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 μV. 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. The hypoparathyroidism may predate or postdate 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 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. 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.

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.

References

Accepted 15 February 1986

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. We report a patient who 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

<table>
<thead>
<tr>
<th>Author</th>
<th>Age of presentation of hypoparathyroidism</th>
<th>Age of presentation of Kearns-Sayre syndrome</th>
<th>Epilepsy</th>
<th>CT scan calcification</th>
</tr>
</thead>
<tbody>
<tr>
<td>Seigal¹</td>
<td>7</td>
<td>1</td>
<td>Absent</td>
<td>Present</td>
</tr>
<tr>
<td>Toppel et al²</td>
<td>7</td>
<td>9</td>
<td>Absent</td>
<td>—</td>
</tr>
<tr>
<td>Sachs³</td>
<td>13</td>
<td>11</td>
<td>Absent</td>
<td>Present</td>
</tr>
<tr>
<td>Horwitz⁴</td>
<td>9</td>
<td>7</td>
<td>Present</td>
<td>Present</td>
</tr>
<tr>
<td>Pellock⁵</td>
<td>Present case</td>
<td>2</td>
<td>Present</td>
<td>Absent</td>
</tr>
</tbody>
</table>
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. Pro-
chlorperazine had been prescribed for verti-
tigo at the time of the first stroke but had
not been continued.

Examination showed a frail, elderly
woman with intermittent blepharospasm,
which worsened by ocular examination, bilater-
grimacing movements of the face, and a
severe spastic dysarthria. The blood pres-
sure was 140/70 mmHg and general exami-
nation was normal. She was alert and co-
operative and there was a full range of
ocular movements without nystagmus, but
with sacadic 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
apalatal 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. Dys-
metria and dysdiadochokinesis 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
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 con-
nections 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 pro-
duce the right hemiparesis, left-sided cere-
bellar 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

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 sug-
gest that the mechanism of the myoclonus
may involve the development of
denervation hypersensitivity in olivary neu-
rns.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 associ-
ated facial dystonia, developing after uni-
lateral ischaemic or demyelinative lesions
of the rostral brain stem or diencephalon.10
12 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 hyper-
sensitivity of facial motor neurons or a
release of facial motor neurons from supra-
nuclear inhibition. The association of the
two disorders, as in the present case, has
been reported only rarely13 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 incom-
plete corticospinal tract lesion is of interest
and indicates that the corticospinal tract is
the final common pathway for the expres-

References

1. Jankovic J, Patel SC. Blepharospasm associated
with brainstem lesions. Neurology (Cleve-

2. Lang AE, Sharp JA. Blepharospasm associated
with palatal myoclonus. Neurology (Cleve-
land) 1984;34:1522.

3. Matsuo F, Ajax ET. Palatal myoclonus and
denervation supersensitivity in the central

4. Meige H. Les convulsions de la face: une forme
clinique de convolution faciale, bilatérale et

5. Jankovic J, Ford J. Blepharospasm and
orofacial-cervical dystonia: clinical and phar-
macological findings in 100 patients. Ann

6. Marsden CD. Blepharospasm-ormandibular
dystonia syndrome (Brueghel’s syndrome). J

7. Nathanson M. Palatal myoclonus. Arch Neurol

8. Herrmann C, Crandall PH, Fang HCH. Palatal
myoclonus. Neurology (Minneapolis) 1957;
37:31–51.


10. Powers JM. Blepharospasm due to unilateral
diencephalon infarction. Neurology (Cleve-

11. Adams RD, Victor M. Tremor, myoclonus,

Accepted 8 August 1985
Meige's syndrome and palatal myoclonus associated with brain stem stroke. A common mechanism?

T J Day, R B Lefroy and F L Mastaglia

J Neurol Neurosurg Psychiatry 1986 49: 1324-1325
doi: 10.1136/jnnp.49.11.1324