Short report

Hemimasticatory spasm—a peripheral paroxysmal cranial neuropathy?

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SUMMARY The clinical and electrophysiological features of a case of hemimasticatory spasm are presented. The findings are in many respects similar to those described in hemifacial spasm. A peripheral cranial neuropathy as proposed in hemifacial spasm also may be responsible for hemimasticatory spasm.

Isolated unilateral contraction of the masticatory muscles (hemimasticatory spasm) is an uncommon type of involuntary facial movement. Earlier descriptions of this condition have been in association with facial hemiatrophy and are cited in a recent report of such a case¹ in which the similarity of hemimasticatory spasm and hemifacial spasm was noted. We present further electrophysiological studies from a patient with hemimasticatory spasm without facial hemiatrophy which document in detail these similarities.

Case report

A woman aged 60 years presented with a three-year history of recurrent spasm of the left temporalis and masseter muscles which would result in jaw closure and last for up to one minute. There were no definite precipitating factors with spasms occurring at rest and during voluntary jaw movements such as talking or chewing. Short spasms occurred in isolation or became repetitive leading to prolonged jaw closure. Numerous episodes occurred daily. Several years prior to the onset of spasm she experienced paraesthesiae and numbness in the left maxilla. This symptom fluctuated for several months before it resolved spontaneously. Shortly after the onset of spasm the sensory disturbance recurred and spread to involve the left maxillary and mandibular divisions of the trigeminal nerve.

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The sensory disturbance again resolved, however the jaw spasms continued unabated. Numerous medications including diazepam, phenytoin, clonazepam, baclofen, valproate, haloperidol and amitriptyline were tried without effect. A cryosurgical lesion of the motor root provided only temporary relief. Examination findings prior to the cryosurgical lesion were normal apart from hypertrophy of the left masseter and temporalis muscles (fig 1). In particular there was no weakness of the jaw musculature and sensation was normal in all divisions of the trigeminal nerve. Investigations, including electroencephalography, posterior fossa myelencephalography and carotid and vertebral angiography, were normal. Electrophysiological studies showed normal blink reflex latencies² for both early and late components. The jaw jerk recorded by mechanical stimulation³ when spasm-free showed an absent left masseter response. Needle electromyography of the masseter muscle showed no evidence of denervation and several patterns of muscle activity during spasm. These

Fig 1 Hypertrophy of left masseter and temporalis muscles in a 60-year-old female patient with hemimasticatory spasm.

Fig 2 Simultaneous recordings (needle electrodes) from the right (upper trace) and the left (lower trace) masseter muscles. Short bursts of activity in the left masseter muscle of increasing duration lead to more prolonged spasm. Right masseter muscle activity shows normal reflex inhibition on jaw closure.

spasms were composed of multiple units of normal morphology discharging at high frequencies. There was considerable variation in burst duration with short bursts of activity of between 20 and 100 ms and spasms of 175–500 ms. A crescendo of short bursts preceded the longer spasms on some occasions (fig 2) while others occurred spontaneously in isolation. Co-activation of the left temporalis muscle was present throughout the spasm. Activity in the right masseter muscle was inhibited on jaw closure during spasm while that in the left masseter was not (fig 2). Recordings from the lateral pterygoid muscle during masseter spasm were unsatisfactory, however, during voluntary jaw closure co-activation of the lateral pterygoid was observed.

Discussion

Hemimasticatory spasm in this patient was not associated with facial hemiatrophy. The antecedent trigeminal sensory neuropathy, normal blink reflexes and ipsilateral absence of the jaw jerk suggests an extra-axial trigeminal nerve lesion. Furthermore, the absence of the jaw jerk and the masticatory hypertrophy imply involvement of the Ia fibres, resulting in deafferentation of the muscle spindles and interruption of the ipsilateral monosynaptic masseter stretch reflex.³ The masseter and temporalis muscle hypertrophy may therefore have developed from sustained isometric contraction as a result of the loss of normal reflex inhibition of masticatory activity after jaw closure. The EMG patterns of brief spontaneous spasms, comprising normal motor units discharging at high frequencies and a tendency to become repetitive, often in a crescendo fashion, are similar to those described in hemifacial spasm.^{4–6} Comparable patterns of activity have been recorded arising ectopically from irritable peripheral nerves and nerve roots in a variety of situations^{7–9} while spontaneous repetitive discharges have been

recorded from the sensory fibres of the trigeminal nerve in the cat after gasserian ganglion injury.¹⁰ Such lesions in experimental models have been shown to generate ectopic impulses and to participate in neuronal cross activation at rapid and variable rates.⁹ It seems clear that such mechanisms also account for facial synkinesis, a cardinal feature of hemifacial spasm, where a peripheral lesion has been postulated.^{4,11} Simultaneous coactivation of the masseter and temporalis during spasm and the lateral pterygoid during jaw closure in the present case of hemimasticatory spasm indicates a disturbance of the normal pattern of muscle activation in a manner analogous to that seen in hemifacial spasm.

The aetiology of the peripheral trigeminal nerve lesion is uncertain in this case. Inflammatory disorders and vascular compression require consideration. The latter phenomenon has received much attention following the successful treatment of tic douloureux and hemifacial spasm by vascular decompression.^{11,12} With this in mind it is significant that the motor root of the trigeminal nerve lies in a position susceptible to compression from above by the superior cerebellar artery,¹³ a situation found to pertain in three cases of a series of 210 anatomical examinations.¹⁴ Hemimasticatory spasm without hemifacial atrophy should be included in the spectrum of paroxysmal cranial neuropathies whose mechanism seems to be due, at least in part, to vascular irritation and compression.

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