Subacute autonomic and sensory ganglionopathy: a postmortem case

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LETTERS TO THE EDITOR

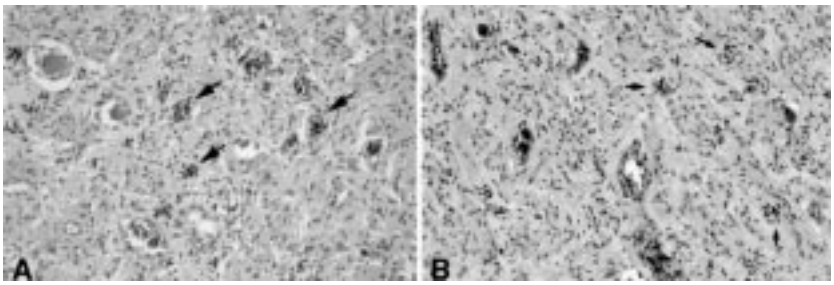
Subacute autonomic and sensory ganglionopathy: a postmortem case

Acute autonomic and sensory neuropathy (AASN) is characterised by severe autonomic dysfunction, sensory deficit, and relatively well or fully preserved motor nerve function.[1–4] The disease sometimes has a chronic course.[2 4] Detailed information is limited because there have been so few reports of the postmortem examination of AASN.[2–4] This case provides histopathological evidence for autonomic and sensory ganglionopathy in AASN.

A 30 year old Japanese man had abdominal pain and a rise in body temperature to 40°C on 1 January 1990. On 23 January, he felt prickling and paraesthesia in all his limbs. On 14 February he could no longer walk due to severe prickling of the entire body. On 2 March, he became bedridden because of orthostatic hypotension. He experienced mild weakness in the all limbs, dysarthria, and dysphagia. On 13 March, he was transferred to our hospital. Physical examination on admission showed anhidrosis, urinary retention, hyposalivation, and paralytic ileus. There was no history of intoxication by drugs or food preceding the illness. His blood pressure was 122/70 mm Hg when supine and 80/46 mm Hg when sitting with no increase in heart rate. His skin was dry. Neurologically, the right pupil was round and 5.5 mm in diameter, whereas the left was oval shaped and 6.0 mm in diameter. The light reflex was sluggish, and the convergence reflex absent bilaterally. Muscle power was mildly weak in his limbs, and there was moderate limb ataxia. Deep tendon reflexes were decreased. Sensation was lost to all modalities over his entire body, including the face. Complete blood cell and serum biochemistry findings were normal. Epstein-Barr virus antibody titres were not raised in the serum. His CSF was normal. Motor conduction velocities (MCVs) and compound motor action potentials were normal in the right median nerve and the right tibial nerve. No sensory nerve action potentials (SNAPs) were evoked in the bilateral sural nerves. The coefficient of variation of the R-R interval on his ECG was reduced (1.9%). In October, dysarthria, dysphagia, and muscle weakness gradually lessened. Despite the improvement in muscle power, severe sensory impairment and paraesthesia of the entire body, as well as orthostatic hypotension persisted. His response to plasmapheresis and high dose corticosteroids was very poor. Two years later, at the age of 33, he died of heart failure.

No tumour was found on general pathological postmortem examination. The dorsal fasciculus at all levels of the spinal cord showed an almost complete loss of myelinated fibres, and the number of dorsal horn cells were decreased. Both the gracilis and cuneatus neurons of the medulla oblongata were slightly decreased. The dorsal roots were atrophic. No detectable changes could be seen in the lateral and ventral fasciculus; nor any clear reduction in the number of neurons in the ventral or lateral horn at Th4 levels. In the cervical (figure A), thoracic, and lumbar dorsal root ganglia, only a few neurons could be seen under low power, with Nageotte nodule formation. In the cervical (figure B) and lumbar sympathetic ganglia, the number of neurons were severely reduced without lymphocyte infiltration. Examination of the dorsal root showed severe loss of myelinated fibres, whereas the ventral root of L3 appeared normal.

This case seemed to be compatible with AASN. Parasympathetic failure was reflected in the loss of salivation, decreased intestinal peristalsis, and bladder atony. Absence of sweating, orthostatic hypotension, and slowness of pupillary dilatation indicated sympathetic failure. Sensory neuropathy was confirmed by an electrophysiological study. The absence of SNAPs in the normal MCVs is best explained by selective involvement of the neurons in lumbar dorsal root ganglia. Neuropathologically, there were no changes in the lumbar ventral roots, but there was severe loss of myelinated fibre in the dorsal roots. Severe degeneration was found in the lumbar dorsal root ganglia neurons at all levels. The slight decrease in the gracilis and cuneatus neurons in the medulla oblongata is thought to be due to anterograde transsynaptic degeneration, as is the slight decrease in the number of dorsal horn cells. As judged by the pathological findings, the site of the sensory nervous system lesion in this case was the lumbar dorsal root ganglia. Similar severe degeneration was found in the sympathetic ganglion neurons, but neurons in the lateral horn remained intact. The site of the autonomic nervous system lesion in our case, therefore, is considered in the sympathetic ganglion. Our case showed the same degree of neuropathologically severe autonomic and sensory ganglinopathy. There have been a few postmortem records of AASN.[2–4] Fagius *et al*[2] showed that the main lesions were in the dorsal root ganglia, dorsal roots, and posterior columns, but they did not examine the autonomic neurons. They speculated that the main lesion responsible for autonomic symptoms was in peripheral nerves. Tohgi *et al*[4] reported severe pathological changes in preganglionic and postganglionic neurons of the autonomic nervous system, posterior columns, and the peripheral nerves, but there they did not describe any pathological findings for the lumbar dorsal root ganglia. Thus they suggested that the main lesion of sensory symptoms was in the peripheral nerves judging from sural nerve biopsy. One report showing both autonomic and sensory ganglion involvement was by Stoll *et al*,[3] who described a 68 year old man with AASN. He died of sudden cardiac arrest four months after onset. In contrast with our case, their patient showed rapid onset (two days) and a very good response to corticosteroids. A postmortem examination detected numerous lymphocytic infiltrates and degeneration in the autonomic and sensory ganglia and, to a lesser extent, in the dorsal roots, dorsal columns, and brainstem. Thus their case is similar to ours in neuropathological lesions. Despite the differences in response to therapy, inflammatory change, and disease course between their patient and ours the pathological findings indicate that the disease, selectively involving both autonomic and sensory systems, is ganglionopathy, not neuropathy.



(A) Section from the lumbar dorsal root ganglia: There are only a few normal lumbar dorsal root ganglia neurons. (B) Section from the sympathetic ganglion: normal nerve cell bodies have disappeared and been replaced by clusters of cells ("nodules of Nageotte" (arrows)). Originally×100.

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