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Excitability of facial nucleus and related brain-stem reflexes in hemifacial spasm, post-facial palsy synkinesis and facial myokymia

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Abstract

Objective: To compare the electrophysiological excitability characteristics of the facial nucleus and related structures in hemifacial spasm (HFS), post-facial palsy synkinesis (PFPS) and facial myokymia (FM).

Methods: Facial F-waves, blink reflex recoveries and magnetically elicited silent periods (SP) were prospectively studied in 17 HFS, 17 PFPS, 8 FM cases and in 13 controls. Earlier unpublished observations on abnormal impulse transmission in 36 HFS and 29 PFPS cases were also included.

Results: Enhanced F-waves were recorded on the symptomatic side in PFPS and HFS cases with a tendency to be more pronounced in PFPS. HFS and PFPS groups both showed an earlier blink reflex recovery, more prominent in PFPS patients, when stimulated and/or recorded on the symptomatic side. Unelicitable SPs were encountered after 24/39 stimulations in 5 patients with PFPS and rarely in HFS cases. Duration of elicitable SPs did not change remarkably. FM group had similar characteristics as normal controls in the 3 electrophysiological tests. Latencies of the lateral and synkinetic spread responses were significantly prolonged in the earlier PFPS group as compared to HFS. In two-point stimulation, both groups showed a greater latency shift in late responses, again more pronounced in PFPS.

Conclusions: PFPS and HFS cases had similar enhanced excitability patterns at the facial nucleus and related brain-stem structures, more marked on the symptomatic side and more obvious in the PFPS group. Findings elicited in the FM group were thought to be caused by asynchronous hyperactivity of facial motoneurons.

Significance: In this comparative electrophysiological study, similar excitability patterns were found in HFS and PFPS groups, albeit with different intensities.

Keywords: Hemifacial spasm; Facial synkinesis; Facial myokymia; F-waves; Blink reflex; Magnetically elicited silent period

1. Introduction

Hemifacial spasm (HFS) and post-facial palsy synkinesis (PFPS) are two entities that can have both similar and distinct aspects. HFS presents with typical twitches and is usually induced by a vascular abnormality around the brain-stem without a history of a previous facial palsy. PFPS follows a facial nerve lesion intense enough to cause a severe paralysis; it presents with synkinesis between facial muscles, and spontaneous spasms are generally lacking (Tan et al., 2002; Valls-Solé and Montero, 2003; Valls-Solé et al., 1992). However, facial twitches, which may be difficult to discriminate from the clonic spasms of HFS, can be synkinetically produced in PFPS by the activation of another muscle. Synkinesis between the facial muscles is a common feature of HFS as well, and its electrophysiological indicators, synkinetic spread of blink reflex and lateral spread responses are found in both conditions (Eekhof et al., 2000; Öge et al., 1992, 1993). Therefore, from a different point of view, these two conditions seem to be similar synkinetic disorders.

Pathophysiological mechanisms leading to these conditions have long been debated: ectopic excitation-ephaptic transmission at the root entry zone, and increased excitability of the facial nucleus are two candidate

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mechanisms for HFS, while faulty nerve regeneration and hyperexcitability of nucleus and/or brain-stem reflexes are advocated for PFPS (Ferguson, 1978; Kimura et al., 1975; Moller, 1991; Nielsen, 1984a,b). In the last two decades, considerable evidence about increased excitability of facial nuclei and related brain-stem structures has been reported for both conditions (Brach et al., 1997; Valls-Solé and Montero, 2003; VanSwearingen and Brach, 2003). However, the relationship of the common electrophysiological characteristics with the clinically similar and distinct aspects of HFS and PFPS are not yet clarified, and reports comparing these two conditions directly are scarce in the literature (Auger, 1979; Eekhof et al., 2000).

Facial myokymia (FM) is characterized by tiny, asynchronous contractions of muscle fibers with electromyographically rhythmic–semirhythmic repetitions of potential groups. FM is usually a manifestation of multiple sclerosis (MS) and other lesions involving the brain-stem. Functional deafferentiation and hyperexcitability of the facial nerve nucleus, or a lesion in the intramedullary nerve fibers are implicated as the cause of myokymia in these disorders (Gutmann, 1991). Rarely, FM occurs in Guillain–Barré syndrome and in the other demyelinating extra-axial facial nerve lesions (Daube et al., 1979; Gutmann, 1991; Öge et al., 1996, 2001). It has previously been shown in a small number of patients that synkinetic spread of impulses between facial nerve branches are not present in FM (Auger, 1979; Nielsen, 1984a). The presence of different clinical and electrophysiological findings in a condition which is thought to be possibly resulting from facial nucleus hyperexcitability led us to include a FM group in our study.

The aim of this study is to compare HFS, PFPS and FM cases by using electrophysiological methods directed to evaluate excitability changes in the facial pathways. Some of our earlier unpublished observations comparing the abnormal impulse transmission in HFS and PFPS cases are also included in order to enhance the contrasts and similarities between the two groups.

2. Methods

2.1. Cases

Electrophysiological tests were performed in 17 patients with HFS, 17 patients with PFPS and 8 patients with FM who were referred to our laboratory consecutively between 2001 and 2003. None of the patients had a personal or family history of any other movement disorder. Control values were obtained from 13 healthy subjects. Informed consent was obtained from each subject and the local ethics committee approved the study.

2.1.1. HFS group

This group consisted of 7 females and 10 males aged between 27 and 83 years (56.6 ± 18.0 years). Duration of symptoms before the electrophysiological examination ranged from 3 to 96 months (36.2 ± 29.1 months). Eleven cases had left-sided and 6 cases had right-sided spasms. There were no cases with bilateral spasms or with any history of facial paralysis, head trauma and neurosurgical operation.

Synkinesis could be demonstrated clinically in 14 cases between the muscles innervated by different facial nerve branches. In 3 cases, mild paresis was also observed on the affected side. One patient had ipsilateral sensorineural hearing loss. Neurological examination of the other patients were normal except for findings related to HFS. Cranial magnetic resonance imaging (MRI) was performed in all patients. A cerebello-pontine cistern epidermoid tumor ipsilateral to the spasms, thought to be the cause of HFS, was found in one patient. In 9 patients a dolico-ectatic vertebrobasilar system artery was detected extending to the cerebello-pontine cistern on the symptomatic side. Only two patients had history of botulinum toxin injections for the treatment of HFS. One of them received regular injections for 2 years, the last one being 4 months ago. The other patient was given 4 injections within a one-year period, 4 years ago.

2.1.2. PFPS group

There were 8 females and 9 males with a mean age of 44.4 ± 15.5 years (range: 17–70 years). The interval between the onset of facial paralysis and the electrophysiological examination ranged between 6 and 240 months (mean: 63.3 months). The causes of facial palsy were Bell's palsy in 12 cases, Ramsay Hunt syndrome in one case, head injury in one case, 'ear operation' in two cases and a surgical procedure directed against an acoustic neuroma in one.

Eleven cases had right-sided and 6 cases had left-sided PFPS. In addition to synkinesis, mild to moderate paresis of the ipsilateral facial muscles was found in 15 cases, contracture in 7, and crocodile tears syndrome in 3. Neurological examination was otherwise normal in all the cases except for ipsilateral hearing loss in two cases whose facial palsies were caused by Ramsay Hunt syndrome in one and by an ear operation in the other.

2.1.3. FM group

There were 7 females and one male aged between 17 and 61 years (mean: 31.7 ± 13.3 years). Continuous undulating and quivering movements of FM were seen on the left side in 5 patients and on the right in 3. The time interval between the awareness of abnormal movements and the electrophysiological examination was 1–60 days (mean: 23.8 days). Four patients had ipsilateral facial contracture. Concentric needle EMG of the facial muscles on the symptomatic side revealed myokymic discharges that were characteristic for FM.

Seven patients had relapsing-remitting MS with a mean disease duration of 64.1 ± 81.4 months. Symptoms and signs (other than FM) indicating brain-stem involvement