Letter to the Editor

Preservation of Ia afferent function in peripheral neuropathies

We read with great interest the Letter by García and colleagues, commenting on the report by Sukockiene et al. (2020) who showed that “areflexia in multifocal motor neuropathy (MMN) could be caused by predominant involvement of group Ia afferents with relatively preserved cutaneous afferents” (García et al., 2021). We would like to comment on the reverse finding in polyneuropathy: preservation of Ia afferent function in the face of the loss of cutaneous afferent function.

It is not possible to stimulate Ia afferents selectively in routine nerve conduction studies, though preferential stimulation can be achieved using a microelectrode inserted into the motor point (Macefield et al., 1989). Alternatively, the integrity of Ia afferents can be assessed indirectly by measuring spinal reflex function, using the tendon jerk (as in Infante et al., 2018; García et al., 2021) or the H reflex (as in Burke, 2016; Burke and Halmagyi, 2018; see Fig. 1).

García and colleagues mention their report in June 2018 of 5 carefully documented patients with cerebellar ataxia, neuropathy and vestibular areflexia syndrome, “CANVAS” (Infante et al., 2018). Tendon jerks were preserved in the upper and lower limbs in patients lacking sensory action potentials. This report mirrors our April 2018 paper (Burke and Halmagyi, 2018). There we reported, also in 5 patients with CANVAS, the preservation of tendon jerks, with preservation of H reflexes in the upper and lower limbs in the four tested patients, all of whom had absent or severely attenuated sensory nerve action potentials in the upper and lower limbs (Fig. 1).

The patients in these reports are unusual. Clinically the majority of patients with CANVAS appears to have a sensory polyneuropathy with areflexia. The sensory loss is due to a neuronopathy (Szmulewicz et al., 2011, 2014), with loss of neurons in the DRG and thereby loss of sensory nerve action potentials in peripheral nerves and of the ascending spinal projections. In a minority of cases tendon jerks are preserved and may even appear to be brisk (Szmulewicz et al., 2011, 2014, 2015; Cazzato et al., 2016 [one of 4 patients]; Taki et al., 2018 [single case]).

Preservation of tendon jerks occurs in small fibre neuropathies and may occur in primarily axonal neuropathies, such as acute motor axonal neuropathy (AMAN). It also occurs in some neuronopathies, such as Friedreich’s ataxia, but here the retained tendon jerks have been attributed to concurrent corticospinal involvement. Although clinicians expect polyneuropathy with distal sensory loss to be associated with depressed or absent tendon jerks, dissociation of muscle afferent function and cutaneous afferent function is not uncommon. We speculate that this is more likely to occur in ganglionopathies (such as CANVAS) because of the biophysical differences between neurons in the dorsal root ganglia subserving different afferent modalities.

These reports represent a timely reminder to clinicians that relative preservation of group Ia afferent function may occur in

Fig. 1. H reflexes in a patient with CANVAS with absent cutaneous sensory action potentials but preserved tendon jerks. A. Soleus H reflex, appearing below threshold for the M wave with a stimulus of 9.8 mA (upper trace), initially increasing with stimulus intensity but then decreasing as it is occluded by the growing direct motor volley. B. H reflex of abductor pollicis brevis recorded during a steady voluntary contraction. Four superimposed averages, each of 100 sweeps. The H reflex (in the centre of the traces) is much bigger than the M wave at this stimulus intensity (10.1 mA). C. H reflex of flexor carpi radialis, recorded at rest, to increasing stimulus intensity, with the reflex present at 10.5 mA (together with a very small M wave), initially increasing with stimulus intensity but then decreasing as it is occluded by the growing direct motor volley. From Burke and Halmagyi (2018) with permission.

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polyneuropathy, and that this can be documented by measuring tendon jerks (Infante et al., 2018; García et al., 2021) or H reflexes (Burke, 2016; Burke and Halmagyi, 2018).

Declaration of Competing Interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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