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Clinical Observations

Hemifacial Spasm in a Child Treated With Microvascular Decompression of the Facial Nerve

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ABSTRACT

BACKGROUND: Hemifacial spasm is a rare condition in children that is characterized by involuntary contractions of muscles innervated by the ipsilateral facial nerve. **PATIENT DESCRIPTION:** We describe a 6-year-old girl who presented with intermittent involuntary spasms of the right face. Magnetic resonance imaging demostrated a loop of the anterior inferior cerebral artery contacting and elevating the cisternal segment of the right facial nerve; this finding was confirmed at surgery where microvascular decompression of the facial nerve was performed without complication. Following surgery she had immediate remission of symptoms, but the hemifacial spasms slowly recurred within 8 months of surgery only to resolve by age 11 years. **CONCLUSION:** This is the youngest patient reported with hemifacial spasms related to a vascular etiology, which initially responded to surgical treatment. The authors review this syndrome in children and discuss possible etiologies and management options.

Keywords: hemifacial spasm, facial nerve, anterior inferior cerebellar artery, vascular loop, microvascular decompression, craniotomy

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Introduction

Hemifacial spasm is characterized by involuntary contractions of the muscles innervated by the ipsilateral facial nerve.[1-3] The prevalence for all ages is 11/100,000, more in women (14.5/100,000) than that in men (7.4/100,000).[4,5] The condition is usually unilateral although bilateral involvement can occur. The disease can be classified as primary or secondary and typical or atypical.[1,2,4] Primary hemifacial spasm is typically attributed to compression of the seventh cranial nerve at its exit zone from the brainstem by a vascular loop. The secondary form is associated with facial nerve palsy or injury to the facial nerve caused by conditions such as infection, trauma, demyelination, or tumor. Hemifacial spasm may also occur without an obvious cause. The typical form of hemifacial spasm starts in the orbicularis oculi muscle and progresses downward to involve the orbicularis oris muscle, buccinator muscle, and occasionally the platysma muscle. Atypical hemifacial spasm is rare (fewer than 10% of the patients) and starts inferiorly in the orbicularis oris and buccinator muscles and spreads upward towards the orbicularis oculi muscle. Primary hemifacial spasm occurs most commonly in middle-aged adults (fourth to sixth decades) and is more common in Asians. A small subset of approximately 0.9% to 6.5% of patients develop symptoms before age 30 years, typically the secondary form.[4,6]

Patient Description

This 6-year-old girl presented with recurrent intermittent involuntary twitching of the right face. Symptoms started at age 5 years with mild spasms of the right cheek progressing to involve the right corner of her mouth. The onset was gradual, but the episodes increased in frequency and became continuous. The facial contractions were of variable

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intensity, more noticeable during eating, drinking, talking, present during sleep, and worsened with stress and fatigue. The symptoms were distressing to the child. She was initially diagnosed with a tic disorder at an outside institution and treated with guanfacine and carbamazepine without improvement.

The patient demonstrated intermittent hyperacusis on the right and occasional headaches and right facial pain. Her pain was attributed to the repetitive right-sided muscular contractions and muscle fatigue and was responsive to over-the-counter analgesics. Review of neurological symptoms was otherwise unremarkable.

Her past medical history was unremarkable. There was no family history of neurological disorders. The results of physical and neurological examinations were normal with the exception of rhythmic repetitive contractions of the muscles of the right side of her face (Fig 1 and Video 1). The facial spasms were more prominent in the right cheek area and caused retraction of the right corner of her mouth and the area below the right lower eyelid. The spasms could be triggered by repetitive touching of the face.

Laboratory findings including a complete blood cell count, electrolytes, calcium, potassium, magnesium, thyroid, renal, and liver function tests were normal. Brain magnetic resonance imaging (MRI) demonstrated a loop of the right anterior inferior cerebellar artery (AICA) within the right internal auditory canal contacting and elevating the right facial nerve (Fig 2).

The patient was diagnosed with atypical hemifacial spasm because the lower face was predominantly affected. The patient underwent microvascular decompression of the right seventh cranial nerve via a right-sided retromastoid approach. Surgery confirmed indentation and compression of the undersurface of the right facial nerve by the AICA loop. The artery was mobilized away from the seventh and eighth nerve complex and separated from the seventh cranial nerve by a small Telfa pledget. Postoperatively, she experienced complete resolution of hemifacial spasms, transient right-sided hearing loss, which quickly resolved, and no facial weakness (Figs 3-5).

Within eight months of surgery, the hemifacial spasms recurred in clusters, again involving the right lower portion of her face. The facial contractions were less severe in intensity and duration than the patient had experienced before surgery. A repeat MRI demonstrated the Teflon pledget in good position interposed between the cisternal segment of the right seventh cranial nerve and the right AICA. No further surgery was recommended. At age 13 years now, the girl remains asymptomatic.

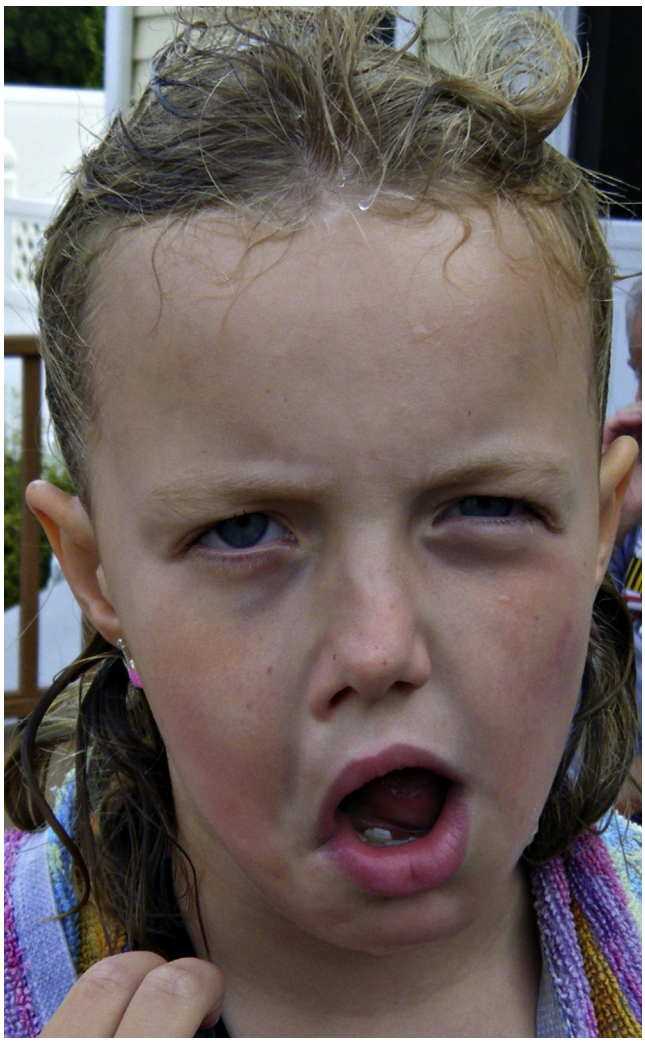

FIGURE 1.
Episode of right facial spasm. The video of this patient can be found at http://dx.doi.org/10.1016/j.pediatrneurol.2016.01.007. (The color version of this figure is available in the online edition.)

Discussion

Hemifacial spasm has a characteristic clinical pattern. The facial spasms last from seconds to minutes and can present with facial contractions that are usually clonic and occasionally tonic or a combination of both.[1,7] Muscular spasms usually start within the orbicularis oculi muscle and may progress over a period of months to years to involve the rest of the lower muscles of the face. The left side of the face is more commonly affected.[1,3,7,8] Affected individuals cannot voluntarily suppress the facial contractions and may persist during sleep. Muscular spasms can be elicited by stress, fatigue, anxiety, repetitive voluntary facial movements, changes in head position, bright light, and cold temperatures.[1,7,9]

In children, hemifacial spasm is commonly attributed to trauma, neoplasms of the brainstem or cerebellopontine angle (pilocytic astrocytomas, gangliogliomas, gangliocytomas, hematomas), infections (otitis media with effusion and tuberculous meningitis), disorders of the skull base and meninges (thickening of the arachnoid membrane, arachnoid cyst), and vascular abnormalities (venous sinus thrombosis, aneurysms of the vertebral artery, and vascular compression).[4,8,9,10-17] In adults, hemifacial spasm is most commonly caused by vascular compression of the seventh cranial nerve. Symptoms usually present around the age of 50 to 60 years and are commonly associated with tortuous, ectatic atherosclerotic arteries, related to aging and hypertensive disease.[1,9,14,16]

High-resolution MRI detects neurovascular compression of the ipsilateral facial nerve in 88% to 93% of patients,[18] most commonly involving the AICA, the superior cerebellar artery, and rarely the vertebral artery.[1,7] This girl had a common anatomic variant of the AICA, forming a loop within the cerebellopontine angle and internal auditory canal that resulted in compression of the facial nerve. Microvascular compression of the facial nerve is considered a rare cause of hemifacial spasm in children. Two young patients with vascular etiologies resembling our patient have been reported in the literature: a 10-year-old boy with an AICA loop compressing the facial nerve and a 13-year-old girl with a vein arteriolar complex causing facial nerve compression.[8,9] The tortuous vascular loop at the root exit zone of the seventh cranial nerve is