images combining specific sequences, such as 3D-TOF with 3D-FIESTA, is essential to predict whether vascular compression exists.

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