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# Hemifacial spasm through changes of cerebrospinal fluid pressure in idiopathic intracranial hypertension

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## ABSTRACT

Hemifacial spasm (HFS) is a rare complication of idiopathic intracranial hypertension (IIH); hence, little is known about its pathophysiology and treatment options. Here, we report a 29-year-old woman with a 3-month history of IIH and HFS. Following a diagnostic and therapeutic lumbar puncture, she suffered from attacks of HFS for a few days every time she rose from a supine position. The relatively sudden drop in cerebrospinal fluid (CSF) pressure led to frequent HFS attacks, which, in contrast to the attacks in the previous months, were strictly observed only when getting up from the supine position. This observation suggests changes in CSF pressure as the likely pathophysiological mechanism responsible for HFS in IIH, rather than the absolute CSF pressure. She was then successfully treated with topiramate, a known therapeutic option in IIH, but not yet described for HFS in IIH.

**KEYWORDS** Cerebrospinal fluid pressure; hemifacial spasm; idiopathic intracranial hypertension; topiramate

**H**emifacial spasm (HFS), a term coined by Joseph Babinski in 1905,<sup>1</sup> describes a movement disorder that arises from hyperexcitation of the facial nerve and is characterized by spontaneous involuntary contractions of the ipsilateral facial muscles. A typical clinical sign that can help differentiate HFS from other movement disorders affecting the facial nerve is the so-called “other Babinski sign,” which describes the raising of the ipsilateral eyebrow during contraction of the orbicularis oculi.<sup>2</sup> The mechanism of HFS is heterogeneous. In primary HFS, the most prevalent type, the facial nerve at its brainstem exit is in contact with an abnormal or ectatic blood vessel,<sup>3</sup> which mechanically leads to spontaneous depolarizations of the facial nerve. Secondary HFS can result from various underlying conditions including tumors in the cerebellopontine angle or in the parotid gland, as well as infections and ischemic lesions.<sup>4</sup> Here, we describe HFS in a young woman suffering from idiopathic intracranial


hypertension (IIH). The focus of the report is on a pathophysiological link between the two conditions.

## CASE DESCRIPTION

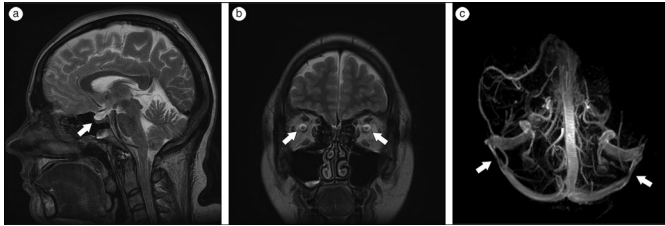
An overweight 29-year-old woman (body mass index 33 kg/m<sup>2</sup>) was admitted to our neurology clinic because of dull holocephalic headache and recurrent spasms of her left facial muscles occurring over 3 months (**Video 1**). An ophthalmologist diagnosed bilateral papilledema, and cranial magnetic resonance (MR) imaging revealed the empty sella sign, bilateral distended optical nerve sheaths with increased perineural fluid, and bilateral stenosis of the transverse sinus before the transition to sigmoid sinus (**Figure 1**). No vascular contact of the facial nerve was seen on MR angiography. Physical examination was normal without any focal neurological deficits. Extended laboratory tests did not reveal any abnormalities either. A lumbar puncture revealed a slightly elevated opening pressure of 260 mm H<sub>2</sub>O. Interestingly, draining around 40 mL of cerebrospinal fluid (CSF) induced

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**Figure 1.** T2-weighted MR images showing the (a) “empty sella sign” (arrow) and (b) bilateral distended optic nerve sheaths with increased perineural fluid (arrows). (c) Bilateral stenosis of the transverse sinus before the transition to the sigmoid sinus (arrows).

recurrent HFS attacks every time she rose from the supine position. These HFS attacks ceased after about 3 days. The patient was treated for IIH with topiramate at a final dose of 50 mg twice daily. She did not tolerate acetazolamide. With this regimen, both the HFS and the headache subsided completely and did not recur in the 6 months of follow-up.

## DISCUSSION

A very rare and poorly characterized cause of secondary HFS is IIH. IIH itself is a rare neurological disorder characterized by an increase in intracranial pressure (ICP) and not associated with underlying pathologies such as intracranial tumors, hydrocephalus, or brain edema. Common symptoms are headache, visual impairment, and abducens nerve palsy.

To date, only a few case reports linking IIH and HFS have been published in the English medical literature.<sup>5–9</sup> In all case reports, HFS occurred intermittently and was remedied by lumbar puncture and treatment with the carbonic anhydrase inhibitor acetazolamide. Mayer Benegas et al reported the recurrence of symptoms after acetazolamide treatment was stopped,<sup>8</sup> suggesting an association between ICP and HFS. However, there is no obvious correlation between absolute ICP and the occurrence of HFS, as the previously reported CSF opening pressures fluctuated widely, from 260 to 480 mm H<sub>2</sub>O.<sup>5–9</sup>

In this context, we highlight two important findings of our report. First, triggering of postdrainage HFS attacks by orthostasis indicates that alterations of ICP rather than absolute values relate with HFS. A recent report of HFS during CSF drainage in a patient with IIH<sup>7</sup> underlines the hypothesis that changes in ICP lead to hyperexcitation of the facial nerve in IIH. Furthermore, contrary to the pathophysiological concept of pulsatile depolarizations of the facial nerve due to contact with an aberrant vessel leading to short-lasting HFS attacks, [video 1](#) shows a relatively long-lasting HFS attack. The clinical presentation likely reflects the pathophysiology, as the dystonic character of HFS attacks in this case would match longer-lasting changes in ICP in IIH.

Second, this case shows that HFS in IIH can be successfully treated with topiramate. Topiramate is commonly used

as a second-line drug for IIH, alternatively to acetazolamide. Topiramate inhibits the carbonic anhydrase, which reduces CSF secretion in the choroid plexus. Studies in mice showed that it lowers ICP more effectively than acetazolamide.<sup>10</sup> Additionally, its side effect of facilitating weight loss is welcomed in the mostly obese IIH patients. In conclusion, HFS is a rare but debilitating complication of IIH. The condition is manageable and adequate therapy can significantly improve the patient’s quality of life.

## SUPPLEMENTAL MATERIAL

**Video: Hemifacial spasm and “the other Babinski sign”:** This homemade video recorded by the patient demonstrates a hemifacial spasm (HFS) from its onset to its termination. The relatively long-lasting HFS begins rather simultaneously in the muscles innervated by the temporal, zygomatic, buccal, and marginal mandibular branches of the left facial nerve. In particular, “the other Babinski sign” can be appreciated as the eyebrow is lifted at the same time that the orbicularis oculi contracts. Note that the “selfie” has the side reversed.

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