Acta neurol. belg., 2001, **101**, 113-116

Symptomatic palatal myoclonus : an unusual cause of respiratory difficulty

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Abstract

A 67-year-old man presented with dysphagia and difficulty breathing. Physical examination revealed palatal myoclonus. In this patient, the respiratory difficulty was caused by the fragmentation of breathing. Electromyographic examination of the cricothyroid muscle demonstrated rhythmic myoclonic jerks. Magnetic Resonance Imaging (MRI) yielded a pontine midline and right sided tegmental infarct. The patient responded to sodium valproate.

Key words : Palatal Myoclonus ; respiratory difficulty ; pontine infarct.

Introduction

Palatal myoclonus (PM) is a rare condition, characterized by continuous rhythmic, or dysrhythmic jerks of the soft palate and pharyngeal muscles. It is usually caused by a vascular ischaemic lesion of the brainstem or cerebellum that disrupts the dentato-rubro-olivary pathway. The movement can also spread to involve other branchial muscles, as well as the face, the eye, the neck, the diaphragm or the arm muscle groups. Respiratory difficulty is rare and drug treatment is usually of value.

In this report, a rare case of PM with respiratory difficulty is presented.

Case report

67-year-old male patient presented with a history of respiratory difficulty present for the last 3 months, that was experienced during daily routine activities. He complained of interruption in breathing and talking, he was out of breath with minimal efforts or even at rest. He noticed difficulty in swallowing both solids and liquids. He also described occasional aspirations. He had suffered an ischaemic cerebrovascular accident (CVA) 17 years before, that has caused a mild left-sided weakness and unstable gait. He was found to have hypertension and coronary artery disease, which were adequately controlled.

Upon examination, he had bilateral, rhythmic, myoclonic movement of the soft palate and pharynx at a rate of 2-3 Hz. Rhythmic movement of laryngeal cartilages was visible from outside. There were no abnormal diaphragmatic movements. Breathing and phonation were clearly fragmented as a result of these rhythmic movements. According to the obtained history, such movements did not change during eating or sleeping. Neurological examination disclosed slight dysarthria, truncal imbalance, and moderately ataxic gait. Deep tendon reflexes were hyperactive on the left side. Eye movements were normal and opsoclonus was not observed.

Routine laboratory examinations were normal except for slight hypercholesterolemia. Indirect laryngoscopic examination showed rhythmic movements of larynx in synchrony with palatal muscles. Abnormal thoracic movements were not observed. Electroencephalography was normal. Electromyography (EMG) of the cricothyroid muscle showed rhythmic bursts of EMG activity (Fig. 1). Magnetic resonance imaging of the brain demonstrated a midline and right-sided ischaemic lesion of the pons-tegmentum that was present both on T1 and T2 weighted images (Fig. 2, 3, 4). Administration of sodium valproate (1000 mg/d) resulted in a significant decrease of involuntary movements. During the follow-up period of two years, the patient no longer complained of respiratory difficulty with daily activities.

Discussion

Palatal myoclonus (PM) is a rare disorder characterized by involuntary rhythmic contractions of the palatal musculature, that are usually bilateral, at a rate varying between 40 and 240 per minute. Most patients present with tinnitus. PM may be associated with myoclonus of other muscle groups, including pharyngeal, laryngeal, diaphragmatic, facial, and ocular muscles (Hanson *et al.*, 1985).

Symptoms including dysarthria, dysphagia, irregular respirations, and airway obstruction may be accompanied with PM (Toland *et al.*, 1984). However, respiratory changes in palatal myoclonus have rarely been described (Sakurai *et al.*, 1993 ; Andrews *et al.*, 1987 ; Nagaoka *et al.*, 1984 ;

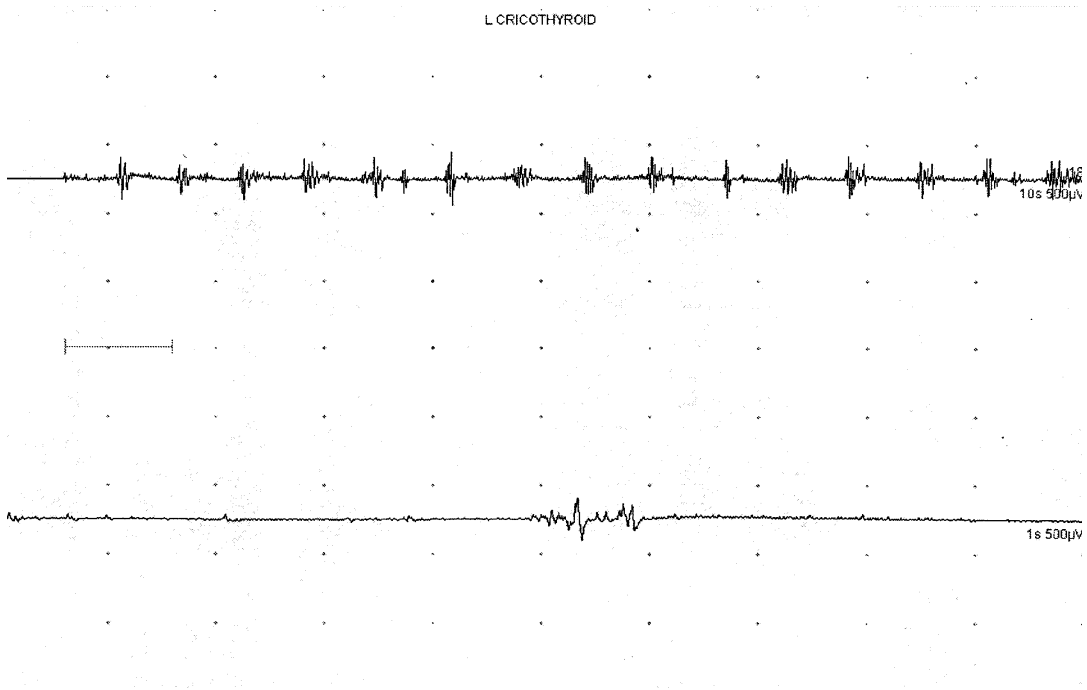

FIG. 1. — Myoclonic jerks in cricothyroid muscle

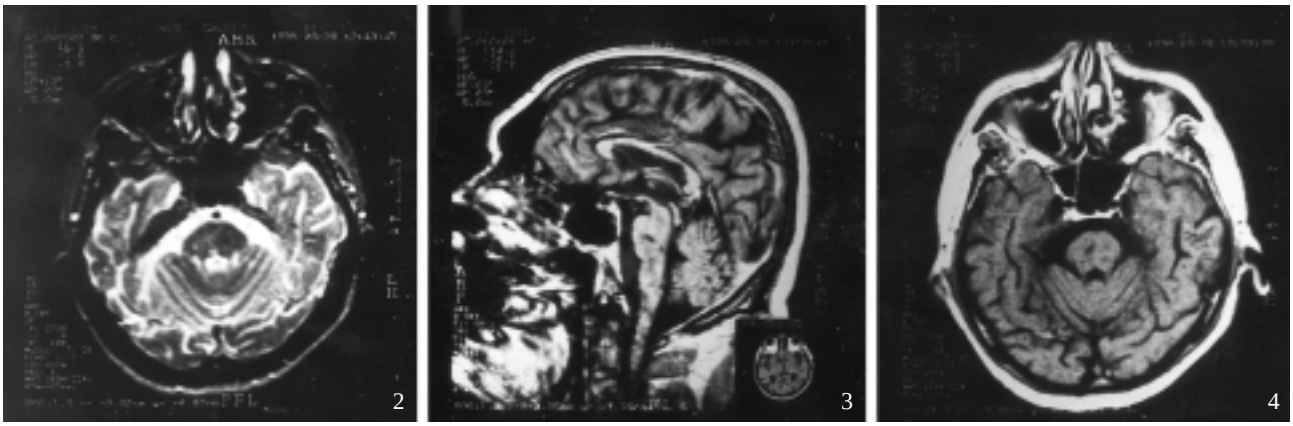

FIG. 2, 3, 4. — Brain MRI demonstrated pontine midline- right sided infarct on T2-weighted axial, T1-weighted axial and sagittal images.

Toland *et al.*, 1984 ; Bollinger *et al.*, 1974). Respiratory changes may be due to involvement of respiratory muscles or laryngeal muscles. On the basis of laryngeal examination, we postulated that laryngeal muscles act as an airway valve, thus causing fragmentation of respiration. Another rare movement disorder that can cause dyspnea is the "diaphragmatic flutter", which involves the diaphragm and intercostal muscles (Kobayashi *et al.*, 1990). However, abnormal diaphragmatic movements, which were not seen in our current case, are essential for the diagnosis of diaphragmatic flutter.

Inferior olivary hypertrophy usually accompanies palatal myoclonus clinically (Deuschl *et al.*, 1990 ; Hattori *et al.*, 1988 ; Lapresle, 1986). This is thought to be due to transneuronal degeneration caused by lesions interrupting the dentato-rubro-olivary pathway (myoclonic triangle) (Dubinsky *et al.*, 1988). The lesion is usually of ischaemic origin (Lapresle, 1986 ; Matsuo *et al.*, 1979). Several other causes less frequently observed include multiple sclerosis, trauma, tumours, degenerative diseases, subarachnoid haemorrhage (Deprez *et al.*, 1999 ; Nishigaya *et al.*, 1998 ; Tranchant *et al.*, 1995 ; Birbamer *et al.*, 1993). On the other hand, paraneoplastic syndromes might cause cerebellar degeneration or the syndrome of opsoclonus-myoclonus-ataxia (Bataller *et al.*, 2001). However, these should only be considered in the absence of an identifiable cause, yet in the current case there was an infarct in the myoclonic triangle.

The time period between the occurrence of the lesion in the myoclonic triangle and the clinical manifestations of palatal myoclonus was not clear for our patient. It is stated that the symptoms most

commonly occur within 10-11 months, although there might be a delay of several years (Lapresle, 1986 ; Matsuo *et al.*, 1919). In our opinion, the pontine infarct of our patient was silent initially and was not associated with the prior stroke that happened 17 years earlier.

Currently there is no definitive treatment for PM. Various medications including anticonvulsants, sedatives, antispasmodics, piracetam, and opioids were reported to be useful in occasional anecdotal reports (Karakostas *et al.*, 1999 ; Martinez et al 1993). In our patient, beneficial effects of valproate might result from its gamma-aminobutyric acid enhancing action. In conclusion, palatal myoclonus should be considered in patients with respiratory difficulties.

Diagnosis is usually based on clinical examination. However, MRI is essential for the definite diagnosis.

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