Hemifacial Spasm Secondary to a Neurenteric Cyst: case report and literature review

Espasmo Hemifacial Secundário a Cisto Neuroentérico: relato de caso e revisão de literatura

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ABSTRACT

Introduction: Hemifacial spasms are clonic movements of facial muscles innervated by the facial nerve. They are unilateral by definition, traditionally caused by vascular compression of the exit zone of the facial nerve. Few cases described in the literature address neurenteric cysts as the cause. The present study reports a rare case of neurenteric cyst causing this neurological alteration. and reviews the literature on this pathology and its causes. Case presentation: 58-year-old woman with a 3-year history of left hemifacial spasms. She was investigated with a brain MRI which showed a cystic lesion at the cerebellopontine angle. We submitted the patient to resection of this lesion via the retrosigmoid approach in the supine position. The anatomopathological studies brought us the diagnosis of neurenteric cyst, a rare entity of the cerebellopontine angle. Conclusion: Our case followed the narrative review of the literature on the subject and the retrosigmoid approach in lateral decubitus is sufficient to resect a lesion of the posterior fossa compressing the facial nerve. With greater access to investigation with modern and advanced images, it is likely that a broader spectrum of diagnoses causing hemifacial spasm will be found and treatment should always be directed to the etiology.

Keywords: Hemifacial Spasm; Compressive; Neurenteric cyst

RESUMO

Introdução: Os espasmos hemifaciais são movimentos clônicos dos músculos faciais inervados pelo nervo facial. São unilaterais por definição tradicionalmente tendo como causa a compressão vascular da zona de saída do nervo facial. Poucos casos descritos na literatura abordam cistos neuroentéricos como causa. O presente estudo vem relatar um raro caso de cisto neuroentérico causando essa alteração neurológica, e revisar na literatura sobre essa patologia e suas causas. Relato de Caso: mulher de 58 anos com história de 3 anos de espasmos hemifaciais esquerdos. Ela foi investigada com uma ressonância magnética cerebral que mostrou uma lesão cística no ângulo pontocerebelar. Submetemos a paciente à ressecção desta lesão por via retrosigmoidoide em decúbito dorsal. Os estudos anatomopatológicos nos trouxeram o diagnóstico de cisto neuroentérico, entidade rara do ângulo pontocerebelar. Conclusão: Nosso caso seguiu a revisão narrativa da literatura sobre o tema e a abordagem retrosigmoidoide em decúbito lateral é suficiente para ressecar uma lesão da fossa posterior comprimindo o nervo facial. Com o maior acesso à investigação com imagens modernas e avançadas, é provável que se encontre um espectro mais amplo de diagnósticos causadores de espasmo hemifacial e o tratamento deve ser sempre direcionado à etiologia.

Palavras-chave: Espasmo Hemifacial; Compressivo; Cisto neuroentérico

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INTRODUCTION

Hemifacial spasms are clonic, jerking, involuntary movements of the facial muscles innervated by the facial nerve. They are unilateral by definition and known to be caused by vascular compression of the exit zone of the facial nerve, especially susceptible due to the lack of epineurium and transition of myelinating cells. They spread downwards after initiating at the orbicularis oculi muscles. However, Singh et al. have reported a case of spasm secondary to an epidermoid tumor of the cerebellopontine angle in which the hemifacial spasms followed the inverse order.

Regarding the causes of hemifacial spasm, Loeser and Chen stated in their series of 450 operated cases that 89% were due to arterial compression, 4% due to venous compression, 0.4% due to aneurysms, and 0.2% due to arteriovenous malformations. About 1% of cases of hemifacial spasms were attributed to CPA tumors.

It is essential to discuss and understand the treatment of hemifacial spasm according to the cause of the disorder due to the morbidity that may accompany it. Although it is naturally benign, it may lead to social exclusion and, in severe cases, functional blindness (due to involuntary and persistent eye closure). Interesting mechanisms for depression have been associated with feedback to the amygdala when the spasm produces frowning movements.

CASE PRESENTATION

We present the case of a 58 year old female with a 3 year history of left hemifacial spasms. She was investigated with a brain MRI that showed a cystic lesion in the cerebellopontine angle (CPA) (Figure 1), with a hypointensity on T1 (Figure 2) acquisitions and hyperintensity in FIESTA (Figure 1) and FLAIR (Figure 3) acquisitions. There was no restriction to diffusion and the lesion did not enhance after contrast administration. We submitted the patient to resection of this lesion through a retrosigmoid approach in the dorsal decubitus position. The pathological studies brought to us the diagnosis of a neurenteric cyst, a rare entity of the CPA causing hemifacial spasm. In the immediate postoperative care, we observed that the patient did not have spasms and the neurological improvement remained after a 3 year follow-up.

Figure 1. MRI FIESTA sequence.
Figure 2. MRI T1 post-Gadolinium infusion sequence.
Figure 3. MRI FLAIR sequence.
DISCUSSION

Hemifacial spasms (HFS) refer to sudden and involuntary twitching of facial muscles on one side of the face. Typically, this unusual muscular activity initiates near the eye muscles and progressively extends to the muscles on that particular side of the face\(^1\). The underlying cause of HFS is associated with prolonged, subtle nerve damage. Multiple researchers have demonstrated that these facial spasms often stem from the compression of a nerve due to vascular factors at the point where the nerve exits the root zone. However, the presence of this vascular compression is not consistently observed in all cases\(^1\).

Ehni and Woltman first described a series of 106 patients with hemifacial spasm before the noninvasive imaging era\(^2\). They identified a higher prevalence in women, a pattern of spread, an average onset age of 45 years old, and worsening of symptoms due to anxiety and fatigue.

Causes of hemifacial spasm other than vascular compression of the facial nerve are rare and seldom described in the literature\(^1\). Gardner and Sava, Kay and Adams, and Nagata add to the description of 43 cases, of which only 4 were caused by expansive lesions. Nagata’s\(^2\) study found glossopharyngeal neurinoma, tentorial meningioma, and 2 other cerebellopontine angle (CPA) tumors, which were not further described\(^1\).

However rare, it is essential to rule out secondary hemifacial spasm through imaging, as shown in the retrospective study conducted by Sprik et al.\(^10\) in 1988. The study revealed that a significant number of professionals who applied botulinum toxin did not investigate their patients further. Fortunately, with time and accessibility to imaging, secondary causes have been excluded more frequently before any therapeutic decision.

A paper by Kuroda et al.\(^11\) cites two patients with primary hemifacial spasm with compression of the facial nerve on its exit zone in MRI. Both declined surgical treatment and had improvement but experienced side effects with clonazepam. They reported significant improvement, with one patient remaining symptom-free and the other remaining symptom-free but then being lost to follow-up after treatment with levetiracetam.

Loeser and Chen\(^1\) reported an incidence of about 1% for hemifacial spasm due to CPA tumors in their 450-case series.

Galvez-Jiménez et al.\(^1\) reported 3 cases of tumors causing hemifacial spasm, including one parotid gland neoplasm, one CPA meningioma, and one vestibular schwannoma. Parotid gland tumors can cause compression of a specific branch of the facial nerve (mandibular buccal branch, as described by this author) and cause segmental spasms only.

Until 2015, when Jain et al.\(^12\) reported a case of a pleomorphic adenoma of the parotid gland presenting with hemifacial spasm, only 3 cases with a benign pathological diagnosis presenting in a similar way had been reported. In this case, there was complete improvement of the spasm postoperatively; however, the tumor had an extension and was tethered to the stylomastoid foramen, and partial mastoidectomy was also conducted. In these lesions, it is important to identify the facial nerve proximal and distal to the tumor before exercising it en bloc, especially when a malignant tumor is in the differential diagnosis. Due to the rarity of cases of parotid tumors causing facial spasm, when there is no involvement of the stylomastoid foramen, the origin of the spasm might be misinterpreted. Martin et al.\(^13\) presented a similar case of a pleomorphic adenoma of the parotid gland and hemifacial spasm ipsilaterally, which did not improve after total parotidectomy\(^13\).

When considering tumors of the posterior fossa causing hemifacial spasm, it is essential to note that, in most cases, when the etiology of this condition is compressive secondary to the neoplasm, there are associated neurological symptoms or findings in complementary investigations (tinnitus, hearing loss, unsteady gait, alterations in brainstem evoked potentials or electronystagmography). Rarely, paroxysmal clicking sounds in the ear may occur due to involvement of the stapedius muscle\(^1,5\).

Rarely, other lesions such as central nervous system neurocysticercosis, as reported by Yang et al.\(^14\), may compress the facial nerve causing hemifacial spasms, as well as arachnoid cysts, gliomas, cavernous angiomas, epidermoid tumors, and other neoplasms presenting with this condition in isolated reports when they are located in a way that can compress the facial nerve. In the cases reported with the aforementioned conditions, there was a complete improvement in symptoms postoperatively, with onset of symptoms having occurred up to 5 years\(^12,15,16\).

Also uncommon, there are reports of lesions in the IV ventricle and brainstem causing hemifacial spasm. Usually, these present with other long tract and cranial nerve symptoms. Dudoit et al. collected cases from three French hospitals. The patients were submitted to...
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an extensive investigation to exclude clinical and surgical diagnoses of brainstem lesions that are not gliomas and were submitted to stereotactic biopsy. Out of the 14 adult patients studied, 2 presented with hemifacial spasm, and 4 presented with both peripheral facial palsy and spasm. Unfortunately, the author did not specify the response of the hemifacial spasm to radiotherapy (and chemotherapy in some cases), but the treatment resulted in improvement of facial nerve symptoms in 57.1% of cases

Navarro-Olvera et al. have described a case of a 43-year-old female with hemifacial spasm secondary to a choroid plexus papilloma compressing the ipsilateral facial colliculus, refractory to botulinum toxin, and complete recovery 1 year after resection of the tumor.

IV ventricular and brainstem tumors are not uncommon in children. However, presentation with hemifacial spasm is. Yingjie et al. presented 4 cases of children with tumors of the cerebellar peduncle (1 hamartoma and 3 ganglioglioma). They confirmed the anatomical relation through tractography in one case, and after surgical resection, all had resolution of symptoms. In children with eye twitching, there might be a correspondence to limbs and moaning, which might resemble seizures. Therefore, it is important to evaluate the semiology of the episodes. In the author’s review, it was very common to misinterpret facial spasms and seizures, and electroencephalograms were part of the investigation. Some authors have described mechanisms between mass, pons, and cerebellum that might lead to cortical discharge and seizures as well.

Regarding neurenteric cysts of the cerebellopontine angle causing hemifacial spasm, during our review, only in one case reported by Umezu et al. was presented, and it resulted in complete recovery after surgical resection of the cyst. Interestingly, in a case reported by Eynon-Lewis et al., simple fenestration and biopsy of the lesion caused only partial and temporary improvement of VII-VIII complex symptoms in their patient, and recurrence of the cyst led to a new procedure for complete resection and broad resection of the lesion and its capsule.

In the previously mentioned cases, the compression in the facial nerve was not only in its cisternal portion but also at its exit zone. Rarely, pure compression of the cisternal or intracanalicular portion may cause symptoms due to the greater resistance of the Schwann cells that it involves. In 2020, Ligas et al. reported the case of a patient with severe stenosis of the internal auditory meatus causing hemifacial spasm ipsilaterally, and with complete resolution of symptoms after drilling of the internal acoustic meatus (IAM) and durotomy to decompress the VII-VIII nerve complex. Their review concluded that the IAM must be about 7.5 mm in children and 7.1 mm in adults. In their patient, the stenotic IAM measured about 2 mm. In these patients, it is important to exclude bony tumors through the CT scan.

Rarely, arteriovenous malformations (AVMs) and aneurysms of the skull base might be the cause of hemifacial spasm and peripheral facial palsy. Nagata described two cases of cerebellar AVMs, one case of medullary AVM, and one vertebral artery aneurysm treated surgically. After clipping of the aneurysm, the hemifacial spasm resolved. However, the facial palsy persisted. Subtotal resection of the medullary AVMs and total resection of the cerebellar AVMs produced complete resolution of symptoms.

Being it through aberrant regeneration, ephaptic transmission, or the ‘kindling model,’ the mechanism of compressive hemifacial spasm occurs with no clear risk factors. Although Oliveira et al. might have reported the association with Systemic Arterial Hypertension, due to the tortuosity that the vessels of the posterior fossa acquire, there is no clear correlation when evaluated by other authors.

Although there is a tendency for neurosurgeons to attribute spasms of any muscle that composes the facial structure to hemifacial spasm (essentially a surgical condition), other rare entities must be excluded, and the characteristics of the spasm must be carefully examined. Psychogenic hemifacial spasm, hemimastikatory spasm, and facial myokymia; focal seizures, tardive dyskinesia, aberrant regeneration after facial nerve injury (causing synkinesias), motor tics, and blepharospasm are included in this list. Therefore, a specific sign once called ‘the other Babinski sign’ may aid in diagnosis: when the orbicularis oculi muscle contracts to close one eye, the inside of the frontalis muscle contracts as well in hemifacial spasm. The semiotic aspects of each condition are essential in clinical differentiation. For example, in blepharospasm, Charcot’s sign is seen where the eyelids close with bilateral synchronous muscular contraction, in contrast to hemifacial spasm.

The explanation for the tendency of primary hemifacial spasms (related to vascular compression) beginning in the upper half of the face is hypothesized by Naraghi et al. when they stated...
that the posterior fossa vessels usually stand dorsal to the facial nerve, where the fibers to the upper half of the face are disposed.\(^5\)

Hemifacial spasm is essentially a surgical condition due to its known natural history, which includes chronification and worsening of symptoms. Spontaneous resolution has been seen in less than 10% of patients.\(^5,25\)

During the investigation, some anatomical aspects have been described to increase the chances of symptomatic vascular compression of the facial nerve, causing hemifacial spasms specifically. Vaguely described aspects include a small posterior fossa and cisternal spaces, and the literature lacks a review of these aspects, which increases the probability of such symptoms in patients with tumors of the posterior fossa. Additionally, Hamasaki et al. studied the relation between the flatness of the posterior fossa in the cranio-caudal axis and found that it was smaller in patients with hemifacial spasm. Again, this correlation with secondary hemifacial spasm must be studied specifically.\(^26,27\)

Regarding treatment, the three options usually accepted are botulinum toxin applications, pharmacotherapy, and microsurgical approaches. Pharmacotherapy (anticonvulsants, benzodiazepines) has inconsistent efficacy and considerable side effects. Botulinum toxin may be applied in the upper half of the face and around the platysma, as well as selected perioral regions (to avoid drooping of the angle of the mouth) and has been widely used. Its effect starts 2-3 days after injection and lasts for 2-3 months. It has been noted that the required dose increases with repeated applications. Aside from these, ptosis, diplopia, and mild facial paresis are also known side effects.\(^5\)

Especially for secondary hemifacial spasm, as presented in our case, surgery is the only treatment that can provide a total and permanent cure for this condition. However, due to the availability of botulinum toxin, it has been reserved in many cases for patients with symptoms that are refractory to botulinum toxin.\(^5\)

Jannetta et al. described hemifacial spasm as a surgical condition that must be treated at its origin. Although new percutaneous and ‘noninvasive’ treatments have been emerging, especially for secondary spasms, surgical resolution of the compression has excellent results, as shown in our case. It agrees with the results in the literature, and a retrosigmoid approach in the lateral decubitus position suffices to resect a lesion of the posterior fossa compressing the facial nerve or remove the compression of the dolichoectatic verteobasilar complex and bring the patient to a cure. With time and greater access to investigation with modern and advanced imaging, a wider spectrum of diagnoses causing hemifacial spasm is likely to be found, and the treatment must always be directed to the etiology.

REFERENCES


CONCLUSION

Jannetta et al. described hemifacial spasm as a surgical condition that must be treated at its origin. Although new percutaneous and ‘noninvasive’ treatments have been emerging, especially for secondary spasms, surgical resolution of the compression has excellent results, as shown in our case. It agrees with the results in the literature, and a retrosigmoid approach in the lateral decubitus position suffices to resect a lesion of the posterior fossa compressing the facial nerve or remove the compression of the dolichoectatic verteobasilar complex and bring the patient to a cure. With time and greater access to investigation with modern and advanced imaging, a wider spectrum of diagnoses causing hemifacial spasm is likely to be found, and the treatment must always be directed to the etiology.


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