Sub-acute presentation of Morvan’s syndrome after thymectomy

A 70 year old male presented in February 2000 with persistent cough. Chest radiograph revealed a mediastinal abnormality. A computed tomography (CT) scan of the thorax confirmed an anterior mediastinal mass. CT guided mediastinal biopsy was performed but proved inconclusive. The patient declined further invasive investigation for 2 years. After follow up CT showed significant enlargement he underwent surgery in September 2002. An excellent post operative recovery was made. Histology confirmed a minimally invasive thymoma.

However, 6 weeks later he developed progressive proximal leg weakness, lethargy, and night sweats. He described a sensation like “someone touching his legs” and became agitated and restless at night. He was re-admitted in November 2002 and on the first night became acutely confused and markedly disorientated with visual hallucinations.

On examination he was agitated and confused. His eyes opened spontaneously, there was no coherent verbalisation, and he withdrew to pain. He was apyrexial although there was marked hyperhidrosis and hyper-salivation. Blood pressure was 120/70 mm Hg, and pulse was 98 bpm and regular. He had profuse diarrhoea although his abdomen was soft and non-tender. There were irregular myoclonic jerks of all four limbs predominating in his upper limbs as elbow flexion jerks. There was no startle reaction. Cranial nerve examination was unremarkable. Power and deep tendon reflexes were normal and plantar responses flexor. Intravenous phenytoin was administered with resolution of the myoclonic movements.

Laboratory investigations including electrolytes, auto-antibody screen, and antineuronal antibodies were unremarkable. Cerebral spinal fluid (CSF) was clear, colourless, and acellular; CSF glucose was 4.0 mmol/l (2.2–4.4) (serum 6.7), and CSF protein 0.25 g/l (0–0.40). Oligoclonal bands were negative in CSF and serum and no abnormality of the immunoglobulin G pattern was detected. CSF analysis for the 14-3-3 (Prion) protein was negative.

Chest radiograph showed postoperative changes. Brain magnetic resonance imaging (MRI) disclosed multiple areas of periventricular and subcortical white matter signal change especially within the right parietal region, felt likely to represent diffuse cerebrovascular changes.

Serial electrocardiograms (ECGs) demonstrated sinus tachycardia. Serial electroencephalograms (EEGs) showed diffusely slow activity only. Nerve conduction studies revealed mild motor conduction slowing (right median 49 m/s, right common peroneal 45 m/s). Sensory nerve amplitudes were small with normal velocities. Repetitive discharges were noted following evoked compound muscle action potential in upper and lower limb motor nerves. Doublet, triplet, and multiplet were recorded in abductor pollicis brevis, tibialis anterior, and extensor digitorum brevis indicating motor axon membrane instability compatible with neuromyotonia (Fig 1).

Confusion, visual hallucinations, insomnia, anxiety, sweating, diarrhoea, a slurred dysarthria, and abnormal muscle activity fluctuated dramatically from day to day, but there was a steady overall deterioration. In a lucid interval he scored 68/100 using Addenbrooke’s bedside testing of cognitive function (equivalent to a Mini Mental State Examination (MMSE) score of 18/30), performing poorly in orientation, attention/concentration, verbal fluency, and visuospatial abilities. He became chair bound despite little clinical weakness either proximally or distally, and later bed bound. Although never as prominent as on first admission, there was frequent muscle twitching, at best resembling myokymia, at worst multifocal myoclonus. He was transferred to a high dependency unit.

The diagnosis of Morvan’s syndrome was based on the clinical phenotype and supported by the nerve conduction studies and the presence of antibodies to voltage gated potassium channel antibodies (VGKC) (165 pmol; normal <100 pmol), 10 weeks post-thymectomy.

At 10 weeks post-thymectomy he had 5 days of plasma exchange (PE). There was rapid improvement in orientation and memory and a cessation of abnormal muscle movements within 5 days. Prednisolone was commenced 40 mg daily and phenytoin continued. He made a dramatic clinical recovery, standing unassisted 10 days post-exchange and mobilising independently by 14 days, with no further muscle twitching. Addenbrooke’s test 2 weeks post-PE revealed a score of 78/100 (MMSE 24/30).

Post-PE nerve conduction studies and EMG showed an increase in the motor conduction velocities (right median 56 m/s, right common peroneal 45 m/s) and resolution of the previous features of neuromyotonia. Repeat serum VGKC antibody levels at 1 and 4 months post-PE were within normal limit. EEGs both 2 and 6 months after treatment showed little improvement. MRI 6 months after treatment also showed no change.

At 8 months post-PE, on 10 mg prednisolone daily he remained asymptomatic aside from fatigue. His wife reported that apart from irritability his personality and memory were essentially back to normal, although on repeat Addenbrooke’s he scored only 82/100. He has even returned to playing golf.

Discussion

Since Morvan’s original publication only a few cases have been described. The majority appear to be a paraneoplastic manifestation.3,4 The discovery of VGKC antibodies in several of these cases, as seen in many cases with neuromyotonia,5 has suggested that Morvan’s syndrome may be an autoimmune disorder. Whether VGKC antibodies play a pathogenic role in the encephalopathy as they do in the peripheral nervous system is as yet unclear. Others3,4 have suggested that the VGKC antibodies may cross the blood–brain barrier and act centrally, binding predominantly to thalamic and striatal neurones6 causing encephalopathic and autonomic features. The reversibility of the encephalopathy with plasmapheresis does suggest that the encephalopathy is also mediated by serum factors. Liguori et al reported the presence of weak CSF oligoclonal bands, absent in the serum, supporting a central neurological role.7 There are also

Figure 1  [A] Upper trace: recordings (superimposed) from abductor pollicis brevis following stimulation of the median nerves at the wrist. Note the repetitive discharges that completely obscure any F responses which may have been generated. Lower trace: following treatment the repetitive discharges have completely disappeared and normal F waves are now visible. (B) Similar to A; recordings before (upper trace) and after (lower trace) treatment from extensor digitorum communis following stimulation of the common peroneal nerve at the ankle. The repetitive discharges subsided following treatment. (C) Upper trace: showing the F responses to be seen; (C, D, E) upper trace: bursts of neuromyotonic discharges in the form of multiplets (C and D) and doublets (E) with intraburst frequencies of up to 120 Hz. C and D are bursts of the same motor unit displayed at different sweep speeds.

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reports of non-paraneoplastic limbic encepha-
litis associated with raised serum VGKC,
suggesting that these antibodies may give rise to a spectrum of neurological disease pre-
senting with symptoms arising peripherally,
centrally, or both.1 However in our case and
the case reported by Lee et al1 oligoclonal
bands were absent in CSF and serum, and
CSF immunoglobulin profiles were un-
remarkable.
The natural history of Morvan’s is highly
variable. Two cases have been reported
to remit spontaneously. In one of these cases
although in one of these cases the patient
died shortly after receiving PE.2 Other fatal-
ities without remission have been described by,
amongst others, Morvan himself. Cardiac
involvement in some cases may increase
vulnerability to sudden death.
Thymectomy has previously been a pro-
posed treatment for Morvan’s syndrome.
This is the first reported case of Morvan’s
syndrome presenting post-thymectomy.
Morvan’s syndrome normally presents with
a slow insidious onset over months to
years.1,3 Our case is unique in that presenta-
tion was over days, just weeks post-thymec-
tomy, and responded to a single PE course
with low dose immunosuppression. We hypothesise that surgery may have precipi-
tated a rise in the serum VGKC antibodies
levels, which were cleared by one course of
PE resulting in remission as supported by the
drop in serum VGKC levels. Although poten-
tially a low risk of thymectomy, it is an
important complication to recognise because
of the dramatic reversibility to treatment.

Case report
A 34 year old Nepali construction worker,
with no past medical history, presented with
suprapubic pain because of acute urinary
retention and was admitted for observation.
In the weeks before admission he had
complained of mild headache and dizziness.
The headache responded well to paracetamol.
There was no history of oregonital ulcers, and
he had received no recent vaccinations.
On admission, he was conscious and alert,
with normal cognitive examination. There
was no neck stiffness. Fundoscopy, pupil
reflexes, accommodation, and vertical and
horizontal eye movements were normal. Bulbar
function was intact. There was general-
ised hyperreflexia, including the jaw jerk,
with equivocal plantar responses. There was
no indication of spinal cord or cauda equina
syndrome on sensory examination. Co-
ordination and gait were normal. He had
urinary retention (550 ml), which was
managed with an indwelling catheter.
General examination revealed an afebrile
patient with normal vital signs. Skin testing
for tuberculosis was negative. Peripheral
blood analysis showed normal findings: the
eythrocyte sedimentation rate was 4 mm/h
and the white blood cell count was 8500/μl
(60% neutrophils and 27% lymphocytes).
Magnetic resonance imaging (MRI) of the
spinal cord including the cauda equina
was normal, as MRI of the brain stem
revealed regression of the hyperintense
lesions on the T2 weighted images.

Comment
Although this patient was likely to have
suffered from a mild probably viral rhomben-
cephalitis, the true diagnosis remains unclear.
We speculate that parainfectious
demyelination or direct viral invasion was
likely to have been the cause of the patient’s
neurological presentation. Intriguingly,
a similar case was reported by Komiyama
et al., in which a 30 year old man had urinary
retention, mild horizontal gaze paresis, and
hypesthesia around the mouth and fingers.4
In their case, the MRI findings revealed
several amorphous lesions in the pons and
cerebellum, in addition to a well defined
lesion in the right dorsolateral tegmentum of
the upper pons. CSF analysis showed a mild
increase in protein but only 3 cells/μl.5 In
contrast to their patient, the lesions in our
case were bilateral and extended from the
medulla to the pontomedullary junction, but
showed a similar rate of disappearance (four
weeks).

Brain stem control of micturition has been
reviewed by Sakakibara and Fowler.6 Urinary
retention is extensively described in patients
with brain stem tumours or strokes. In the
series of patients with brain stem tumours,
the majority of the lesions were located in
the pons and medulla, while in patients with

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Long term continuous bilateral pallidal stimulation produces stimulation independent relief of cervical dystonia

Idiopathic cervical dystonia is the most common form of focal dystonia in adults. It is characterised by involuntary, sustained contractions of the cervical muscles and produces abnormal head movements or postures. Although deep brain stimulation (DBS) of the globus pallidus internus (GPI) is now accepted in the treatment of a wide spectrum of dystonias including cervical dystonia (for a review, see Krauss spectrum of dystonias including cervical dystonia is now accepted in the treatment of a wide

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Severe vasculitic neuropathy following influenza vaccination

Current Department of Health (UK) guidelines suggest that all people aged over 65 years should receive annual influenza vaccination. There is a range of adverse medical sequelae associated with this. Two serious complications are Guillain-Barré syndrome1 and systemic vasculitis.2 The strength of association between influenza vaccination and Guillain–Barré syndrome has been questioned, and little is known of the pathogenesis. Several investigators have reported a close temporal association between influenza vaccination and systemic vasculitis of various classifications.3,4 Vasculitic peripheral neuropathy was not a feature in any of the reported cases.5

We report a case of biopsy proven vasculitis, presenting as mononeuritis multiplex, following influenza vaccination. The clinical picture evolved rapidly into a syndrome indistinguishable from axonal Guillain–Barré syndrome. This suggests a differential diagnosis for post-vaccination neuropathy, with implications for management. We believe this is the first report in which there was an associated peripheral neuropathy at presentation. It raises issues about the aetiology and pathogenesis of vaccination associated neuropathy.

Case report

A previously fit and active 72 year old woman was admitted after routine influenza vaccination. She gave a history of progressive distal arm and leg symptoms, tiredness, and anorxia. One week after vaccination she experienced pain in her left buttoc which radiated down her left leg. In the following 24 hours she developed weakness in her right hand in a median nerve distribution. This was followed by similar symptoms in the left hand. In the 24 hours before admission her left foot became numb and weak and she complained of general exhaustion and anorexia. There was no previous history or relevant family history of neurological disorder, and the only drug she used regularly was pravastatin, which she had been taking for three months. She gave a history of progressive distal weakness, sensory loss, and hyporeflexia of all four limbs, suggestive of a mononeuritis multiplex.

In the 24 hours before admission her left foot became numb and weak and she complained of general exhaustion and anorexia. There was no previous history or relevant family history of neurological disorder, and the only drug she used regularly was pravastatin, which she had been taking for three months. She gave a history of progressive distal weakness, sensory loss, and hyporeflexia of all four limbs, suggestive of a mononeuritis multiplex.

Examination revealed bilateral median nerve palsies and left lower limb sensory loss consistent with abnormality of the distal sciatic nerve. Cranial nerve examination, including fundoscopy, was normal. General examination revealed a purpuric rash on both ankles. There was no evidence of involvement of other organs.

On admission the erythrocyte sedimentation rate (ESR) and C reactive protein were raised at 105 mm/h and 35 mg/l respectively. Full blood count, urea and electrolytes, uric acid, thyroid function, liver function (apart from reduced albumin (34 g/l)), total protein and electrophoresis, calcium, creatine kinase, glycosylated haemoglobin (HbA1c), hepatitis B surface antigen, vitamin B12, and folate were all normal. A vasculitic screen—including antinuclear antibody, perinuclear and cytoplasmic antineutrophil cytoplasmic antibody, and rheumatoid factor—was negative. Influenza A and B complement fixation tests were negative. Urinalysis was normal. The patient declined lumbar puncture.

Nerve conduction studies carried out on admission confirmed a peripheral neuropathy with features of a mononeuritis multiplex syndrome. Motor nerve studies are shown in table 1. Sensory conduction was preserved in both upper and lower limbs.

A skin biopsy of one of the purpuric ankle lesions showed prominent vasculitis involving the small arteries without granuloma formation. On radial nerve biopsy there was severe axonal loss but no definite vasculitis. No evidence of myelin debris was found.

She was treated with high dose oral steroids, azathioprine, and monthly pulses of intravenous cyclophosphamide. Six weeks later there was evidence of further clinical deterioration, although inflammatory markers were low (ESR 21 mm/h). She had severe distal weakness, sensory loss, and hypesthesia of all four limbs, suggestive of involvement of all peripheral nerves. She was very disabled, unable to walk or use either hand.

The electrophysiological findings at this stage were indistinguishable from a severe acute motor and sensory axonal neuropathy of Guillain–Barré syndrome type (table 1). Electrophysiological studies demonstrated complete denervation in the small hand muscles bilaterally and severe partial denervation in lower limb muscles (the gastrocnemius and tibialis anterior were tested). All sensory action potentials were absent (radial, median, ulnar, and sural groups; tested antidiromically). F waves were normal at presentation; however, they were absent in the 24 hours before admission.

Table 1: Motor nerve conduction studies

<table>
<thead>
<tr>
<th>Nerve</th>
<th>Study</th>
<th>DML (ms)</th>
<th>CV (m/s)</th>
<th>Amplitude (distal/proximal) (mV)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Right median</td>
<td>1</td>
<td>4.0</td>
<td>52</td>
<td>1.0/1.5</td>
</tr>
<tr>
<td></td>
<td>2</td>
<td>6.8</td>
<td>–</td>
<td>0.2/0.0</td>
</tr>
<tr>
<td></td>
<td>3</td>
<td>Absent</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>Left median</td>
<td>1</td>
<td>3.8</td>
<td>–</td>
<td>1.0/0.0</td>
</tr>
<tr>
<td></td>
<td>2</td>
<td>Absent</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>Right ulnar</td>
<td>1</td>
<td>3.0</td>
<td>61</td>
<td>7.0/7.0</td>
</tr>
<tr>
<td></td>
<td>2</td>
<td>3.2</td>
<td>62</td>
<td>7.0/7.0</td>
</tr>
<tr>
<td></td>
<td>3</td>
<td>Absent</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>Right medial popliteal</td>
<td>1</td>
<td>5.2</td>
<td>40</td>
<td>6.0/6.0</td>
</tr>
<tr>
<td></td>
<td>2</td>
<td>4.6</td>
<td>41</td>
<td>0.5/0.5</td>
</tr>
<tr>
<td></td>
<td>3</td>
<td>Absent</td>
<td>–</td>
<td>–</td>
</tr>
</tbody>
</table>

Amplitude, peak to peak amplitude of compound muscle action potential; study 1, at presentation; study 2, two weeks later; study 3, six weeks later.

CV, conduction velocity; DML, distal motor latency.

Comment

Interestingly, long term (about 22 months) continuous bilateral GPI DBS with IPGs produced stimulation independent relief of this patient’s cervical dystonia, although short term (less than 24 hours) was associated with limited battery life and discontinuous DBS with the external transmitter had not yielded such beneficial effects. We cannot exclude the possibility that spontaneous remission of the cervical dystonia occurred during the use of IPGs. However, our experience with this patient suggests that chronic GPI DBS may result in normalisation of the altered basal ganglia related motor circuits that are implicated in the occurrence of dystonia, and that over time the normalised state may persist independently of DBS. Alternatively, chronic GPI-DBS may lead to the reorganisation of the functional anatomy of the motor circuits at unknown levels, thus resulting in suppression of dystonia.

Long term remission of idiopathic cervical dystonia after BTX treatment has also been reported.7 It is presently unknown whether peripheral BTX injection and GPI-DBS share a common mechanism in producing remission of cervical dystonia. From a practical standpoint, in patients undergoing GPI-DBS for cervical dystonia treatment, it may be necessary to add DBS as continuously as possible and to avoid any unnecessary discontinuation of the IPGs. In addition, when battery depletion makes it necessary to revise the IPGs, it may be advisable to determine whether further stimulation is actually needed in these patients.

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isolated cranial nerve palsies are often attributed to lesions of the respective nerves along their extraaxial courses. However, ischaemic or haemorrhagic lesions of the brainstem also cause isolated cranial nerve palsies through involvement of the intraxial segments of the respective nerves. Small lesions of the brainstem can affect the trigeminal nerve only, although isolated trigeminal nerve palsy occurs less frequently than third or sixth nerve palsies. A case of small medullary infarction was reported in this journal, involving the spinal trigeminal tract and nucleus and presenting as painful trigeminal sensory neuropathy. In our report, the patient presented with orofacial pain and abnormal sensation both of which were confined within ipsilateral maxillary (V2) and mandibular (V3) nerve distribution. A similar isolated sensory distribution in the V2 and V3 dermatomes was observed in our patient with a small dorsolateral midpontine infarction involving a principal sensory trigeminal nucleus. Our case was unique because light touch sensory deficit was restricted to the V2 and V3 dermatomes while the V1 dermatome remained intact. In a small number of previous reports, dorso-lateral or entry zone infarction or bleeding at the midpons presented as isolated trigeminal nerve neuropathy which involved all trigeminal dermatomes, although some cases tended to show lesser involvement of the V1 dermatome and more marked involvement of the V2 and V3 areas.

**Case report**

A 51 year old man noticed a rubbing sensation on the right edge of his tongue. Then 9 days after onset, this abnormal sensation extended to the right half of his oral cavity including the tongue, lips, upper and lower gums, and the hard palate. An evident boundary existed between the impaired sensory area (V2 and V3) and the intact sensory area (V1). Pinprick and temperature sensation were well preserved in the V2 and V3 areas. Slight weakness of the right masseter was detected. All modalities of sensation were preserved in the ophthalmic nerve division (V1). Corneal reflexes were normal. On examination of other cranial nerves we found no abnormalities, including eye movement and taste. Motor weakness of the trunk and the four extremities was not found. Jaw jerk and deep reflexes in the four limbs were normal. Truncal and limb ataxia was not seen. A blink reflex study showed no delayed latency of ipsilateral R1, R2, or contralateral R2 responses on stimulation of the left or right supraorbital nerve (V1). Brain

**Isolated hypesthesia in the right V2 and V3 dermatomes after a midpontine infarction localised at an ipsilateral principal sensory trigeminal nucleus**

Isolated cranial nerve palsies are often attributed to lesions of the respective nerves along their extraaxial courses. However, ischaemic or haemorrhagic lesions of the brainstem also cause isolated cranial nerve palsies through involvement of the intraxial segments of the respective nerves. Small lesions of the brainstem can affect the trigeminal nerve only, although isolated trigeminal nerve palsy occurs less frequently than third or sixth nerve palsies. A case of small medullary infarction was reported in this journal, involving the spinal trigeminal tract and nucleus and presenting as painful trigeminal sensory neuropathy. In our report, the patient presented with orofacial pain and abnormal sensation both of which were confined within ipsilateral maxillary (V2) and mandibular (V3) nerve distribution. A similar isolated sensory distribution in the V2 and V3 dermatomes was observed in our patient with a small dorsolateral midpontine infarction involving a principal sensory trigeminal nucleus. Our case was unique because light touch sensory deficit was restricted to the V2 and V3 dermatomes while the V1 dermatome remained intact. In a small number of previous reports, dorso-lateral or entry zone infarction or bleeding at the midpons presented as isolated trigeminal nerve neuropathy which involved all trigeminal dermatomes, although some cases tended to show lesser involvement of the V1 dermatome and more marked involvement of the V2 and V3 areas.

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Figure 1 (A) Axial T2* weighted image, (B) axial T2 weighted image, and (C) coronal T2 weighted image, taken 2 weeks after onset, showing a hypointense lesion (arrow) in the right dorsolateral pontine tegmentum. Extrapontine trigeminal nerves are indicated by arrowheads. (D) A sketch of the intrapontine and extrapontine segments of trigeminal nerves and the trigeminal nuclei. The white thick line indicates the axial slice level used to obtain images (A) and (B). (E) A mid-sagittal view image with a white thin line indicating slices (A) and (B). (F) A neuroanatomical sketch corresponding to the MR imaging at slice ‘‘C’’. 1, principal sensory trigeminal nucleus; 2, motor trigeminal nucleus; 3, middle cerebellar peduncle; 4, superior cerebellar peduncle.
A characteristic of our case was the restricted sensory impairment in the V2 and V3 dermatomes. The distribution of sensory deficits in our case, a dorsolateral pontine infarction, was very similar to that in a patient with a small medullary infarction reported by Nakamura et al. Our patient presented a rubbing sensation and decreased light touch sensation without pain, while the previous case presented orofacial pain and sensory deficit. Root fibres conveying impulses for pain and temperature senses enter the principal sensory nucleus, while root fibres conveying impulses for pain and temperature senses enter the spinal trigeminal nucleus. The lesion in our case involved the intermediate nucleus in which fibres of V2 and V3 enter, while the previous case involved the tract or nucleus related to pain and temperature senses, because it was relatively small and located in a dorsolateral area of the midpons. For this reason, pain and temperature senses were preserved in our patient.

The size and location of the infarction in our case was very similar to that in a patient reported by Ishii et al. The sensory impairment in our case was restricted to the V2 and V3 dermatomes, while the previous case presented numbness and paresthesia over the left upper face, tongue, and buccal mucosa. The motor trigeminal nucleus was impaired in our case but not in the previous case. The lesion in our case was in a slightly dorsal location compared with the lesion of the previous case. The motor trigeminal nucleus was presumably affected in our case, since it is located on the dorsomedial side of the principal sensory trigeminal nucleus.

The most interesting finding of our case is that an evident boundary existed between the impaired sensory area (V2 and V3) and the intact sensory area (V1). As yet, it is not known whether or not a small lesion of the principal sensory nucleus can cause hypalgesia limited to the V2 and V3 areas based on “segmental distributions” of the face. Functional MRI activation was studied on brush or noxious stimulation of the V1, V2, and V3 areas of the face. However, somatotopic activation in the principal sensory trigeminal nucleus has been unsuccessful. According to Carpenter, neurons carrying sensation from the V1 area are localised in the most ventral part of the principal sensory nucleus, while the V2 and V3 areas are located more dorsally. The explanation for the deterioration of the tactile sense being limited to the V2 and V3 areas is that the lesion in our case involved the intermediate and dorsal portions of the principal sensory nucleus in which fibres of V2 and V3 terminate.

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