However, the scan has to faced, the MRI cannot be undone, the risk of subsequent events in this particular context looks to have increased and some careful discussion along those lines has to take place. Although, of course, it should be re-focused on the presenting symptom, not the presenting scan. Maybe CT scans are not so bad for patients with headache after all.

Competing interests: None.

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Small neurons may be preferentially affected in ganglionopathy

Haruki Koike, Gen Sobue

In this issue of J Neurol Neurosurg Psychiatry, the paper by Gorson and colleagues describes 23 patients with neuropathic pain that is distributed in a non-length dependent manner (see page 163). In these patients, neuropathy can be distinguished from conventional length dependent small fibre neuropathy because the sensory disturbance involves proximal regions of the limbs, face and trunk, either sparing the acral extremities or with simultaneous involvement of distal and proximal areas. This pattern may suggest that the pathology is localised to small sized neurons in the dorsal root ganglia, although neuropathies where the clinical symptom is that of multiple mononeuropathy in a non-length dependent manner of painful symptoms, such as vasculitic neuropathy, should be considered.

Recent progress in techniques evaluating small fibre involvement such as skin biopsy or quantitative sensory testing has allowed assessment of patients with small fibre dysfunction at an early stage of the disease. The retrospective multicentre study of Gorson and colleagues describes the emerging entity of ganglionopathy, predominantly involving small sized neurons. Because this study excluded patients with established causes of ganglionopathy, these cases may be classified as idiopathic painful ganglionopathy, although prospective evaluation of the natural history should be evaluated. This hypothesis was supported by: (1) the non-length dependent distribution of painful symptoms; (2) relative sparing of large nerve fibre functions assessed by clinical and quantitative sensory examinations; and (3) skin biopsy findings corroborating a non-length dependent pattern. However, as the authors recognise, pathological examination of the dorsal root ganglia is required to confirm this hypothesis.

Pathological evaluation of the dorsal root ganglia in patients with major causes of ganglionopathy by biopsy or autopsy has been reported in patients with Sjogren’s syndrome and paraneoplastic syndrome. The major symptom in these syndromes is sensory ataxia due to impairment of deep kinaesthetic sensation suggestive of large sized neuron involvement. In fact, a previous report of an autopsy of a patient with paraneoplastic neuropathy associated with small cell lung carcinoma showed preferential loss of large diameter sensory neurons in the dorsal root ganglia, a marked decrease in large myelinated fibres in the dorsal root and sural nerve, and a near total loss of myelinated fibres in the fasciculus gracilis. In contrast, ganglionopathy may affect small diameter neurons preferentially, and such ganglionopathy has been reported in an autopsy of a patient with Fabry disease. Recent studies of neuropathies associated with Sjogren’s syndrome and paraneoplastic syndrome suggest variability of clinical symptoms, ranging from patients with pain without sensory ataxia to those with sensory ataxia. Typical cases with painful neuropathy in these syndromes manifest well preserved motor nerve function, small axon loss with relative preservation of large axons and lack of axonal sprouts. These findings indicate the presence of sensory ganglionopathy that affects small ganglion neurons in these cases, although histological examination of the dorsal root ganglia was not performed. Some patients with pain without sensory ataxia eventually developed sensory ataxia due to the impairment of deep kinaesthetic sensation over the long term follow-up. Alternatively, some of the patients with sensory ataxic neuropathy had impairment of superficial sensations with painful dysesthesias. These overlapping symptoms, observed in these two forms of neuropathy, may support the hypothesis that the two neuropathies are part of a spectrum of disorders with a similar pathology.

In conclusion, Gorson and colleagues provide an advanced insight into the current progress in the field of small fibre neuropathy and ganglionopathy.

Competing interests: None.

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