LETTERS TO
THE EDITOR

Recurrent spontaneous accessory neuropathy

Isolated spinal accessory nerve palsy of obscure or spontaneous origin is an uncommon but well recognised entity.1 We describe the first reported instance of recurrent spontaneous accessory palsy.

At the age of 28, the patient developed an aching discomfort in the right shoulder and neck. This resolved after a few days but after several weeks he became aware of weakness of shoulder elevation. Examination revealed weakness referred to the right trapezius muscle. Nerve conduction studies of the accessory nerve, recorded in the trapezius, revealed latencies of 4.3 ms on the right and 2.9 ms on the left (normal < 3.2 ms). Electromyography showed large motor unit potentials and reduced recruitment in the right trapezius only. After four to five months the condition improved and the size and strength of the right shoulder returned to normal.

He remained well and continued to work as a policeman until he was 31, when he again developed an ache in the right shoulder after exercising with five-pound dumb-bells. The ache persisted for two to three weeks but was never severe and did not interfere with his sleep. Naproxen provided some relief. About three weeks after the onset of pain, the patient began to notice wasting of his right shoulder and weakness of shoulder elevation. Two weeks later the pain had entirely resolved but he was now aware of the right shoulder hanging lower than the left. He had no sensation of loss or pain other than that in the shoulder. The other limbs and spinal function remained normal.

Examination revealed a fit man with marked wasting of the right trapezius muscle and moderate weakness of shoulder elevation. There was right scapular winging of the trapezius type, which was present with the arm at rest and accentuated by lateral elevation of the arm. The right sternomastoid muscle was normal. No wasting or weakness was observed in the other right shoulder girdle muscles and the remaining cranial nerves and other limbs were normal. General examination was normal and there was no sign of injury or abnormality on the lateral side of the neck.

Cervical radiographs and myelography of the foramen magnum were normal. CT scan of the neck and skull base revealed no abnormality. The cerebrospinal fluid (CSF) was clear and colourless, with four lymphocytes and a protein of 0.34 g/l. Serum glucose, full blood count, sedimentation rate, C-reactive protein, antinuclear factor, and routine biochemistry were normal.

Nerve conduction studies of the accessory nerve, recording trapezius, revealed a considerable decreased amplitude of the compound muscle action potential on the right, but the distal latencies were similar and normal on both sides (right = 3.2 ms, left = 3.1 ms). Electromyography revealed fibrillation potentials and large motor unit potentials with reduced recruitment in the right trapezius, and normal findings in the serratus anterior, infraspinatus, deltoid, biceps, brachioradialis and extensor digitorum communis.

Our patient experienced two similar episodes, separated by eight years, of shoulder ache followed by right trapezius weakness and wasting. On both occasions clinical and electromyographic examination found no abnormality except in the right trapezius, implying the presence of a lesion affecting the accessory nerve distal to the innervation of the sternomastoid. He appeared to complete recovery after the first episode, only to suffer a recurrence eight years later. Investigation has not shown an underlying cause and we therefore believe he is the first reported instance of a recurrent recurrent accessory neuropathy. Although one of the second episode coincided with light exercise with weights, these were small (5 pounds) and seem unlikely to have produced a stretch injury of the accessory nerve.

Due to its complicated course, the accessory nerve may be damaged by a wide range of processes affecting the cervical spinal cord, brachial plexus, cervical spinal canal, or at the foramen and skull base carotid sheath, or posterior triangle of the neck. The nerve is particularly vulnerable as it crosses the posterior triangle, where injury occurs most often following radical neck dissection for biopsy, resulting in weakness and wasting of the trapezius.

Spillane described three patients in 1949 who had clinical evidence of isolated accessory neuropathies, the cause of which was obscure.2 They made a complete recovery after no evidence of improvement of the weakness and atrophy of the sternomastoid and trapezius. Four similar cases were studied clinically and electrophysiologically by Eisen and Bertrand in 1972.3 One of their patients recovered completely in 22 months, but two did not improve and one was lost to follow up. There have been occasional case reports of isolated “spinal accessory palsy” in the literature since then, but our case is the first in which there was recovery and spontaneous recurrence in the same nerve. Typically there is abrupt onset of pain in the lateral neck or shoulder region, which is followed by wasting of the trapezius and sternomastoid, which then subsides over several weeks and is followed by wasting and weakness of the trapezius and sternomastoid, or both. Recovery has occurred in a minority of the reported cases and takes several months to one or two years.

Analyses may be drawn between isolated spontaneous accessory palsy and two other more common entities, idiopathic facial (Bell’s) palsy and peripheral amyotrophy (Parson Turner syndrome). Common to all three conditions is pain at the onset, followed by muscle weakness and atrophy, which subsequently recovers to a variable extent. In the majority of presentations, Bell’s palsy and neurogenic amyotrophy are quite distinct entities, but occasionally cases incorporate elements of both. Neuromuscular junction involvement is now being recognised as an increasingly diverse disorder,6 although accessory or other cranial neuropathies are rarely described as part of this syndrome. In our patient, involvement of a single cranial nerve and relatively mild pain seems to us more akin to Bell’s palsy than to neurogenic amyotrophy.

Pesticide toxicity and motor neuron disease

Pyrethrins are neurotoxins that kill insects by paralysis—“knock down”. This is thought to be secondary to an effect on sodium channels in motor neurons. Mammals are spared from this effect as, unlike insects, they can metabolise pyrethrins in the hepatic microsomal system to non toxic compounds.7

We previously reported a case simulating motor neuron disease (MND) closely associated with over-exposure to permethrin and chlorothane based insecticide.8,9 The chlorothane component is metabolised to various compounds (including oxyclozadane and epoxychlorodane) via the cytochrome P450 P450 enzyme system. However, it is not clear how permethrin is metabolised by different routes (fig). Following esterase cleavage, two products are formed which are further metabolised then eliminated. In mammals,10 metabolite A is excreted in the urine as a glucuronide conjugate. Metabolite B undergoes oxidation and hydroxylation to yield mainly C and D, with other compounds. Conjugates of minor metabolites with glycine,11 glutamic acid12 and taurine13 have been noted. However, the elimination of metabolites C and D in the rat is by sulphate conjugation14 which is the major metabolic pathway for permethrin.

This is of interest since we recently reported that MND patients have a defect in their ability to convert cysteine into inorganic sulphate and also show a poor capacity to form the sulphone conjugate of paracetamol. In the light of these observations, the 4’-Hydroxy-3-phenoxyl benzyl alcohol (metabolite C) and/or 4’-Hydroxy-3-phenoxyl benzoic acid (metabolite D) may be responsible for the neurotoxicity that resulted in our patient suffering from his MND-type illness. Some environmental chemicals generally

Values are expressed as the mean (1 SEM) for data obtained using seven animals.

*p < 0.01 compared to vehicle treatment, Dunnett’s multiple comparison test.