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Hemifacial spasm in a child

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Abstract. Hemifacial spasm in a 10-year-old child is herein reported. Computed tomography and angiography were normal. Magnetic resonance imaging showed an anomalous vascular structure, probably a redundant loop arising from AICA or PICA, which was identified at surgery. The spasm, presumably due to vascular compression at the root entry zone of the right facial nerve, promptly and completely remitted after surgical decompression, without functional deficits. Atypical clinical features, as well as pre- and postoperative neurophysiological findings supporting the microvascular compression theory, are discussed.

Key words: Hemifacial spasm – Microvascular compression – Neurosurgery – Blink reflex

Hemifacial spasm is a relatively common condition due to a hyperactive dysfunction of the facial nerve. It is found more frequently in women, has a peak incidence in the fifth decade (the youngest patient reported being 17 years old), and is characterized at the onset by mild, intermittent twitching of the inferior orbicularis oculi [5]. The disorder is almost exclusively unilateral and in most series has been reported to affect the left side.

With time, spasms become more and more frequent, severe and persistent, spreading down to the inferior part of the face, and finally involving the platysma. In contrast, the frontalis muscle generally is not involved [3].

Progression is slow, but the disorder may become very distressing within a few months, twiches being eased by changes in head and body position and exacerbated by using facial expression muscles and, in some patients, by emotion and fatigue. The occurrence of twitches during sleep is common.

Mild facial weakness may be an early symptom, gradually worsening with time, while pain is infrequent, except occasionally in the buccal region during prolonged "tonus". The "tonus" phenomenon is a severe, persistent contraction of the involved muscles, causing almost complete closure of the eye and the drawing up of the corner of the mouth. No spontaneous remission has been reported.

The causative factor of hemifacial spasm is still a matter for debate [7], except in cases due to pontocerebellar angle tumors or vertebral artery aneurysms. Neither is there agreement on where the facial nerve hyperactivity originates. According to the most accepted theory, known as "microvascular compression" (MCV), the seventh nerve is compressed by an artery, or more rarely a vein, in the root entry zone (REZ), which is believed to represent a vulnerable area, due to the change from peripheral to central myelin.

This theory, introduced by Jannetta in 1976 [5], has been recently discussed by Adams and Chir [1], and Kaye and Adams [6]. Reviewing the literature on this topic, they reported both MCV without hemifacial spasm and hemifacial spasm without MCV to be very frequent, suggesting that the disorder could rather be due to nuclear dysfunction.

In this paper, we report the unusual case of hemifacial spasm in a child.

Case report

A 10-year-old boy was admitted to our hospital with an almost 3-year history of right hemifacial spasm. The previous anamnesis was completely negative. Perinatal and neonatal life, psychological and motor development were normal.

Mild contractions of the right orbicularis oculi had suddenly begun at 7 years of age. There were no triggering events. Twitches, which were exacerbated by anxiety and emotion, rapidly worsened both in frequency and extension, spreading to the corner of the mouth and to the frontal region. Six months after the onset, they occasionally started to fix in a prolonged spasm. They were more frequent when the child was awake, but also occurred during sleep.

Ten months after the onset, the boy was admitted to a pediatric department. Electroencephalography (EEG), electromyography (EMG), radiography of skull and spine and magnetic resonance imaging (MRI) of the brain were all reported to be normal. No

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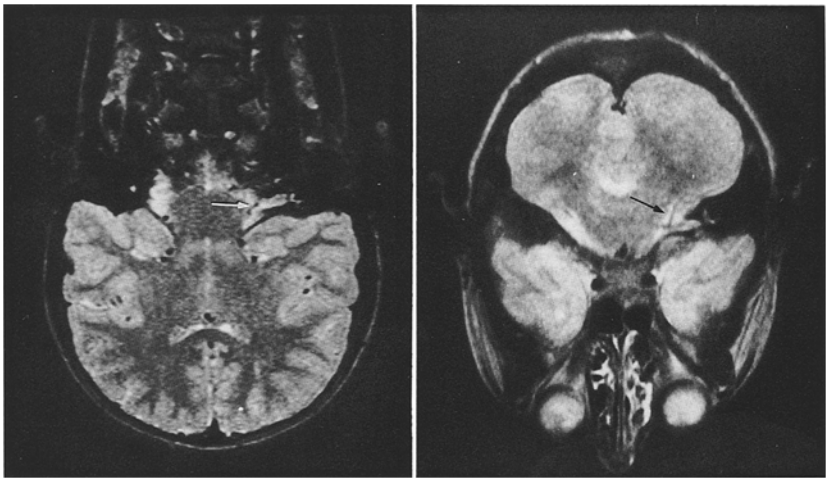

Fig. 1. Tranverse and coronal T2-weighted images (TR 2100, TE 100) show an anomalous vascular structure in the right cerebellopontine angle cistern (*arrow*)

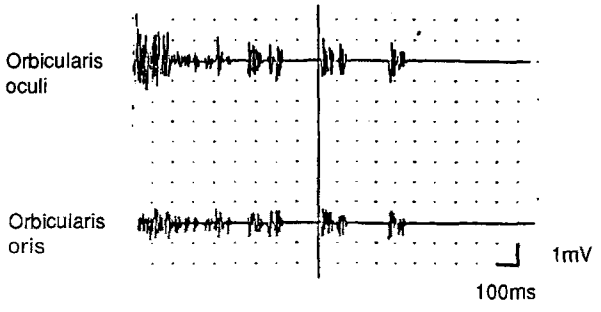

Fig. 2. Spontaneous activity: typical, synchronous and arrhythmic bursts of motor unit potentials were recorded. As shown, the duration of the spasm was variable

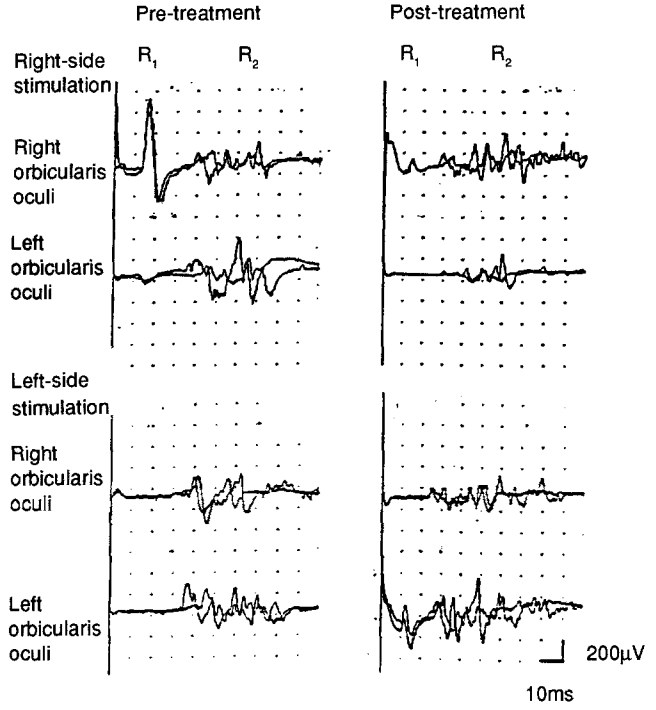

Fig. 3. Pre-treatment blink reflex (BR): ipsilaterally to the spasm (right side) an abnormally enlarged R1 response was obtained. R1 recorded after left-side stimulation was suppressed. No relevant changes in R2 responses were detected. Post-treatment BR was normal

diagnosis was made. He was treated with carbamazepine without any significant improvement.

When he was admitted to our hospital, about 3 years after the onset, neurological examination was completely normal, except for severe contractions of the right orbicularis oculi, with involvement of the corner of the mouth. The contractions were triggered by every attempt to use the facial muscles voluntarily, as requested when examining the seventh nerve. Even looking at the patient could cause him to start twitching.

EEG, otovestibular examination, auditory tests and brain-stem auditory evoked potentials (BAEPs) were all normal. MRI showed an anomalous vascular structure in the right cerebellopontine angle cistern, adjacent to the seventh and eight cranial nerves, possibly representing a redundant loop of a branch of AICA or PICA (Fig. 1). On angiograms it was not possible to identify this vessel exactly, nevertheless the hypothesis of a vascular malformation was excluded.

In an EMG study, simultaneous recording by means of needle electrodes was performed from the orbicularis oculi and oris muscles. At rest arrhythmic and synchronous bursts of motor unit potentials (MUPs) were recorded on both muscles. The burst duration was variable and ranged from 50 to 200 ms (Fig. 2). On voluntary activity a full interference pattern was recorded.

A nerve conduction velocity (NCV) study was performed. By electrical stimulation at conventional sites a normal distal latency of facial nerve and normal amplitude compound muscle action potentials were obtained.

Blink reflex (BR) was tested by means of conventional techniques (Fig. 3). On stimulation of the right (affected) side, an abnormally enlarged R1 direct response was evident; the R2 direct component was inconstant and only slightly anticipated with respect to the latency onset of the consensual R2. On left (unaffected) side stimulation, the R1 component was suppressed; R2 showed slight anticipation with respect to the R2 direct component. No changes in latency onset, amplitude or duration of R2, both direct and consensual, were found.

Surgery was performed on 28 May 1990. With the patient in the sitting position, via a suboccipital retromastoid craniectomy on the right side, the cerebellopontine angle (CPA) was approached after dural opening and positioning of a self-retaining spatula over the infero-lateral aspect of the cerebellar hemisphere.

Utilizing the microscope, the eight nerve was identified and followed from the internal meatus toward the brain stem. The AICA was seen coming straight from the anterior portion of the cerebellopontine cistern, just under the auditory nerve, at some distance from the pons: it could be identified as the vessel seen on MRI in the right CPA (see Fig. 1). A small recurrent branch (about 1 mm in diameter) arose from the AICA and was directed upward, passing between the eighth and the seventh nerves. The REZ of the seventh nerve was clearly compressed by this ascending vessel in its

posterocaudal aspect: after this portion of the nerve had been freed from tiny arachnoidal adhesions with the vessel, a slight but visible indentation was apparent over the REZ. An implant of autologous muscle was placed between the artery and the seventh nerve, close to the brain stem, and a drop of fibrin glue ensured the stability of the implant, so that it could not flow away with the cerebrospinal fluid.

At the end of the procedure, neither the facial nor the auditory nerve showed any contusion. The dura mater was closed watertight. The wound was closed in the usual manner. No drain was left in place.

Since the operation no spasms have occurred. The postoperative course has been uneventful. The child was discharged after a week, without any neurological deficits or complaints.

At rest, no clonic facial activity was recordable by EMG. On voluntary activity a full interference pattern was recorded, as before treatment. NCV was normal, as before the operation. Concerning BR, on stimulation of the right and left sides normal amplitude and latency of the R1 and R2 responses were obtained (Fig. 3). BAEPs were normal, except for a mild, non-significant asymmetry of the III component latency. Auditory tests were normal.

Four months after surgery the boy is still doing well, without any recurrence of spasms (either clinical or electrophysiological).

Discussion

From the clinical point of view, the age of our patient is the most unusual feature in this case. Hemifacial spasm is usually reported to occur from the second to the sixth decade, with a peak incidence in the fifth. Our patient (10 years old) was far younger than the youngest previously reported in literature, who was 17.

Moreover, our patient suffered from a typical hemifacial spasm, starting from the orbicularis oculi and spreading down but: (1) the frontal muscle was also involved (as in atypical spasms, starting from the orbicularis oris); (2) a posterocaudal microvascular compression on the REZ was found, not an anterocaudal one, as might be expected in classical spasms [4].

Moreover, this case shows interesting neurophysiological findings that support the microvascular compression theory. EMG, NCV and BR performed before and after treatment showed no evidence of peripheral nerve damage. The EMG spontaneous activity, typical of hemifacial spasm, promptly disappeared after treatment. The preoperative BR showed findings consistent with abnormalities of the brain-stem mechanisms involved in the control of the excitability of R1 and R2 responses. The facilitation of the ipsilateral R2 component has already been reported in hemifacial spasm [2], indicating that central mechanisms may contribute to the facial spasm; however, in our case, an abnormally enlarged ipsilateral R1, associated with a depressed controlateral R1, indicates and suggests that the mechanism responsible for R1 may be involved independently from those responsible for the R2 component. Interestingly, the postoperative BR was normal.

Therefore, the electrophysiological findings in our case indicate that:

1. Hemifacial spasm probably results from an ectopic hyperactive focus that produces the clinical phenomena by ephaptic mechanisms, alone or in combination with brain-stem central mechanisms due to abnormal backfiring neural activity.

2. The surgical procedure on the facial nerve not only abolishes the clinical phenomena but also restores the physiological brain-stem mechanisms of control of the excitability responsible for BR.

These data suggest that the electrophysiological brain-stem abnormalities reported in the present case and in cases of other authors [2] are probably secondary to a hyperactivity along the proximal portion of the facial nerve and that the clinical and electrophysiological improvement is not due to nerve trauma. In fact, the moderate entity of trauma is demonstrated by the absence of any clinical sequelae and by normal postoperative tests, including BAEP, otovestibular examination and audiograms. In conclusion, our data favor the microvascular compression theory in the pathogenesis of hemifacial spasm.

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