

Facial movement disorders, which should be distinguished from tremors of the smile, are many and include Parkinsonian isolated jaw tremor, oculomandibular dystonia (Meige's), tardive dyskinesia ("rabbit sign"), myorhythmia,¹ hemifacial spasms, segmental myoclonus (branchial), myokymia, focal motor seizures, Gilles de la Tourette's syndrome and habit spasms. Patients with reading epilepsy may exhibit tremor of the jaw while reading which at times precedes a generalised tonic clonic seizure.² In focal reflex myoclonus, sensory precipitants are evident and a central nervous system lesion is present.³ In contraction fasciculation, subtle volitional contraction of enlarged regenerating motor units in atrophic muscles can be seen in chronic denervating illnesses like amyotrophic lateral sclerosis or poliomyelitis, and may simulate spontaneous tremor of muscle segments.⁴ Because of axonal membrane hyperexcitability in neuromyotonia, muscle contraction may trigger outlasting spasms of delayed relaxation resolving into myokymia and fasciculations mimicking tremor on muscle contraction.⁵ Common variety muscle cramps may resolve into fasciculations.⁶ All these disorders will be properly diagnosed on clinical basis with the aid of radiological or electrophysiological testings.

The aetiology of this patient's condition could not be determined. Radiological studies and electroneuromyography ruled out brain tumour and degenerative or demyelinating illnesses. It is believed that this type of tremor represents a rare benign functional condition, with a slow progression and isolated involvement of the risori muscles; it is of interest that it was triggered by contraction of the muscles in question independent of suprasegmental activating mechanisms, that is, cortical volitional or automatic subcortical. This tremor is better understood as an action or postural tremor rather than intentional or ballistic since it was induced by a particular level of motor unit recruitment and inhibited by maximal volitional contraction. In this context, it represents a form of familial essential benign tremor, a condition of central origin⁷ often manifested in its initial stages as a focal task specific movement disorder of the type of primary writing tremor⁸ or writing tremor myoclonus (Jacome, DE: submitted for publication).

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References

- 1 Mascucci EF, Kurtzke JF, Saini N. Myorhythmia: a widespread movement disorder. *Brain* 1984;**107**(pt 1):53-79.
- 2 Geschwind N, Sherwin I. Language-induced epilepsy. *Arch Neurol* 1967;**16**:25-31.
- 3 Sutton GG, Mayer RF. Focal reflex myoclonus. *J Neurol Neurosurg Psychiatry* 1974;**16**:207-17.
- 4 Daube JR. *Needle Examination in Electromyography*. American Association of Electromyography and Electrodiagnosis. Minimonography No 11, 1979:17.
- 5 Lance JW, Burke D, Pollard J. Hyperexcitability of motor and sensory neurons in neuromyotonia. *Ann Neurol* 1979;**5**:523-32.
- 6 Brown WF. *The Physiological and Technical Basis of Electromyography*. 1st ed. Boston: Butterworth Publishers, 1984:358-9.
- 7 Shahani BT, Young RR. Action tremors: a clinical neurophysiological review. In: Desmedt JE, ed. *Progress in Clinical Neurophysiology*. Vol 5. Basel: S Karger, 1978: 127-37.
- 8 Klawans HL, Glantz R, Tanner CM, Goetz CG. Primary writing tremor: a selective action tremor. *Neurology* 1982;**32**:203-6.

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Paroxysmal sensory-motor attacks due to a spinal cord lesion identified by MRI

Sir: Sudden short lasting tonic attacks, with posture of the limbs resembling tetany, are among the paroxysmal symptoms of multiple sclerosis and were first recognised in 1958 by Matthews,¹ who referred to them as "tonic seizures". We use the term "attack" rather than "seizure" to avoid any confusion with cortical epilepsy. Tonic attacks, either unilateral or bilateral, are often triggered by voluntary movement or by peripheral sensory stimulation. When preceded or associated with sensory symptoms they can be called "sensory-motor attacks"; some-

times they are referred to as "Brown-Sequard syndrome in reverse" when the classical pattern of sensory-motor deficit is replaced by corresponding sensory and motor irritative disturbance.² It has not yet been possible to draw any conclusion as to the site of the lesion responsible for the paroxysmal attacks and there are no reports in the literature of cases studied with Magnetic Resonance Imaging (MRI).

We observed the case of a previously healthy woman, a school-teacher, aged 48 years, who developed paraesthesia (feeling of heat) with sensory deficit for heat, touch and pain on the left of her body up to the level of her neck. The degree of sensory disturbances increased for 3 days, when weakness at the right limbs appeared and gradually increased during the following 4 days. The patient was admitted to hospital on 14 June 1985. Neurological examination showed severe sensory deficit on the left of the body up to C3 dermatome, with impaired sensitivity to heat, touch and pain, sensory deficit on the right of the body up to C3 dermatome, with impaired proprioceptive sensation and sensor ataxia, mild weakness of the right limbs, with brisker deep reflexes, absent abdominal reflexes and extensor plantar response on the right side.

Cerebrospinal fluid examination, myelography, electroencephalography, cerebral CT scan and cerebral MRI were normal. Spinal MRI (21 June) showed a lesion in the cervical medullar parenchyma at the level of C2 on the right side; the lesion was 1 cm long, a few mm wide, with altered signal appearing as a lighter area, particularly in images with prolonged echo-time and was consistent with either ischaemia or a demyelinating lesion (fig).

Two days after her admission the symptoms improved and the patient was eventually put on steroid therapy (betametasone 1 mg a day) for 10 days. She was discharged on 25 June with only slight weakness of the right limbs, brisker deep homolateral reflexes and complete recovery from sensory deficit.

A few days later the patient experienced several paroxysmal sensory-motor attacks characterised by paraesthesia (feeling of heat) in the left leg, immediately followed by stiffening of the right limbs with adduction of the arm and flexion of the forearm; the fingers were flexed at the metacarpophalangeal and extended at the interphalangeal joints. The leg was extended with plantar flexion and inversion of the foot. The tonic attacks of the right limbs were also preceded by homolateral brief feeling of electric shock like cramps. The sensory-motor attacks were triggered by a voluntary

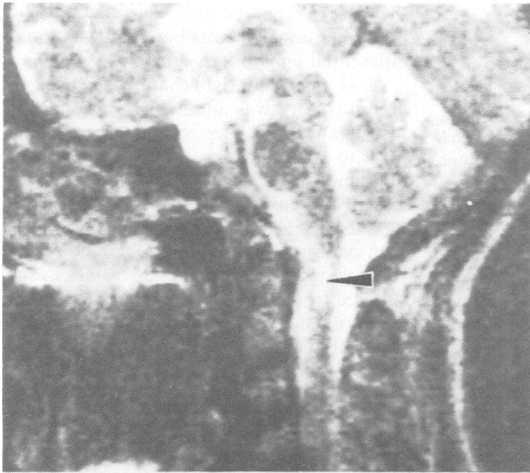


Fig MRI of the spinal cord in the sagittal plane, recorded with a Phillips Gyroscan (a magnetic field strength of 0,5 Tesla) using a spin-echo pulse sequence (echo-time 50 ms). The lesion in the cervical medullar parenchyma is at the level of C2 (arrow = light area)

movement, such as rising and standing, lasted less than 2 minutes and occurred several times a day.

She was admitted to hospital again on 12 July 1985. Neurological examination was unchanged from the previous discharge. Chvostek and Trousseau signs were negative. Several investigations gave normal results (routine laboratory tests, including calcium and magnesium, cerebrospinal fluid examinations for detection of oligoclonal G immunoglobulin bands by agar-gel electrophoresis and by isoelectrofocusing, bilateral subclavian-vertebral angiography, electroencephalography during and between seizures).

Carbamazepine at low doses (100 mg for 3 times a day) completely prevented the attacks. She was discharged on 22 July and since then she has experienced neither attacks nor other neurological disturbances.

Paroxysmal tonic attacks are usually associated with multiple sclerosis³ and seldom with other diseases such as encephalomyelitis^{2,4} and cervical trauma.⁵ There are few reported familial or sporadic cases, associated with choreoathetosis or without any other indications of neurological disease.⁶ The only case in which an ischaemic aetiology was hypothesised⁷ was later recognised to be due to multiple sclerosis.⁸ There is a racial modification of the clinical picture: the tonic attacks tend to be rarer and usually monolateral in Europe and North America and more frequent, often bilateral and painful, in Asian countries and Japan.⁹⁻¹¹ In the only reported case from Africa the tonic attacks were bilateral.¹² Occasionally tonic attacks are observed at the onset of multiple sclerosis.^{1,13-15} The most remarkable finding at electro-

myography is a simultaneous activity of the agonist and antagonist muscles, particularly in the biceps and triceps brachii.¹⁶

Emphasis has been laid on the importance of a spinal cord lesion in the genesis of tonic attacks by Matthews in 1958,¹ who first reported four cases, three of which had shown symptoms referable to Brown-Sequard syndrome at some stage of the disease. Later more attention was given to spinal cord lesions, even though no definitive conclusion as to site of lesion was drawn.^{2,5,9,15,17-19} On the other hand, some authors suggested different sites of lesion, such as the brainstem¹⁴ or the internal capsule,²⁰ while some others admitted the impossibility of finding a precise site due to the presence of multiple lesions.^{1,3,21} Few necropsies have been done on patients with multiple sclerosis and tonic attacks: some of them were inconclusive,^{18,19} while in other cases it was possible to identify spinal cord lesions suspected to be responsible for attacks, even though there were minor lesions in the brainstem and in the cerebral hemispheres.^{8,9}

Ekbohm *et al.*² who reported two cases with paroxysmal sensory-motor attacks, gave the first pathogenetic explanation hypothesising transversely spreading activation of damaged axons in fibre tracts of the spinal cord. Paraesthesia followed by controlateral tonic attacks are explained as diffusion of ephaptic activation from the spinothalamic to the corticospinal tract. This hypothesis was then thought to be operative in other types of paroxysmal attacks in multiple sclerosis.⁵ An alternative explanation could be that during a voluntary movement, when activation along the corticospinal tract reaches the demyelinated

plaques, it is wholly or partially blocked and spreads to the neighbouring axons.²² Pathological evidence in multiple sclerosis plaques of demyelinated axons in contact with each other, without intervening glial tissue has been claimed to support the lateral spread of activation.²³ Accordingly tonic attacks without any sensitive symptomatology could be explained with a demyelinating lesion at any point of the cortical spinal tract, while it could be necessary to have a spinal cord lesion to explain sensory disturbances at one side followed by motor attacks on the contralateral side, because it is only in the spinal cord that sensory and motor tracts run close to each other.

Carbamazepine at low doses was first reported to be promptly effective by Kuroiwa *et al.*²⁴ in 1967 and later universally recognised to be the drug of choice for all the paroxysmal disorders of multiple sclerosis.²⁵ Carbamazepine reduces both sodium and potassium conductance through the axonal membrane.²⁶ Depression of ephaptic transmission through and around demyelinating plaques could be the explanation for the efficacy of carbamazepine in treating paroxysmal disturbances.²⁷

The presence of a single cervical spinal lesion, without any encephalic lesion, observed in our case, corroborates the hypothesis of the spinal origin of the paroxysmal sensory-motor attacks and ephaptic transmission. In contrast with ischaemic lesions in which both axons and myelin are damaged, in multiple sclerosis the axons in the plaques are relatively preserved and it is possible that they lie close together without intervening glial tissue:²³ that could explain the extraordinary frequency of tonic attacks in demyelinating lesions. Hence, in our case, although we did not observe other clinical signs and symptoms indicative of multiple sclerosis, and cerebrospinal fluid examination was normal, we tend to consider as demyelinating the lesion observed at MRI. It is worthwhile noting that a higher incidence of tonic attacks has been reported, in Asian countries and Japan, where lesions due to multiple sclerosis in the spinal cord tend to be more frequent and severe.¹¹

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References

- 1 Matthews WB. Tonic seizures in disseminated sclerosis. *Brain* 1958;**81**:193-206.
- 2 Ekblom KA, Westerberg CE, Osterman PO. Focal sensory-motor seizures of spinal origin. *Lancet* 1968;**1**:67.
- 3 Matthews WB. Clinical aspects of paroxysmal symptoms in multiple sclerosis. In: Matthews WB, ed. *McAlpine Multiple Sclerosis*. Edinburgh: Churchill Livingstone, 1985: 111-7.
- 4 Durston JHJ, Milnes JN. Relapsing encephalomyelitis. *Brain* 1970;**93**:715-30.
- 5 Osterman PO, Westerberg CE. Paroxysmal attacks in multiple sclerosis. *Brain* 1975;**98**:189-202.
- 6 Lance JW. Sporadic and familial varieties of tonic seizures. *J Neurol Neurosurg Psychiatry* 1963;**26**:51-9.
- 7 Castaigne P, Cambier J, Brunet P. Spinal sensory-motor seizures. *Lancet* 1968;**i**:357.
- 8 Castaigne P, Cambier J, Barbizet J, Brunet P, Poirier J. Crises sensitivo-motrices d'origine spinale au cours d'une sclérose en plaques à poussée aiguë terminale. *Rev Neurol (Paris)* 1974;**130**:261-71.
- 9 Shibasaki H, Kuroiwa F. Painful tonic seizures in multiple sclerosis. *Arch Neurol* 1974;**30**:47-51.
- 10 Kuroiwa Y, Hung TP, Landsborough D, et al. Multiple sclerosis in Asia. *Neurology* 1977;**27**:188-92.
- 11 Shibasaki H, McDonald WI, Kuroiwa Y. Racial modification of clinical picture of multiple sclerosis. *J Neurol Sci* 1981;**49**:253-71.
- 12 Dumas M, Girard PL, Ndyaye IP, Gueye M. Manifestations motrices paroxystiques au cours d'une sclérose en plaques chez une Noire africaine. *Bulletin de la Société de Médecine Afrique Noire Langue Française* 1977;**22**:30-4.
- 13 Joyn RJ, Green D. Tonic seizures as a manifestation of multiple sclerosis. *Arch Neurol* 1962;**6**:293-9.
- 14 Castaigne P, Cambier J, Masson M, Brunet P, Lechevallier B, Delaporte P, Dehen H. Les manifestations motrices paroxystiques de la sclérose en plaques. *Presse Med* 1970;**78**,**44**:1921-4.
- 15 Heath PD, Nightingale S. Cluster of tonic spasm as an initial manifestation of multiple sclerosis. *Ann Neurol* 1982;**12**:494-5.
- 16 Toyokura Y, Sakuta M, Nakanishi T. Painful tonic seizures in multiple sclerosis. *Neurology* 1976;**26**:18-9.
- 17 Matthews WB. Paroxysmal symptoms in multiple sclerosis. *J Neurol Neurosurg Psychiatry* 1975;**38**:617-23.
- 18 Kreindler A, Cardas M, Petrescu A, Botez MI. Considérations sur l'étiopathogénie des crises toniques dans l'encéphalomyélite disséminée. *Rev Neurol (Paris)* 1962;**107**,**4**:353-69.
- 19 Kuroiwa Y, Araki S. Lhermitte's sign and reflex tonic spasm in demyelinating diseases with special reference to their localizing value. *Kyushu J Med Sci* 1963;**14**:29-38.
- 20 Watson CP. Painful tonic seizures in multiple sclerosis: localization of a lesion. *Can J Neurol Sci* 1979;**6**:359-61.
- 21 Fabri S, Millefiorini M. Crisi toniche in un caso di sclerosi multipla. *Riv Neurol* 1965;**35**:161-8.
- 22 Matthews WB. Pathophysiology of paroxysmal symptoms in multiple sclerosis. In: Matthews WB, ed. *McAlpine Multiple Sclerosis*. Edinburgh: Churchill Livingstone, 1985:217-9.
- 23 Prineas JW, Connel F. The fine structure of chronically active multiple sclerosis plaques. *Neurology* 1978;**28**:68-75.
- 24 Kuroiwa Y, Shibasaki H. Carbamazepine for tonic seizures in multiple sclerosis. *Lancet* 1967;**i**:116.
- 25 Espir MLE, Millac P. Treatment of paroxysmal disorder in multiple sclerosis with carbamazepine (Tegretol). *J Neurol Neurosurg Psychiatry* 1970;**33**:528-31.
- 26 Schauf CL, Davis FA, Mader J. Effects of carbamazepine on the ionic conductances of myxoloma giant axons. *J Pharmacol Exp Ther* 1974;**189**:538-43.
- 27 Twomey JA, Espir ML. Paroxysmal symptoms as the first manifestations of multiple sclerosis. *J Neurol Neurosurg Psychiatry* 1980;**43**:296-304.

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Can migraine cause multiple segmental cerebral artery constrictions?

Sir: Before computed tomography (CT) scanning, angiography was extensively used in the investigation of patients with migraine, in particular those with complicated forms of migraine. It became clear that in between attacks cerebral angiograms were normal except for the low incidence of structural lesions such as arteriovenous malformations.¹ Likewise during attacks of migraine most angiograms are normal² arguing against Wolff's suggestion that the aura in migraine is due to intracranial artery vasoconstriction.³ However, many authors have pointed out that intra-arterial contrast medium can provoke migraine and many of the limited number of reports of cerebral arterial changes during migraine attacks were induced this way.^{4,5} These reports mainly mention two different patterns, firstly the failure to visualise one or more of the major intracranial arteries^{4,6} and secondly a transient proximal constriction most commonly of the infraclinoid portion of the internal carotid artery.^{5,7-9} Recently there have been three reports which raised the possibility that multiple separate narrowed arterial segments can also be found in migraine.¹⁰⁻¹² We present a fur-

ther case of migraine with such angiographic abnormalities. The possible significance and differential diagnosis is discussed.

A 41 year old right handed Caucasian accounts clerk was admitted to hospital for investigations of menorrhagia. Five years prior to admission she developed attacks of migraine. These occurred two to three times a year and consisted of a unilateral throbbing headache, lasting 6 hours associated with photophobia, phonophobia and recurrent vomiting. During her attacks she suffered from a tingling sensation in the face and both hands. Her father suffered from classical migraine. She also had a history of 17 years intermittent bloody diarrhoea associated with recurrent bilateral iritis, mouth ulcers, and joint pains affecting ankles and elbows. Her bowel disorder had never been investigated but was thought to be ulcerative colitis. A barium enema during her admission was normal and a rectal biopsy showed non-specific proctitis.

After 2 weeks in hospital undergoing gynaecological and gastroenterological investigations she developed sudden severe bifrontal headache, recurrent vomiting, a sensation of flashing lights in both eyes, numbness in her face and hands and profuse sweating. These severe symptoms persisted for 3 days. Initial examination showed neck stiffness but no Kernig's sign, normal fundi, no focal neurological deficits but a blood pressure of 210/100 mm Hg. A subarachnoid haemorrhage was suspected. A CT scan was initially reported as showing evidence of subarachnoid blood over the convexity of the cortices with no evidence of intraventricular or intracerebral bleeding. On review the scan was subsequently felt to be normal. However, four vessel angiography was requested in response to the initial scan report. This revealed no aneurysms but instead showed multiple regions of narrowing affecting mainly the cortical branches of all the cerebral arteries and to a lesser extent the first parts of the anterior cerebral arteries (fig).

A subsequent lumbar puncture was unremarkable; the pressure was 225 mm CSF, there were 3 white blood cells/mm³, 72 red blood cells/mm³, protein was 0.32 g/l and sugar was 4.4 mmol/l. The erythrocyte sedimentation rates were normal (13, 16 and 17 mm/hour). Other routine blood tests were normal including antinuclear factor although her rheumatoid factor was positive at 1/40.

The patient's headache improved but fluctuated over a 7 day period. In view of the possibility of a vasculitic disorder the patient was started on prednisolone 60 mg/day. The headaches continued to



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