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**Spinal angiomatosis presenting as a sensory neuropathy**

Sir: Spinal angiomatosis is a rare condition which usually presents with progressive symptoms suggesting a radiculomyelopathy. It may also cause subarachnoid haemorrhage. The usual presentation of our patient with symptoms suggestive of a sensory neuropathy appears unique.

An Irish building labourer, aged 55 years, was admitted to hospital because of progressive tingling of his legs and unsteadiness in walking for ten days. He had previously had any significant illness and there was no family history of neurological disease. There was no suggestion of any exposure to known neurotoxins. His alcohol consumption was considered moderate, but he smoked 80 cigarettes daily. He appeared unkempt, with marooned, carious teeth. In other respects general medical examination was normal. His intellectual function had always been below average and he was vague about the details of his medical history. The cranial nerves and tone and power of his limb muscles were normal. All the tendon reflexes were reduced, while the right biceps, left knee and ankle reflexes were absent, but plantar responses were flexor. Modalities of sensation were impaired in the fingers as far as the metacarpophalangeal joints, and in the feet, as far as the ankles. Position sensation was impaired at the toes and ankles. He was unable to stand still with his eyes closed, and his gait was wide based and unsteady. Extensive haematological, biochemical and radiological investigation for a possible carcinoma gave normal findings. A CT scan of the brain was normal. Nerve conduction studies showed normal motor conduction velocities but reduced sensory action potentials (right radial 1 mV; right sural 2 mV; left sural absent). The F latency to the right thenar muscles was 34 ms and to the left abductor hallucis was 55 ms. Lumbar puncture revealed a clear fluid containing no cells.

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**References**

8. Botazzo GF, Vendelli C, Mirmiran R. The detection of autoantibodies to discrete

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**Table**

Unselected cases tested by IFL on three different pituitary substrates

<table>
<thead>
<tr>
<th>Pituitary substrates</th>
<th>Total cases tested</th>
<th>Positive reactions</th>
</tr>
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<tbody>
<tr>
<td>Normal human</td>
<td>100</td>
<td>17</td>
</tr>
<tr>
<td>Baboon</td>
<td>100</td>
<td>18</td>
</tr>
<tr>
<td>Rhesus monkey</td>
<td>51</td>
<td>28</td>
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and protein 1.6 g/l. Sural nerve biopsy showed gross loss of myelinated fibres and an excess of collagen. The appearance of axonal degeneration was confirmed by electron microscopy, which also showed occasional demyelinated axons. Sensory loss gradually became more severe and extended proximally. After one month the reflexes all disappeared, including the plantar responses which had remained flexor until that time. In addition, weakness of the intrinsic hand muscles and hip flexion developed. The neurological deficit continued to increase and, after three months, wasting of the intrinsic hand muscles, tibialis anterior and peroneal muscles had become evident. Finger clubbing had also developed. By this stage the right median motor conduction velocity slowed to 40 m/s, the median nerve action potential—which had been 11 μV disappeared completely and the F wave latency remained 36 ms. A course of prednisolone and azathioprine failed to influence the progress of the condition. After six months, ataxia of the limbs became evident with pseudo-athetosis of the outstretched fingers. After eleven months he became incontinent of urine and after a year he died from a chest infection.

At necropsy severe bronchitis and bronchopneumonic consolidation were found in both lungs. In the ascending aorta there was a 1 cm horizontal tear, as is associated with dissecting aneurysm, but dissection had not occurred. Other systems were normal, apart from the nervous system to be described. The cerebrum, brain stem, cerebellum, pons, medulla and cranial meninges were normal on both gross and histological examination. The spinal theca was somewhat thickened, while a number of prominent tortuous vessels were noted under the leptomeninges covering the spinal cord from its mid cervical to lower thoracic regions. The spinal cord itself was distorted and softened in its mid thoracic part. The roots in the cauda equina were noted to be either of normal pale-cream colour or pinkish-grey in approximately equal proportion. The sciatic nerve was remarkably fibrous in texture and grossly resembled a frayed tendon.

Histology showed that both the spinal theca and leptomeninges were thickened: iron deposits (Perl's ferrocyanide test) in these membranes suggested that the thickening had resulted from previous haemorrhage. There was an excess of leptomeningeal vessels with arterial structure, while the veins were dilated, tortuous and markedly more collagenous (fig a) than normal veins around the cord. One of these leptomeningeal veins contained a small organising thrombus filling half its lumen. Numerous dilated tortuous thick walled small collagenous vessels were seen within the cord (fig b) and were particularly associated with the necrosis (myelomalacia) of the posterior columns. The upper thoracic cord was cavitiated at its centre and resembled a syrinx but the cavity was not lined with epithelium and was clearly necrotic in origin. The region of the right anterior spinocerebellar tract showed a modest degree of pallor. Perl's ferrocyanide test was negative in the cord, indicating the absence of previous haemorrhage. The dorsal spinal roots contained frequent dilated sinuses lined by endothelium only; a considerable number (up to 90%) of nerve fibres were lost in many dorsal roots. Ventral spinal roots showed less loss of nerve fibres. The sciatic nerve examined was partly replaced by collagen and showed substantial fibre loss.

Spinal angiomatosis—originally described by Foix and Alajouanine⁴ has been characterised in depth by others.⁵ The present case conforms pathologically to these previous descriptions, but is presented here because of the unusual and unsuspected clinical presentation.

The disease usually presents as a progressive radiculomyelopathy at an average age of 40 years (range 21-75). It is two and a half times more common in men than in women. It is complicated by spinal subarachnoid haemorrhage in about a quarter of cases and sometimes by development of haematomyelia. It may be associated with capillary telangiectasia in the affected dermatomes.

Our patient's symptoms and signs were much more suggestive of a sensory neuropathy than spinal cord disease. In a heavy smoker the appearance of finger clubbing appeared to support a diagnosis of non-metastatic sensory neuropathy, but it is possible that the finger clubbing was related to the arteriovenous shunting in the spinal cord.

The pathological findings were typical of spinal angiomatosis. Most vein walls were markedly tortuous and collagenised or hyalinised. This, together with the apparent arteriatisation of some veins, indicates that blood enters such angiomata at high pressure. However the "river-delta" haemodynamic effect of angiomatous vessels might have reduced perfusion pressure in some parts of the tumour. Aminoff⁶ suggested that the ischaemic necrosis (myelomalacia) of the affected areas of the cord are caused by reduction in the arteriovenous gradient across the angiomata. Thrombosis is a further factor, and one thrombosed vein was noted in the present case. Aminoff⁶ further suggested that the "steal" from the cord. The relative sparing of the lateral column explains the lack of corticospinal tract signs in the developing stages of our patient's illness. In the later stages the associated degeneration of the dorsal, and to a lesser degree, ventral roots might have masked signs of a specific corticospinal tract deficit.

We thank Drs JC Houston and RK Knight for allowing us to report this case.

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References

⁴ Foix C, Alajouanine T. La myélite nécrotique au subaigue. Myélite centrale angiopathique à évolution progressive.


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Cephalic tetanus presenting with Bell’s palsy

Sir: Cephalic tetanus is a clinical variant characterised by paralysis of the cranial nerves.1-3 Weakness of the muscles innervated by the seventh nerve is the most frequently reported symptom. However, cephalic tetanus commencing with facial paralysis, as in the case described here, is unusual.

A 53-year-old man suffered a wound on the right side of his chin, caused by a stick. Two days later, there was inflammation at the site of the injury and weakness of the right orbicularis oris muscle. Later the paralysis increased, with weakness of the ipsilateral orbicularis oculi, but without facial stiffness. On the fifth day, the patient was seen by his general practitioner who diagnosed right-sided Bell’s palsy and prescribed prednisone 15 mg daily. Five days later, the patient had difficulty in swallowing and in opening his mouth, and was referred to our hospital. Examination revealed severe lockjaw and left facial hemispasm, combined with right facial paralysis, predominantly affecting the lower portion (fig). Taste was not affected. The wound was infected. Treatment included debridement of the wound, tracheotomy, penicillin, antitetanic gamma globulin, tetanus toxoid and diazepam. At rest, the electromyogram showed bilateral continuous abnormal activity in the masseters, orbicularis oris and orbicularis oculi muscles; electrical silence was not obtained. The patient began to improve three days after admission, being asymptomatic three months later. A further EMG study after 33 days evolution showed a clear reduction in spontaneous activity and a normal blink reflex.

Facial palsy in the course of cephalic tetanus usually appears when there is already evidence of trismus4 5 7 8 or simultaneously with it.4 6 9-11 Thus the interest in our case lies in the fact that facial paralysis was the only clinical sign for the first eight days of evolution, being diagnosed understandably as Bell’s palsy. Although mistaken diagnoses of facial paralysis at the onset of cephalic tetanus have been mentioned occasionally,2 12 the pattern of evolution described here has only been reported in one case.10 This was a 14-year-old patient who developed left facial paralysis 3 days after suffering an eye injury on the same side; trismus and dysphagia were present the following day. Our EMG findings are consistent with those previously described.13 11 Consequently, when Bell’s palsy and craniofacial injury are found together, the possibility of cephalic tetanus should be considered.

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References


Fig Patient 20 days after onset of symptoms. Note the healed wound on the right side of the chin, right peripheral facial palsy and spasm of the left orbicularis oris and orbicularis oculi muscles.