The Journal of International Advanced Otology

J Int Adv Otol 2020; 16(1): 138–40 • DOI: 10.5152/iao.2020.6112

Case Report

Vestibular Impairment in Hemifacial Spasm Syndrome: A Case Report

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Cite this article as: Barona L, Krstulovic C, Bejarano B, Perez N. Vestibular Impairment in Hemifacial Spasm Syndrome: A Case Report. J Int Adv Otol 2020; 16(1): 138-40.

A 52-year-old man presented with left hemifacial spasm (HFS). A magnetic resonance imaging scan showed compression of the left facial nerve at the cerebellopontine angle by a dolichoectatic basilar artery. The neurotological evaluation showed an otolithic deficit, with canalicular preservation and normal hearing. The deficit improved after surgical decompression. No previous report has described the impairment of vestibular function in patients presenting with HFS.

KEYWORDS: Vestibular-evoked myogenic potentials, hemifacial spasm, vertebrobasilar dolichoectasia, facial nerve diseases

INTRODUCTION

Hemifacial spasm (HFS), vestibular paroxysmia, trigeminal neuralgia, and glossopharyngeal neuralgia are disorders associated with a vascular compression of the corresponding nerve in its route to/from the brainstem.

Hemifacial spasm can be detected through the vascular compression of the facial nerve (VII cranial nerve) in the internal auditory canal (IAC) or cerebellopontine angle (CPA). It is characterized by involuntary, uncontrolled, unilateral facial muscle contractions (spasms), which are usually painless. It is a rare condition with a prevalence of 9.8 per 100,000 [1].

The facial nerve, situated in the pontomedullary junction, comprises part of the acoustic-facial bundle. This group is composed of the facial nerve, intermediate nerve of Wrisberg, cochlear nerve, and vestibular nerve. Considering the proximity of these nerves, a vascular compression may simultaneously affect more than one of them.

CASE PRESENTATION

Written consent was obtained from the subject prior to the preparation of this case report. A 52-year-old man presented with a 3-month history of painless involuntary intermittent contractions of the left eyelid (myokymia), which had spread to the entire left hemifacial musculature. The patient reported a significant improvement after sleep. No pain, altered sensitivity, or other neurological symptoms were described; also, no hearing loss or vestibular symptoms were reported.

A neurological assessment showed slight facial paresis in the left frontal area, accompanied by frequent spasms in the orbicular and perioral regions on the same side. Compound muscle action potentials recorded from the left facial nerve showed 53.4% axonal loss. High frequency discharges in the upper and lower left facial area without signs of active denervation were recorded in an electromyogram (EMG).

Hearing was normal on both sides: a pure tone audiometry showed an average of 15 dB HL in the left ear and 16.25 dB HL in the right ear. No spontaneous (with or without visual fixation), gaze-evoked or positional nystagmus were observed. To further assess vestibular function, both video head impulse test (vHIT; GN Otometrics, Taastrup, Denmark) and vestibular-evoked myogenic potentials (VE-

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Submitted: 27.09.2018 • **Revision Received:** 25.04.2019 • **Accepted:** 23.05.2019 • **Available Online Date**: 18.03.2020
Available online at www.advancedotology.org

MPs; Biomed, Jena, Germany) were performed. The vHIT revealed normal vestibulo-ocular reflex (VOR) gain for all six semicircular canals. In contrast, cervical VEMPs showed a 53% lower response during left ear stimulations, indicating a saccular deficit (normal asymmetry <30%), and ocular VEMPs showed a 38% lower response during left ear stimulations, indicating a utricular deficit (normal asymmetry <30%).

An axial Constructive Interference in Steady State (CISS) magnetic resonance imaging (MRI) of the brain centered on the CPA revealed the presence of a dolichoectasia of the basilar artery impinging on the left acoustic-facial bundle (Figure 1).

A neurosurgical team performed a two-step microvascular decompression (MVD) process of the acoustic-facial bundle using a retrosigmoid approach. The first step provided a course improvement of the acoustic-facial bundle by drilling the posterior wall of the IAC decreasing its angulation at the pore level. Since the position of the dolichoectatic basilar artery did not allow mobilization, the only way to decrease the impingement on the facial nerve was to drill the posterior wall of the IAC. This ensured the displacement of the facial nerve posteriorly by rectifying the angled course of the nerve.

The second step achieved a permanent nerve decompression by interpositioning pieces of Teflon felt between the nerve and the vertebral artery and posterior inferior cerebellar artery (Figure 2). The two steps were performed consecutively during a single surgical intervention.

A Grade IV facial paralysis was observed immediately after surgery, which was subsequently treated with a methylprednisolone tapering regimen.

Facial paralysis improved to Grade I 3 months after surgery. At this time, the patient reported no involuntary muscle contractions, hearing disturbance, or balance impairment.

The auditory and vestibular assessment was repeated after surgery. As before surgery, hearing was normal on both sides, and the vHIT revealed normal VOR gain for all six semicircular canals, although a complete recovery was observed in both otolithic potentials. Cervical VEMPs showed 14% lower response during left ear stimulations (indicating a symmetric saccular function), and ocular VEMPs showed 30% lower response during left ear stimulations (indicating a symmetric utricular function).

After surgery, facial nerve discharges disappeared in the EMG assessment, but signs of chronic denervation were recorded.

DISCUSSION

Hemifacial spasm is a condition that can be caused by any vascular structure compromising the facial nerve. The main clinical features of this syndrome are as follows: (1) it is an adult disease that occurs predominantly in the fifth decade of life; (2) the onset is insidious and only muscles innervated by the facial nerve on one side are affected (there may be bilateral spasms but the onset and clinical manifestations are asymmetrical); (3) typically, the first muscles to be affected are the orbicularis oculi, and later there is a cefalocaudal progression of muscular involvement (90% of cases); (4) the timing, anatomical site, and spread of spasm varies at different times in the same person; (5) there is contraction of several muscle groups innervated by the facial nerve, although anatomically, these muscles may be far from each other; (6) the spasms worsen in stressful situations and improve

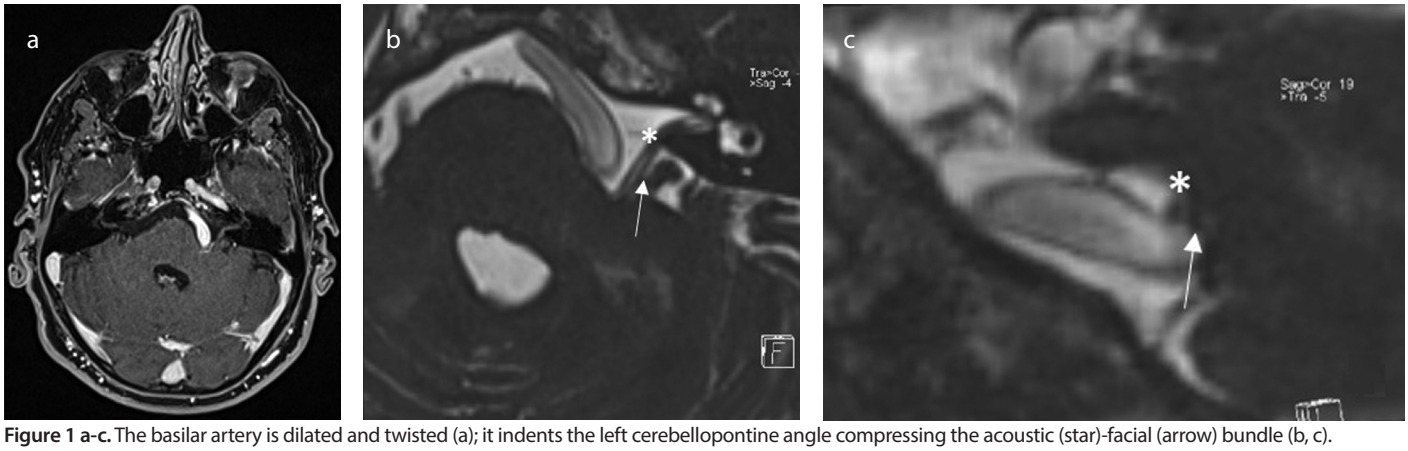

Figure 1 a-c. The basilar artery is dilated and twisted (a); it indents the left cerebellopontine angle compressing the acoustic (star)-facial (arrow) bundle (b, c).

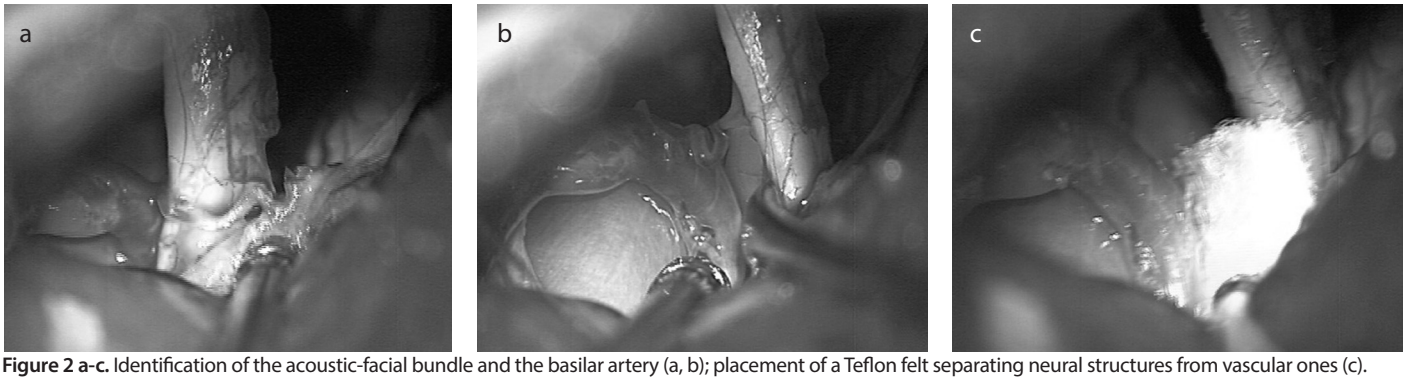

Figure 2 a-c. Identification of the acoustic-facial bundle and the basilar artery (a, b); placement of a Teflon felt separating neural structures from vascular ones (c).

with rest, but they do not disappear with sleep; (7) it cannot be suppressed voluntarily; and (8) as a general rule, there is no pain [2].

MRI is the imaging modality of choice since it is the most sensitive tool for detecting vascular impingement of the facial nerve, particularly CISS and Fast Imaging Employing Steady-state Acquisition sequences [3].

Although there is consensus that MVD of the facial nerve is a technique with few complications in the hands of an expert surgeon, scientific studies estimate that the temporary or permanent iatrogenic paralysis of the facial nerve ranges between 1% and 23%, and hearing iatrogenic disorders range between 2% and 15% [2]. Other serious complications reported in literature, with very low incidence, are cerebellar hematomas, stroke, and death [4]. Both these studies agree that the complication rate is higher in reoperations.

In our case, the compression of the facial nerve in the acoustic-facial bundle caused HFS and the simultaneous appearance of a quantifiable vestibular deficit. It is possible that given the close proximity of the facial nerve and the vestibular-cochlear nerve, the displacement or distortion of the acoustic-facial bundle may cause either HFS or vestibular paroxysmia or any degree of overlap between them. In addition, the extensive arterial dolichoectasia forcing the posterior wall of the IAC to be drilled could explain the important percentage of observed facial deficit (53.4% axonal loss) and the presence of accompanying vestibular deficit.

Vestibular paroxysmia is a disorder caused by the neurovascular compression of the vestibular-cochlear nerve. It was described in 1975 by Janetta et al., [5] who called it "disabling positional vertigo." Brandt et al. [6] described vestibular paroxysmia as a new vestibular disorder, which meets the following criteria: (1) short attacks of rotational to-and-fro vertigo lasting seconds to minutes, (2) attacks frequently dependent on particular head positions, (3) hearing loss or tinnitus, (4) measurable auditory or vestibular deficits by neurophysiologic methods, and (5) efficacy of carbamazepine.

Although the otolithic deficit may have been incidentally found and not related to the offending vessel, the presence of the deficit on the same side of the lesion and the complete recovery of the deficit after surgery are sufficient arguments to attribute the vestibular compromise to the arterial dolichoectasia.

CONCLUSION

Taken together, the findings in the present case suggest the importance of quantifying vestibular function in subjects presenting with HFS. In addition, it would be interesting to measure the function of the facial nerve in subjects presenting with vestibular paroxysmia due to the possible overlap of deficits caused by the compression of the acoustic-facial bundle.

This is the first case reporting a vestibular deficit in a subject with HFS. Further studies are needed to determine whether the presence of a vestibular deficit in subjects with HFS is a frequent condition or an exceptional finding.

Informed Consent: Informed written consent was obtained from the patient who participated in this study.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept – L.B., N.P.; Design - L.B., N.P.; Supervision - N.P.; Resource - N.P., B.B.; Materials - N.P., B.B.; Data Collection and/or Processing - C.K., L.B.; Analysis and/or Interpretation - C.K., L.B.; Literature Search - C.K.; Writing - C.K., L.B.; Critical Reviews - N.P., B.B..

Conflict of Interest: The authors have no conflict of interest to declare.

Financial Disclosure: The authors declared that this study has received no financial support.

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