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Benign segmental myoclonus: electrophysiological evidence of transient dysfunction in the brainstem

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Summary We present a 66-year-old patient with segmental myoclonus evoked by a brainstem infarction. The myoclonus appeared soon after a cerebrovascular accident and it was evident in the soft palate, jaw, neck, shoulders and upper limbs. Brain MRI showed infarction in the left pons and left cerebellum. Small amounts of orally administered clonazepam were remarkably effective. Electroencephalogram (EEG) and auditory brainstem response (ABR) were normal. Somatosensory evoked potential (SSEP) revealed delays in P14 and N19 recorded at C3 by right median

nerve stimulation. These findings were normalized in 4 days. Segmental myoclonus is thought to be evoked by olivary hypertrophy following cerebrovascular accident in the brainstem and is said to be resistant to medication. The limited involvement of the brainstem in our patient may account for the transient segmental myoclonus. The prognosis for this type of segmental myoclonus is excellent. © 2001 Harcourt Publishers Ltd.

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INTRODUCTION

Myoclonus is defined as abrupt, instantaneous, jerky involuntary movements unassociated with loss of consciousness.¹ Myoclonus can be evoked by various causes,² and is classified by its origin.¹ For example, lesions in the cerebral cortex, brainstem and spinal cord may cause myoclonus. Segmental myoclonus is characterized by rhythmic or arrhythmic involuntary contractions of muscle groups supplied by one or several contiguous segments of the brainstem or spinal cord.³ Palatal myoclonus is a special type of segmental myoclonus and appears exclusively in the soft palate.⁴ We report a patient with segmental myoclonus induced by a brainstem infarction with electrophysiological evidence of transient dysfunction in the brainstem.

CASE REPORT

A 66-year-old man, who had been treated for hypertension, received an operation for a varix in the left leg under epidural anaesthesia. The surgical procedure was successful, and the patient was discharged 5 days later. In the early morning of the first day after discharge, he noticed an abnormal movement in his neck, and visited the emergency room of our hospital. Neurological examination revealed vigorous rhythmic involuntary movement in the jaw, neck and shoulders. These movements were jerky, with rates varying from 120 to 180 oscillations/min. A series of abnormal involuntary movements appeared at a rate of 2–10/min with a duration of 1–2 s per series. These movements, which were distinct from tics, chorea, or other involuntary jerky movements, were diagnosed as myoclonus. Palatal myoclonus (PM) was also present, but negative myoclonus was not apparent. Additional neurological examinations revealed nothing abnormal. Sodium valproate (300 mg/day) was administered and he went home. On the evening of the same day, the involuntary movement became continuous, and the patient was again admitted to the emergency room. The distribution and the rates of myoclonus were unaltered, but the symptoms were aggravated. Myoclonus ceased completely upon intravenous administration of diazepam (10 mg). He continued to receive administration of oral clonazepam (1 mg/day). The next day, intermittent myoclonus was observed, and the day after that, he was admitted to our hospital. Neurological examination revealed a slight terminal tremor of the left arm, accentuation of right knee jerk, and positive Barré's sign in the right leg. The myoclonus had disappeared. Urinalysis and blood laboratory data showed no abnormality. Chest X-ray showed cardiomegaly. ECG revealed a right bundle branch block.

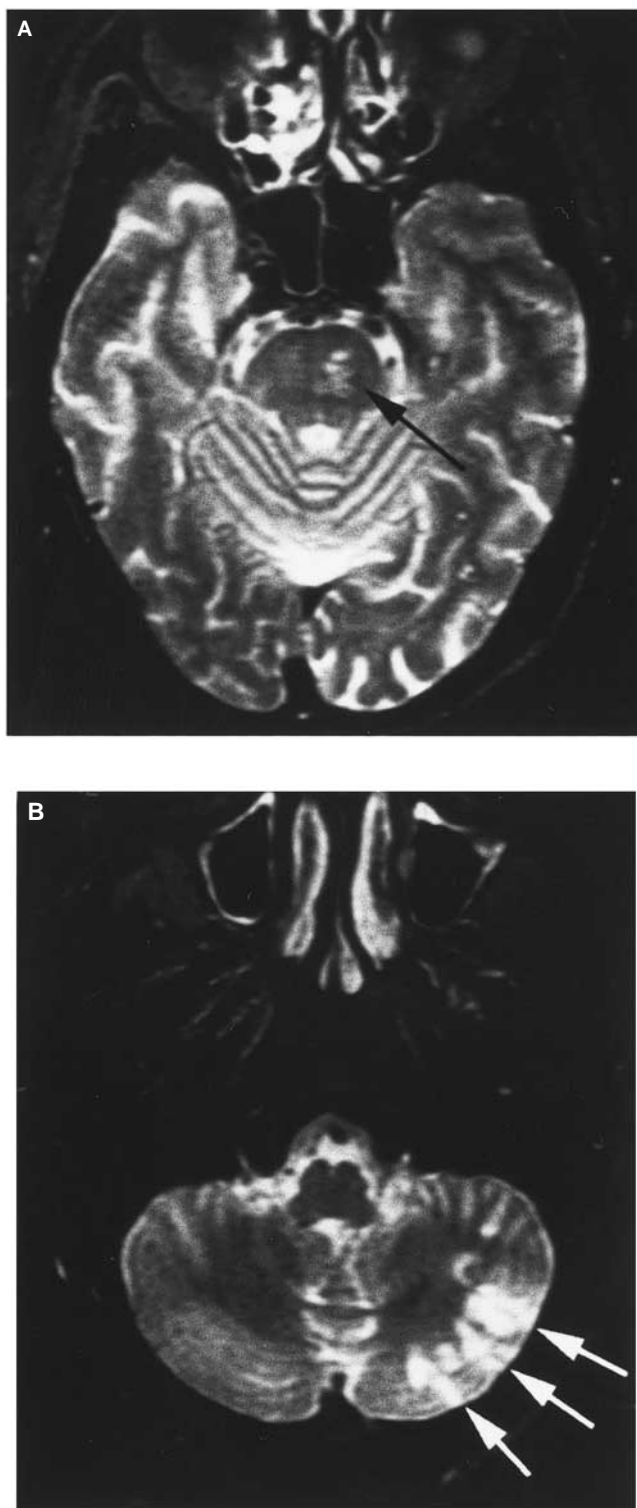


Fig. 1 T₂-weighted magnetic resonance image (MRI) of the patient. The infarction in the left pons is indicated by a black arrow (A). The infarctions in the left cerebellar hemisphere are shown by white arrows (B).

Examination of cerebrospinal fluid detected no abnormality. A brain magnetic resonance imaging (MRI) showed an area of infarction in the left pons and left cerebellum (Fig. 1). A ¹²³I-N-isopropyl-*p*-iodoamphetamine (IMP) single photon emission computed tomogram (SPECT) revealed a slight reduction of blood flow in the left cerebellar hemisphere. Occlusion of both the left posterior inferior cerebellar artery (PICA) and the pontine branch from the basilar artery was suspected. Electroencephalogram (EEG) and auditory brainstem response (ABR) were normal.

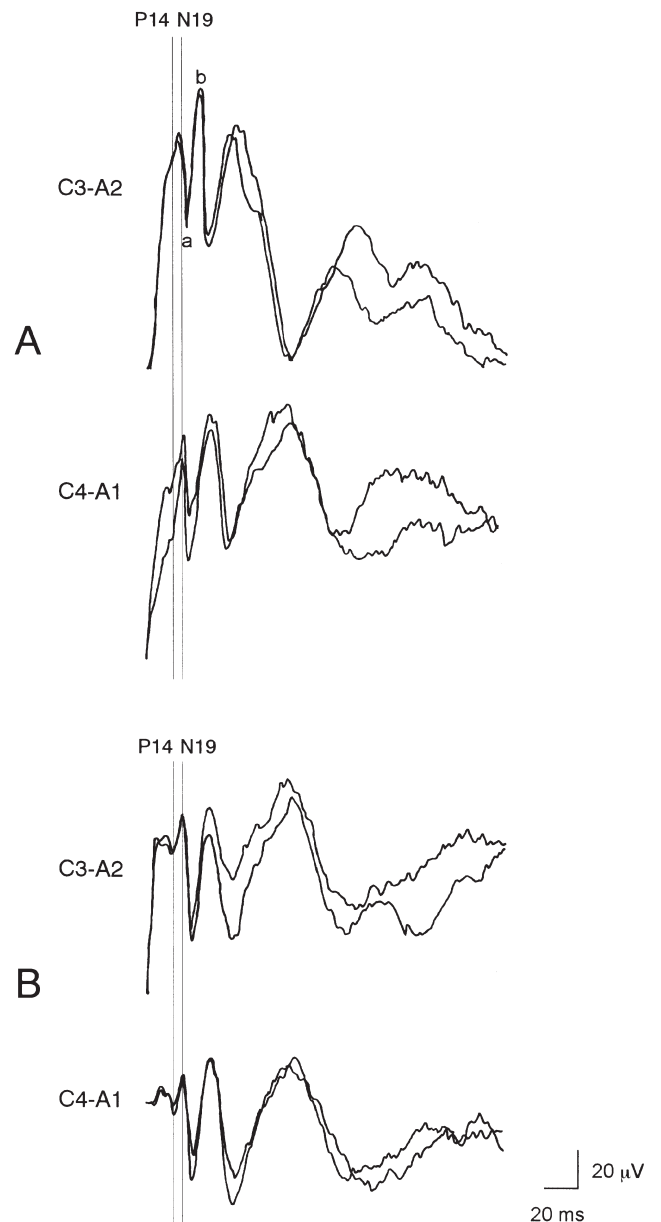


Fig. 2 Somatosensory evoked potential (SSEP) recorded from the patient by median nerve stimulation. The reference was on the ear ipsilateral to the stimulated side. Initial recording indicates the delays of P14 (a) and N19 (b) at C3 by right median nerve stimulation (A). These findings were normalized 4 days later (B). Vertical lines indicate the normal positions of P14 and N19.

Somatosensory evoked potential (SSEP) showed a delay in P14 and N19 recorded at C3 by right median nerve stimulation (Fig. 2A). C-response was not evoked. The abnormal findings in SSEP normalized in 4 days (Fig. 2B). Although administration of clonazepam was stopped at 3 days after admission, myoclonus did not reappear. After 4 weeks in the hospital, the patient was discharged with no neurological deficit. Follow-up MRI was examined 7 months later, and no changes were noted.

DISCUSSION

The patient showed myoclonus in the soft palate, branchial muscles and upper limbs. The responsible lesion for myoclonus was in the brainstem, as evidenced by brain MRI. The location of the lesion was close to the Guillain et Mollaret's triangle,⁴ i.e., the triangle made by the ipsilateral dentate nucleus, contralateral red nucleus and contralateral inferior olive.

Palmer et al. reported a patient with synchronous positive and negative myoclonus due to pontine haemorrhage.⁵ The positive and negative myoclonus was present in the facial and limb muscles. They suggested that a physiologic rhythm generator in the brainstem could have produced bursts of activity in both excitatory and inhibitory descending long fiber tracts, and that this would account for the synchronicity of positive and negative myoclonus.

Our patient exhibited two remarkable features among his neurological signs. First, myoclonus appeared soon after cerebrovascular accident. In a collaborative review from literature of 31 patients with palatal myoclonus and their 2 patients, Matsuo and Ajax found that the appearance of palatal myoclonus was delayed 2 to 49 months after the vascular accident.⁶ They attributed this delay to the development of olivary hypertrophy, which has been proven on the basis of both MRI and pathologic examination.⁷ However, the myoclonus in our patient was characterized by sudden onset, suspected to have occurred soon after the cerebrovascular accident. Segmental myoclonus, then, is not necessarily evoked by olivary hypertrophy. Although patients with segmental myoclonus appearing soon after the presumed cause have been reported, the responsible lesions in these cases were not revealed by radiological study.³ The MRI findings in our patient clearly indicate that brainstem infarction was responsible for the acute onset of segmental myoclonus.

The second important finding in our patient was the transient and benign course of myoclonus, which was accompanied by changes in the SSEP. Small amounts of clonazepam completely relieved the involuntary movement in our patient. Even after administration of clonazepam had ceased, myoclonus did not return. The initial SSEP was abnormal, which suggested a dysfunction in the brainstem. We considered that the left medial lemniscus may have been the lesion responsible for the delayed SSEP. These assumptions were confirmed by the brain MRI study. The abnormal findings of SSEP normalized 4 days later. These findings suggest that the myoclonus in our patient was evoked by a transient dysfunction in the brainstem. Such transient abnormalities of SSEP in patients with segmental myoclonus have not been reported previously. On the other hand, Masucci et al. reported that the prognosis of involuntary movements in patients with brainstem vascular diseases was not good.⁸ However, their patients showed other neurological deficits in addition to segmental myoclonus, and although our patient showed several neurological deficits beyond myoclonus, they were transient. The lesions observed in our patients may constitute the low end of the spectrum in terms of capacity to evoke segmental myoclonus, and this may account for the benign course. Transient myoclonic state with asterixis has previously been reported in elderly patients.⁹ In this report, however, brain MRI analysis was not performed, and some of these patients may have shown small brainstem infarction if examined by MRI.

Myoclonus is a unique neurological finding caused by many different conditions. Those induced by a cerebrovascular accident in the brainstem are thought to be caused by olivary hypertrophy and to be resistant to medication. The benign myoclonus shown in our patient is an important neurological finding because it indicates that a small brainstem infarction can evoke functional abnormalities in neuronal activity, and because complete cessation could be achieved by administration of a small amount of benzodiazepine, specifically, clonazepam.

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Spinal leptomeningeal metastases of giant cell glioblastoma associated with subarachnoid haemorrhage: case report

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Summary A case of subarachnoid haemorrhage (SAH) due to spinal leptomeningeal metastases of a giant cell glioblastoma is described. A 51 year old male presented with a four week history of headache. Neurological examination was normal except for a slight left hemiparesis. Computed tomography (CT) revealed a large cyst with a mural nodule in the right temporal lobe. The tumour was removed followed by 60 Gy of radiation therapy. Thirty-two months later he developed headache and shoulder pain with symptoms of normal pressure hydrocephalus. Head CT showed ventriculomegaly and SAH. Magnetic resonance imaging showed spinal leptomeningeal metastases at the C4–5, T12, and L2 levels, but no local recurrence or tumour dissemination in the brain. He died 34 months after surgery. Autopsy revealed diffuse SAH over the whole brain and spinal cord, associated with spinal leptomeningeal metastases, but no cerebral aneurysms. Spinal radiotherapy and ventriculoperitoneal shunting could possibly have extended survival in this patient. © 2001 Harcourt Publishers Ltd

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INTRODUCTION

Giant cell glioblastoma is a rare variety of glioblastoma multiforme characterised by bizarre multinucleated giant cells with abundant