It might be argued that this woman suffered from two distinct disorders, initially a personality disorder, and then later and coincidentally developing metachromatic leukodystrophy. However, the changes in her personality are very much of the type described in other cases of adult metachromatic leukodystrophy.[2-4 6 7] The extended period before clearly organic symptoms appeared is, however, atypical. An alternative is to suggest that she had juvenile metachromatic leukodystrophy. However, the pattern of onset, especially the lack of neurological signs after 18 years of illness and the normal EEG, make this highly unlikely.

Adult metachromatic leukodystrophy is a rare condition, with 15 cases reported between 1977 and 1983,[1] and it is therefore not surprising that the natural history is incompletely documented. At present there is no biological marker to distinguish the adult form from other metachromatic leukodystrophy subtypes, although the assay of intracellular cerebroside sulphatase activity[8 9] may prove to be of value when sufficient data have been collected. Thus, when subdividing the disease we should rely more on the pattern of clinical features and investigations, using age of onset and time course as a rough guide only, accepting that in these areas there will be an overlap with other subtypes.

It seems possible that as psychiatrists become more aware of the condition, and of the availability of an enzyme marker test for it, other examples similar to the patient just described may come to light.

Our thanks are due to Dr I Card, High Royds Hospital, for referring this case, Dr Fenson, Guy's Hospital for the enzyme assays, and to the National Hospital Queen Square for the MRI scan. Dr Robin Jacoby kindly provided in-patient care at Bethlem Royal Hospital.

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Tremors of the smile

Sir: It is our observation that facial tremors on muscle contraction can occur in conditions producing generalised tremors; for example, multiple sclerosis, valproate toxicity, alcohol withdrawal and anxiety. Recently we encountered an unusual case of a patient exhibiting isolated bilateral idiopathic tremor of the face induced only by spontaneous or volitional contraction of the risori muscles.

A 27 year old female had a 9 year history of progressive tremor of the perioral facial muscles induced either by spontaneous smiling or volitional contraction. Forceful smiling abolished the tremor and seemingly there was a set point of contraction effort (motor unit recruitment) required for tremor production. Fatigue during contraction and stress increased the intensity (amplitude) of the tremor but reduced its frequency. Unilateral muscle contraction of the risorius induced ipsilateral tremor only. Her late father had a similar lifelong tremor. She had no other neurological symptoms, and examinations were normal except for the tremor described. Computed tomography of the brain with selective thin sectioning of the posterior fossa showed no abnormalities and one magnetic resonance imaging of the brain was normal. She refused spinal fluid examination. Standard electroencephalogram (EEG) was normal and EEG recordings during tremor showed no time-locked cortical potentials (myoclonus). Masseter reflex latencies and amplitudes, blink reflex studies and facial nerves compound muscle action potential amplitudes and latencies were normal. Facial, tongue and masseters electromyograms (EMG) were normal. Synchronous 5–6 Hz tremor with burst duration of 75 to 125 ms and 600–800 μV amplitude was recorded from risori muscles with concentric needle electrodes (TECA CF 25, 26 gauge; TD 20 EMG recording system). Tremor appeared on moderate contraction effort (fig A) and was suppressed by maximal contraction of the muscles (fig C). Unilateral contraction induced ipsilateral tremor (fig B). Right arm median sensory potential was normal. The patient greatly improved with oral propranolol 80 mg a day.

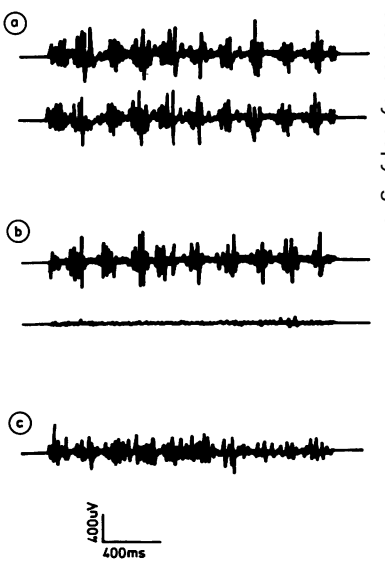

Fig *Tremor recorded with concentric needle electrodes. (A) 5–6 Hz synchronous tremor of risori muscles on moderate effort of contraction. (B) Unilateral tremor of risorius induced by ipsilateral moderate effort contraction. (C) Tremor is suppressed by maximum effort of contraction of risori muscles (only one side shown in the figure).*

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"), myorrhythmia,[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 origin[7] often manifested in its initial stages as a focal task specific movement disorder of the type of primary writing tremor[8] or writing tremor myoclonus (Jacome, DE: submitted for publication).

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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"; sometimes 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