Matters arising

our review of apraxia following deep cerebral lesion and we acknowledge their priority in the investigation of this subject. We are not certain, however, that the evidence they have presented in support of the deep nuclei involvement in limb apraxia is compelling. Most of their patients scored 15 out of 20 on the imitation test, which is just two points below the cut-off score reported by De Renzi, *et al*¹ (Agostoni *et al* did not have a control group). One wonders whether such a marginal deficit on a single test deserves the label of apraxia, all the more so as of the seven patients they report, three had damage to the right brain, an amazingly high proportion for a symptom that is classically associated with left hemisphere disease.

Reference

- 1 De Renzi E, Pieczuro A, Vignolo LA. Ideational apraxia: a quantitative study. *Neuropsychologia*, 1968;6:41-52.

Peripheral neuropathy associated with sicca syndrome

Sir: We wish to report an additional case of a woman suffering from the sicca syndrome who presented with features of a chronic sensory neuropathy, complementing the cases recently described by Kennett and Harding.¹

She presented in 1972, at the age of 64 years, with a one year history of numbness which began in the right index fingertip and gradually spread to involve all fingers of both hands. Examination disclosed impaired sensation to all modalities apart from vibration sense in all fingertips. Except for a normal left knee jerk, generalised areflexia was present. The following year she developed painful dry eyes and mouth and a sore tongue. Her symptoms persisted and in 1976 no sensory nerve action potentials were detectable when recording from the median and ulnar nerves. Median, ulnar and peroneal motor nerve conduction velocities were normal. Cerebrospinal fluid protein level was 0.16 g/l (normal 0.15-0.45) and glucose 3.5 mmol/l (normal 2.2-4.4). A muscle biopsy examination was normal but sural nerve biopsy showed mild endoneurial fibrosis and a 30% reduction in the number of large myelinated nerve fibres (fig). No active inflammation was seen in the nerve or vessels.

Sicca syndrome was diagnosed and despite one prolonged course of corticosteroid treatment her condition has been indolent but slowly and inexorably progressive. By 1986 she had increasing difficulty chewing, swallowing and speaking as a result of a dry mouth. Pain and touch appreciation was reduced in the distribution of the right trigeminal nerve and the right corneal response was reduced. Pseudo-athetosis of the outstretched hands was present and proprioception was lost below

the elbows and impaired in the shoulders bilaterally. Light touch, pain and temperature appreciation were impaired below the right elbow and left mid forearm. Vibration sense was absent below the shoulders bilaterally. The two point discrimination threshold was greater than 8 cm on the index fingers. Generalised areflexia persisted except for a brisk left knee jerk. The remainder of the neurological examination was normal including a remarkable sparing of lower limb sensation. A weakly positive antinuclear antibody (30 U/l) with mixed staining anti DNA binding 4% was present. Serum immunoglobulins, complement, cryoglobulins and protein immunoelectrophoresis were normal.

This patient, now aged 79 years, suffers from a chronic peripheral sensory neuropathy with predominant involvement of large myelinated fibres as reflected by the considerable impairment of proprioception in the upper limbs. Although the sensory symptoms commenced asymmetrically the subsequent sensory deficit in the upper limbs has become symmetrical with time.

Sicca syndrome constitutes keratoconjunctivitis sicca and xerostomia. Sjogren's syndrome refers to sicca syndrome with an associated connective tissue disease. Peripheral neuropathy has been generally considered to be a rare feature in Sjogren's syndrome.² Nerve biopsies performed in such patients have occasionally shown evidence of medium to small vessel vasculitis.³

The characteristic neurological syndrome associated with sicca syndrome, comprising areflexia and asymmetrical sensory loss in the limbs, is often associated with tonic pupils and trigeminal anaesthesia.¹ Peripheral nerve histology has not been reported previously in these patients. It is of note that the sural nerve biopsy appearances of our case showed no evidence of small to medium vessel vasculitis.

This case report reinforces the conclusion of Kennett and Harding¹ that the diagnosis of sicca syndrome should be considered in patients with chronic sensory axonal neuropathy, particularly if proprioceptive loss is prominent and there are pupillary abnormalities and/or trigeminal involvement.

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References

- 1 Kennett RP, Harding AE. Peripheral neuropathy associated with the sicca syndrome. *J Neurol Neurosurg Psychiatry* 1986;49:90-2.

Fig Kulitschitzky-Pal myelin stain showing reduced numbers of myelinated fibres ($\times 150$).

- International Society for Adolescent Psychiatry.** The 2nd International Congress will be held in Geneva, 10-13 July 1988. Information may be obtained from: The Secretary, 2nd International Congress for Adolescent Psychiatry, P.O. Box CH-1211 Geneva 8, Switzerland.