Hemifacial Spasm Associated with Chiari Type I Malformation: Surgical Considerations and Case Report

Espasmo Hemifacial Associado a Malformação de Chiari Tipo I: Considerações Cirúrgicas e Relato de Caso

Carlos Augusto Ferreira Lobão1 Ulysses de Oliveira Sousa2 Diego Arthur Castro Cabral3 Fernanda Myllena Sousa Campos3

1 Division of Neurosurgery, Department of Neuroscience, Hospital Porto Dias, Belém, PA, Brazil
2 Department of Neurosurgery, Hospital Saúde da Mulher, Belém, PA, Brazil
3 Institute of Health Sciences, Faculdade de Medicina, Universidade Federal do Pará, Belém, PA, Brazil

Arq Bras Neurocir

Address for correspondence Diego Arthur Castro Cabral, Medical Students, Universidade Federal do Pará, Belém, Pará, Brasil (e-mail: diego.arthur.castro@gmail.com).

Abstract

Hemifacial spasm (HS) is a movement disorder characterized by paroxysmal and irregular contractions of the muscles innervated by the facial nerve. Chiari malformation type I (CM I) is a congenital disease characterized by caudal migration of the cerebellar tonsils, and surgical decompression of foramen magnum structures has been used for treatment. The association of HS with CM I is rare, and its pathophysiology and therapeutics are speculative. There are only a few cases reported in the literature concerning this association. The decompression of the posterior fossa for the treatment of CM I has been reported to relieve the symptoms of HS, suggesting a relation between these diseases. However, the possible complications of posterior fossa surgery cannot be underrated. We report the case of a 66-year-old patient, in ambulatory follow-up due to right HS, no longer responding to botulinum toxin treatment. Magnetic resonance imaging (MRI) of the skull revealed compression of the facial nerve and CM I. The patient underwent surgery for HS by neurovascular microdecompression of the facial nerve via right lateral suboccipital craniectomy, but presented significant clinical worsening in the postoperative period even though the cerebellum edema related to surgical manipulation was mild. Due to the clinical worsening, the patient underwent a median suboccipital craniectomy with decompression of the foramen magnum structures. After this second surgery, the patient had progressive improvement and was discharged from the hospital for ambulatory care.
Introduction

Hemifacial spasm (HS) is a movement disorder characterized by paroxysmal and irregular contractions of the muscles innervated by the facial nerve. In most cases, it is associated with neurovascular compressions around the facial nerve root exit zone.\(^1\)\(^2\) Chiari malformation type I (CM I) is a congenital disease characterized by caudal migration of the cerebellar tonsils that causes disproportion between the contents and volume of the posterior fossa and compression of structures at the foramen magnum level.\(^3\)\(^4\) Therefore, the decompression of foramen magnum structures is widely accepted as the treatment of choice for symptomatic CM I and can be performed using several techniques.\(^5\)\(^6\)\(^7\)\(^8\) Hemifacial spasm associated with CM I is rare, with few cases reported in the literature. The exact pathophysiology of the association between the two diseases remains speculative until the present. However, it was observed that posterior fossa decompression for the treatment of CM I lead to the relief of the symptoms of HS, being considered as the primary procedure to be applied in these patients. The amelioration of HS symptoms seems to indicate a relationship between the geneses of the two pathologies.\(^3\) Nevertheless, complications of posterior fossa surgery are rarely highlighted in the literature. Here, we report the case of a patient with HS associated with CM I without craniovertebral instability that was treated with posterior fossa decompression, drawing attention to the possible complications associated with this procedure and critically reviewing the factors that may influence possible surgical complications.

Case Report

A 66-year-old female patient was on ambulatory follow-up due to right HS, unresponsive to treatment with botulinum toxin. Magnetic resonance imaging (MRI) of the skull revealed compression of the right facial nerve in its cisternal pathway by vascular loop and CM-I, with no signs of craniovertebral instability or symptoms related to CM-I \((\text{Fig. 1}).\)

The patient underwent surgery for the treatment of HS by microdecompression of the facial nerve via right lateral suboccipital craniectomy, showing complete resolution of face spasm. However, at the third postoperative day, the patient presented with somnolence, speech difficulty, and episodes of respiratory distress. Computed tomography (CT) and magnetic resonance imaging (MRI) revealed a mild edema of the right cerebellum related to surgical manipulation, and minimal hydrocephalus \((\text{Fig. 2}).\)

Due to the clinical worsening, external ventricular drain was opted. As there was no improvement, we considered the clinical worsening as being caused by the decompression of CM I causing compression of the brainstem, despite the small degree of postoperative cerebellum edema. The patient was then subjected to a median suboccipital craniectomy with decompression of the structures of the magnum foramen, laminectomy of C1 and C2, duraplasty with autologous muscle fascia graft, without cerebellar tonsillecrtomy \((\text{Fig. 3}).\)

The patient had a slow hospital recovery but with progressive and significant improvement. She was discharged alert with mild mental confusion, gait difficulty, and symmetrical tetraparesis, without facial spasm and without episodes of dyspnea and dysphonia. She remains in outpatient clinic follow-up with progressive improvement of symptoms and without facial spasm. \((\text{Fig. 4}).\) illustrates her last MRI images.

Discussion

Hemifacial spasm is a movement disorder of the muscles innervated by the facial nerve. Its clinical presentation includes spasms beginning in the upper hemiface, usually in the lower eyelid, that progress inferiorly affecting the cheek and the oral commissure; these findings are predominantly unilateral with a few bilateral cases described in the literature.\(^9\)\(^10\)\(^11\) Although it is not an immediate life-threatening condition, the affected patients tend to participate less in social interactions. As a severe psychosocial stressor, the condition requires timely diagnosis and therapy. Hemifacial spasm is a relatively rare condition that occurs mostly in middle-aged women, with a prevalence of 14.5:100,000 in women and 7.4:100,000 in men.\(^10\)
The pathophysiology of HS is not yet completely elucidated.\textsuperscript{12} In most cases, facial nerve compression is recognized by a vascular loop at the exit point of its nerve root in the brainstem.\textsuperscript{7} The exit zone of the facial nerve root has some characteristics that increase the vulnerability to stimuli: a) is an area in which the nerve is sheathed only by an arachnoid membrane, without epineurium; b) there is no connective tissue separating its fascicles; c) it is a

**Fig. 1** Magnetic resonance imaging of the preoperative skull showing malformation of Chiari type I, in addition to neurovascular compression in the complex VII/VIII cranial nerve on the right.

**Fig. 2** Postoperative cranial magnetic resonance imaging demonstrating absence of compression on the brainstem or excessive edema in the area of manipulation after the performance of neurovascular microdecompression of the facial nerve.
Hemifacial Spasm Associated with Chiari Type I Malformation

Lobão et al.

transitional zone between the central (oligodendroglial cells) and peripheral (Schwann cells) myelination regions. Some authors presuppose that in HS there is a hyperexcitability of the motor nucleus of the facial nerve in the brainstem predisposing to spasms. Additional etiologies implicating the brainstem or cerebellopontine angle may also be the cause of HS. Moreover, there are studies reporting that the symptoms of HS have been triggered after Bell palsy due to facial nerve lesion.

The diagnosis of HS is based on the clinical evaluation, further complemented by electrophysiological studies and imaging examinations. The main differential diagnoses related to HS include blepharospasm, oromandibular dystonia, facial nerve tic, hemimasticatory spasm, focal convulsions, and synkinesis after facial nerve palsy.

The therapeutic options for HS range from clinical drug treatment and subcutaneous injections of botulinum toxin to the affected muscles, to vascular microdecompression surgery. The main medications used in the treatment of HS include drugs such as anticonvulsants, anxiolytics, and baclofen. Botulinum toxin type A injection produces considerable relief of symptoms in most patients with mild to moderate disease (90–100%). However, the improvement is temporary (mean duration of 3–6 months) and, thus, periodic administration of the drug is necessary. In addition, botulinum toxin treatment may be associated with side effects such as eyelid ptosis, keratitis, and diplopia. In the case presented here, the patient had already received treatment with medication and botulinum toxin, but over the years ceased to adequately respond, justifying the choice of surgical treatment.

Microvascular decompression is the treatment with better long-term control of HS, first postulated by Gardner and popularized by Jannetta and collaborators. The procedure aims to alleviate the vascular compression at the exit zone of the facial nerve root in the brainstem. After retrosigmoid craniotomy, the cerebellopontine angle is exposed under microscopic view and the exit zone of the facial nerve root in the brainstem is inspected to identify the location of vascular compression. As with any surgical procedure, facial nerve vascular microdecompression has several risks, including hearing loss and paresis or paralysis of the facial nerve that may be temporary or permanent. In addition, severe complications such as hematoma, hydrocephalus, cerebellar edema, ischemia of the brainstem, and subarachnoid hemorrhage may occur as in any posterior fossa surgery. The patient presented here had only a mild right cerebellum edema related to surgical manipulation, but it was enough for clinical decompensation of CM I and brainstem compression symptoms.

Chiari malformation type I is a congenital anomaly of the rhombencephalon characterized by caudal displacement of the cerebellar tonsils through the foramen magnum. In this context, the ectopia of cerebellar tonsils, the anatomopathological hallmark of CM I, causes a disproportion between the contingent of the posterior fossa, the skull, and its content, the neural tissue. This disproportion caused by the underdevelopment of the occipital somites originating from the mesoderm is believed to be the cause of the disease. This results in a small posterior fossa that predisposes to the herniation of its contents. Once developed, the cerebellar ectopia causes the disturbance of the craniovertebral pressure in the subarachnoid space, with the creation of a gradient that favors additional descent of the tonsils. These events predispose the formation of meningeal fibrosis and subsequent adherence around the foramen magnum, with eventual formation of syringomyelia, hydrocephalus, or both. Several anomalies of the nervous system are associated with this malformation and include, among others, enlargement of the foramen magnum, hypoplasia of cranial nerve nuclei, superior migration of the cerebellum by the tentorial notch, and hypoplasia of the brain sickle and the cerebellum stall.

Patients with CM I can be divided into two large groups based on their clinical presentation: those with signs and symptoms arising from spinal cord dysfunction, with syringomyelia as substrate; and those with signs and symptoms from the compression of the contents of the posterior fossa, foramen magnum, or both. Spinal cord dysfunctions may present as trunk and extremity dysesthesia, upper limb paresis, with atrophy of the musculature of the hands, spasticity in the lower limbs, dissociated sensory losses (pain/temperature) in the trunk and upper limbs, and urinary incontinence. Compression of the posterior fossa content, especially of the

Fig. 3 Intraoperative: Dura mater plastic with autologous pericranium graft.

Fig. 4 Cranial magnetic resonance imaging at the time of discharge of the patient, after performing the decompressive craniectomy of the posterior fossa.
cerebellum, causes ataxia and nystagmus, particularly vertical nystagmus, in the downward vertical saccade, known as “down-beating nystagmus” and, more dramatically, symptoms of brainstem compression.\(^3\)

Posterior fossa decompression in patients with CM I is the conventional treatment.\(^4\) However, postoperative complications may occur, such as surgical wound infection, meningitis, cranial nerve palsy, edema of the posterior fossa, hematomas, postoperative hydrocephalus, and cerebellar mutism.\(^4\) In the present study, the patient had some of these symptoms; thus, we indicated the requirement of the second surgery for median suboccipital craniectomy and decompression of the posterior fossa content.

The association between the 2 pathologies is rare, with just over 20 cases described so far in the literature.\(^3\) The signs and symptoms of the disease are similar to those found in idiopathic HS with the upper part of the face predominantly involved during the initial stages, when CM I is asymptomatic. Strahle et al.,\(^5\) in 2011, reported that only 32% of patients with CM I detected in MRI were considered symptomatic during clinical diagnosis, indicating a high number of patients with pathology, but asymptomatic. Cheng et al.,\(^6\) in 2015, reported a higher level of superposition of structures located in the posterior fossa in HS patients compared with healthy volunteers. The small volume of the posterior fossa causes agglomeration of the content of the cerebellopontine cistern, that is considered a factor which may cause the vascular compression of the facial nerve.\(^5,4,45\)

Other mechanisms have been proposed as possible causes of the cranial rhizopathies associated with CM I, such as the axial traction of the cranial nerves caused by the caudal displacement of the altered römencephalon.\(^46,47\) Moreover, microischemia is possible to occur in these nerves.\(^58\) In addition, the dynamics in the flow of the cerebrospinal fluid caused by blockage in the craniocervical junction may be responsible for the traction of the cranial nerves.\(^59\)

Most patients with HS and CM I receive individualized treatment based on the predominance of symptoms related to each pathology. Patients without significant CM I symptoms should be treated for HS with neurovascular microdecompression of the facial nerve.\(^3,49\) On the other hand, patients with the two pathologies, with signs of craniovertebral instability, or clear symptoms of CM I, should be treated initially with foramen magnum decompression with or without craniovertebral fixation.\(^9,13,15\) It is expected that the symptoms of the HS ameliorate after surgery for CM I in patients with the two pathologies. Subsequently, it is possible to consider neurovascular microdecompression of the facial nerve for patients who do not recover from facial spasm after decompression of the foramen magnum.\(^3,48,50–52\)

Our patient was treated initially with facial nerve neurovascular microdecompression that led to the clinical resolution of HS. However, at the third postoperative day, her level of consciousness was lowered, she had speech difficulty and episodes of dyspnea/apnea. This deterioration was then attributed to the decompression of CM I due to postoperative, yet mild, cerebellar edema. After decompression of the foramen magnum, the patient presented slow recovery, and was discharged from the hospital alert, with mild mental confusion, some gait difficulty, symmetrical tetraparesis, and without HS.

Although neurovascular microdecompression is the treatment of choice in patients with HS and CM I without predominant symptoms of CM I,\(^53\) there are inherent risks associated with this surgery. Dealing with the association between HS and CM I, Cheng et al.,\(^3\) in 2017, suggested that foramen magnum decompression should be considered as the primary procedure, especially if there is associated syringomyelia.\(^3\)

**Conclusion**

Neurovascular microdecompression of the facial nerve is the procedure of choice for patients with HS and CM I, when HS symptoms prevail and there is no craniovertebral instability. However, possible complications of this surgery that may be caused by edema of the posterior fossa, albeit mild, could induce decompression of the symptoms of CM I. The case reported here exemplifies a situation like this and should serve as an alert for the potential need for a second immediate surgical intervention.

**Conflict of Interests**

The authors have no conflict of interests to declare.

**References**

Hemifacial Spasm Associated with Chiari Type I Malformation

Lobão et al.

23 Jannetta PJ. Outcome after microvascular decompression for typical trigeminal neuralgia, hemifacial spasm, tinnitus, disabling positional vertigo, and glossopharyngeal neuralgia (honored guest lecture). Clin Neurosurg 1997;44:331–383
24 Jannetta PJ. Typical or atypical hemifacial spasm. J Neurosurg 1998;89(02):346–347
27 Lovely TJ, Getch CC, Jannetta PJ. Delayed facial weakness after microvascular decompression of cranial nerve VII. Surg Neurol 1998;50(05):449–452
31 Bejiani GK. Definition of the adult Chiari malformation: a brief historical overview. Neurosurg Focus 2001;11(01):E1