

bilateral flexor plantar responses. There was a mild finger/nose, heel/shin and truncal ataxia. There were no sensory abnormalities and general examination was normal but his height was between the 5th–10th percentile for his age.

Routine investigations at the age of 11 were normal and included a full blood count, ESR, urea and electrolytes, calcium, phosphate, alkaline phosphatase, liver function tests, blood sugar, glucose tolerance test, serum lipids, CK, vitamin B12, folate, thyroid function, prolactin, cortisol, growth hormone, urinary protein and respiratory function tests. The serum magnesium was low at 0.5 mmol/l (normal 0.7–0.95) with a 24 hour urinary magnesium excretion of 1.46 mmol/l (normal 2.1–6.2). The cerebrospinal fluid protein was 1.13 g/l. An ECG revealed right bundle branch block with left axis deviation. An EEG showed a slight excess of slow forms for his age but no evidence of epilepsy. Visual evoked responses were normal. Electromyography (EMG) showed normal motor and sensory conduction velocities. Concentric needle electromyography showed short duration, polyphasic motor unit action potentials. On maximal volition there was a full interference pattern of small amplitude with maximum amplitude of 750 μ V. A right quadriceps muscle biopsy demonstrated a large number of "ragged red fibres" affecting only type I fibres. A CT scan was normal with no evidence of intracranial calcification.

There have been few previous reports of the association of the Kearns-Sayre syndrome with hypoparathyroidism.^{1–5} The hypoparathyroidism may predate or post-date the onset of symptoms and signs typical of the Kearns-Sayre syndrome and may or may not be associated with seizures (table). In the three previous cases reports in which CT scans were performed intracranial calcification was found, localised to the basal ganglia. Seigal *et al*¹ found intracranial calcification in four of eight patients

with the Kearns-Sayre syndrome, one had hypoparathyroidism, one pseudo-hypoparathyroidism, and the remaining two had normal calcium, phosphate and parathormone levels as did all the patients without intracranial calcification.

The absence of basal ganglia calcification in our patient with both hypoparathyroidism and the Kearns-Sayre syndrome may be due to the early treatment with calcium and vitamin D and the return of the serum calcium and phosphate levels to normal. This suggestion would be supported by the observation that intracranial calcification does not occur in secondary hypoparathyroidism due to previous thyroid surgery where treatment is started early with the onset of tetany.³ The predilection of the basal ganglia for calcification in the recognised disorders associated with intracranial calcification may be due to disorders of calcium metabolism, increased vascular permeability, the preferential perfusion of grey matter and the high rate of blood flow to the basal ganglia. Furthermore alkaline phosphatase activity may be regionally elevated in the basal ganglia in patients developing intracranial calcification.⁶

The early treatment of hypoparathyroidism in cases of Kearns-Sayre syndrome is required to control the symptoms of hypocalcaemia and may prevent basal ganglia calcification, as in our case. Whether the calcification in the basal ganglia has a clinical consequence in this syndrome is uncertain and where it is found a careful search for hypoparathyroidism should be made.

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Meige's syndrome and palatal myoclonus associated with brain stem stroke. A common mechanism?

Sir: Meige's syndrome (blepharospasm and oro-facial dystonia), and palatal myoclonus are two uncommon movement disorders whose pathophysiology is poorly understood. Isolated reports have associated each with focal brain stem lesions and have suggested denervation hypersensitivity of brain stem nuclei as the underlying mechanism.^{1–3} We report a patient in whom both Meige's syndrome and palatal myoclonus developed following upper brain stem strokes. A related origin for these movement disorders is postulated.

A 78-year-old right-handed woman, who was recovering from a stroke was first seen because of involuntary facial movements. Five years previously she had experienced the sudden onset of rotational vertigo, vomiting and loss of balance, resulting in falling. A coarse action and intention tremor of the right upper limb appeared after this episode, causing difficulty with writing and bringing food to the mouth. The tremor apparently remained unchanged over the next five years but was totally abolished following the recent stroke. Involuntary facial grimacing and intermittent forced eye closure had also been present for five years, being aggravated by concentration or anxiety. Six weeks previously she developed sudden loss of speech (anarthria) with right hemiplegia. There was

Table

Author	Age of presentation of hypoparathyroidism	Age of presentation of Kearns-Sayre syndrome	Epilepsy	CT scan calcification
Seigal ¹	?	1	Absent	Present
Toppet <i>et al</i> ²	7	9	Absent	—
Sachs ³	?	?	—	Present
Horwitz ⁴	13	11	Absent	—
Pellock ⁵	9	7	Present	Present
Present case	2	6	Present	Absent

some return of power in the right limbs and dysarthric speech within 24 hours and slow improvement occurred thereafter, so that she could walk and dress with assistance. There was a past history of maturity-onset diabetes, hypertension and glaucoma and her medications included prazosin, pindolol, frusemide, tolbutamide and aspirin. Prochlorperazine had been prescribed for vertigo at the time of the first stroke but had not been continued.

Examination showed a frail, elderly woman with intermittent blepharospasm, worsened by ocular examination, bilateral grimacing movements of the face, and a severe spastic dysarthria. The blood pressure was 140/70 mm Hg and general examination was normal. She was alert and cooperative and there was a full range of ocular movements without nystagmus, but with saccadic intrusions on pursuit in all directions of gaze. Continuous rhythmic palatal myoclonus at a rate of 165/min was seen on inspection of the oral cavity, and tongue movements were slow. Facial and palatal sensation were normal. The jaw jerk was not exaggerated. There was a mild residual right hemiparesis involving the face, arm and leg, with hyper-reflexia and a Babinski response on the right side. Dysmetria and dysidiadochokinesia were present in the left arm and leg. The gait was wide-based and hemiplegic, with a tendency to fall to the right. Sensory testing showed no abnormality. A cranial CT scan performed 6 weeks after the second stroke showed a non-enhancing low-density lesion on the left side of the rostral pons (fig 1). Auditory brain stem evoked responses with monaural click stimulation showed attenuation of waves I-V with left ear stimulation, and absence of wave III. With right ear stimulation the I-V interval was prolonged.

The initial episode of vertigo, ataxia and right upper limb tremor is characteristic of an infarct involving cerebellar nuclei or connections with probable involvement of the right dentato-rubral tract in the midbrain. The lesion demonstrated in the pons by CT after the second stroke is likely to represent an infarct resulting from occlusion of one of the perforating branches of the basilar artery and was appropriately placed to produce the right hemiparesis, left-sided cerebellar signs and palatal myoclonus due to involvement of the central tegmental tract. On the other hand, the myoclonus may have developed after the first stroke and not have been noted. The blepharospasm and facial dystonia which developed after the first stroke was typical of Meige's (or Brueghel's) syndrome.⁴⁻⁶

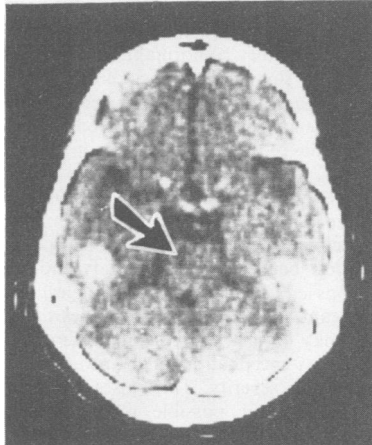


Fig CT scan performed six weeks after the second stroke showing infarct on the left side of the rostral pons (arrow).

Palatal myoclonus may be caused by a variety of lesions involving one limb of the dentato-rubro-olivary (Guillain-Mollaret) triangle, including stroke, demyelination, tumour, arteriovenous malformation, trauma, encephalitis and syphilis.^{7,8} A delay of 2-49 months has been noted in the onset of the myoclonus after acute lesions suggesting that the mechanism of the myoclonus may involve the development of denervation hypersensitivity in olivary neurons.³ An alternative mechanism postulates release of olivary neurons from supra-segmental inhibition.⁹ Meige's syndrome is usually of idiopathic origin and insidious in onset, but there have been reports of blepharospasm, and in some cases associated facial dystonia, developing after unilateral ischaemic or demyelinating lesions of the rostral brain stem or diencephalon.^{1,2,10} In such cases the onset of the involuntary movements was usually delayed for several months after the acute episode, again raising the possibility of a denervation hypersensitivity of facial motor neurons or a release of facial motor neurons from supranuclear inhibition. The association of the two disorders, as in the present case, has been reported only rarely^{2,5} and suggests that similar pathophysiological mechanisms may be involved in the development of the two involuntary movements.

The abolition of the right-sided tremor by the subsequent development of an incomplete corticospinal tract lesion is of interest and indicates that the corticospinal tract is the final common pathway for the expres-

sion of this involuntary movement. Parkinsonian tremor and dyskinesias resulting from basal ganglion lesions may also be abolished by lesions of the lateral corticospinal tract, usually with some degree of resulting hemiparesis.¹¹

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