Recurrent common peroneal palsy in association with the Ehlers-Danlos syndrome

A case report

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We report a patient with type III Ehlers-Danlos syndrome, presenting with recurrent footdrop, who had electrophysiologic evidence of a conduction block of the common peroneal nerve followed by complete resolution. We feel that this case supports the possible link between Ehlers-Danlos syndrome and multiple pressure sensitive neuropathy (tomaculous neuropathy), alluded to by Schady and Ochoa (1984).

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A 25-year-old female, known to have type III Ehlers-Danlos syndrome, had previously suffered problems with subluxation and dislocation of several major joints, including the shoulders, hips, and patellae. In April 1989, she presented with a 10-week history of fluctuating sensory disturbances in her left thigh, left lower leg, and right calf. The symptoms on the left side subsided after 2 weeks, but those on the right side progressed, with paresthesia spreading to the dorsum of the foot and weakness of dorsiflexion. There were no preceding symptoms and no history of a recent dislocation or trauma.

On examination, she had altered sensation on the anterolateral aspect of her right lower leg and the dorsum of the foot. She had reduced proprioception of her great toe and grade 1 power in the ankle dorsiflexors and evertors, with mild wasting of her anterior tibial muscles. Nerve conduction studies revealed a marked block of the right common peroneal nerve with absence of the f-wave. Without treatment, her symptoms improved rapidly; however, in late May, the footdrop recurred with the associated sensory changes. On questioning, she also admitted to having some paresthesia in the ulnar borders of both hands.

A complete blood count, ESR, urea, and electrolytes were all normal. A lumbar puncture was performed, with normal CSF pressure, proteins, glucose, and microscopic findings. Repeat studies revealed mild slowing of ulnar nerve conduction across the elbow, slowing of conduction of the left common peroneal nerve, but a severe block of the right common peroneal nerve at the knee, with good distal conduction, suggesting that recovery of function was likely to occur. She was fitted with a footdrop splint and discharged.

At review in August, she had regained full power of the left ankle and four-fifth power of the dorsiflexors of the right ankle. She had discarded her splint. However, in November, she had once again developed a right-sided footdrop with grade 2–3 power of the dorsiflexors and evertors. It was debated at the time whether decompression of the common peroneal nerve was appropriate; however, when reviewed in December, there was again improvement in her symptoms. By January 1990, the toe extensors and tibialis anterior had recovered to grade 4, with the peroneae at grade 3. The sensory impairment of the lateral aspect of the leg and the dorsum of the foot remained. When last reviewed in June 1990, she had complete recovery of power, normal sensation, and normal nerve conduction studies.

Discussion

Ehlers-Danlos syndrome is a hereditary generalized disorder of connective tissue manifested by widespread hyperelasticity of the skin and joint laxity. It has been classified into many types depending upon the particular pattern of its manifestation. In type III Ehlers-Danlos syndrome, joint laxity is the predominant feature, and dislocation and subluxation are common.

Neurologic problems associated with Ehlers-Danlos syndrome have been described: most commonly,

central nervous system defects associated with intracranial vascular events as a result of the abnormal collagen in the vessel walls (Herrero 1980). Peripheral neurologic problems are less commonly reported, but again may be secondary to vascular abnormalities (Bowers et al. 1976). Both brachial and sacral plexus dysfunction have been described, but the causative mechanism is less clear (Degraff 1973, Kayed and Kasse 1979, Papapetropolous et al. 1981, Pretorius and Butler 1983). Schady and Ochoa (1984) reported a soldier with Ehlers-Danlos syndrome who developed clumsiness of his arm. A nerve biopsy showed the presence of tomaculous neuropathy, a histologic finding often associated with hereditary multiple pressure sensitive neuropathy. Although biopsy information is not available for our patient, the involvement of several nerves, as shown by nerve conduction studies, would suggest that her symptoms may have been the result of this type of neuropathy.

The pattern of recurrent but completely resolving episodes of peripheral neuropathy would be typical of someone with hereditary multiple pressure sensitive neuropathy. However, our patient has no family history of neuropathy (Behese et al. 1972). Direct mechanical trauma could be postulated as a cause of her footdrop, but there is no history of dislocation or subluxation of her right knee, nor is there any suggestion of local trauma. Another possibility is that the lesion is due to a repetitive traction injury as a consequence of the hypermobility of the joints.

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