Letters

Familial bilateral carpal tunnel syndrome

Sir,—Familial carpal tunnel syndrome, unassociated with systemic disease has been rarely reported. To the best of our knowledge, only 13 families have been reported in the world literature. In most of the reported cases, precise family pedigree was not shown and the pattern of inheritance is unclear. We describe here a new family with carpal tunnel syndrome. No common aetiologic feature was found in the patients and the family pedigree suggests autosomal dominant pattern of inheritance (fig).

A 45 year-old-woman (case 1) began to have numbness and tingling of each hand, involving mainly the thumbs, second and third fingers 15 years ago. The burning sensation gradually spread from both hands to shoulders and this symptom often awakened her at night. Five years ago, decompression of the right median nerve was performed by section of the transverse carpal ligament. Left median decompression was performed three years ago. Subjectively, there was considerable improvement, as the burning sensation stopped, but the tingling sensation of the fingertips evoked by light touch persisted. Physical examination, performed when aged 44 years, showed bilateral weakness of abductor pollicis brevis, opponens pollicis and flexor pollicis brevis, and abduction of the left thumb was limited to 70°. Impairment of sensation to light touch, pinprick and disturbance of two-point discrimination was noted over the median distribution in both hands. A sister of case 1, aged 40 years (case 2), started to have a tingling and burning pain of left hand, involving the thumb, second and third fingers 14 years ago. Later, her right hand was similarly affected. Physical examination showed slight atrophy of each thenar eminence. Slight loss of pinprick sensation and disturbance of two-point discrimination was noted in the thumb, second and third fingers of the left hand and the third finger of the right hand. Seven family members with carpal tunnel syndrome were identified by history taken from these two patients.

In both case 1 and case 2, prolonged distal motor latency was obtained in stimulation of the median nerve at the wrist. The sensory fibre examination of the median nerves showed the abnormal prolongation of distal latency in both sides of case 1. No action potential was obtained in the right hand of case 2. Motor conduction velocity in the median nerve between the wrist and elbow was found to be normal in both cases. Complete blood count, urinalysis, sedimentation rate and biochemistry including total protein, protein fraction, serum iron, rheumatoid factor, ANA, thyroxine, growth hormone, immunoglobulins were normal in both cases. Using a standard microcytotoxicity technique, histocompatibility antigen typing was done in these two cases and in four offspring (fig). Specific AW13 and B12 antigens were noted in each of two patients.

In a review of the literature about primary familial carpal tunnel syndrome, four male and 21 female cases had been reported. However, the pedigree that Gray reported there was an equal number of patients of each sex and this suggested the autosomal dominant pattern of inheritance with a high degree of penetrance.1 In our cases, the gene was transmitted through a male or female who had symptoms of the syndrome, and the autosomal dominant pattern of inheritance is apparent. Idiopathic cases of carpal tunnel syndrome have features of sporadic occurrence, middle-life onset and female predominance.2 On the other hand, familial carpal tunnel syndrome has a variable age of onset from first decade to menopause.3,4 Gray found no association of familial carpal tunnel syndrome with an histocompatibility antigen,1 but Shinohara examined HL-A typing in a Japanese family of familial carpal tunnel syndrome and found that A2 was present in four female patients.5 Our cases, the second family to be reported in Japan, showed no HL-A-A2 antigens but AW33 and B12 antigens were common. The cause of familial carpal tunnel syndrome is unknown and no genetic markers were previously known for this disease. It is hoped that with further study of HL-A typing as a marker, new approaches may be initiated in the study of this disease.

References

2 Phalen GS, Kendrick JI. Compression neuropathy of the median nerve in the carpal tunnel. JAMA 1957; 164:524–30.