Matters arising

Our review of apraxia following deep cerebrovascular 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 (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


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.

She presented in 1972, at the age of 64 years, with a one year history of numbness which began in the right index finger 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. Pseudoathetosis 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 asymptomatically 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.1 Nerve biopsies performed in such patients have occasionally shown evidence of medium to small vessel vasculitis.

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 Harding 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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Fig Kultschitzky-Pal myelin stain showing reduced numbers of myelinated fibres (× 150).
On the mystery of multiple sclerosis

Sir: In his excellent article, Professor McDonald, with admirable discretion, really leaves untouched the mystery of the origin of multiple sclerosis after citing his title as a quotation of Gowers. However, in but a few pages he summarises nicely the evidence which leads him to conclude as to the nature of multiple sclerosis. “That it is a disease produced by an environmental agent in genetically susceptible individuals in whom there is an abnormality of the immune mechanism.” He then points out that, in fact, each phrase of this definition remains to be elucidated. But he does provide a description of the essential pathophysiology of the disorder with which few could cavil.

There are, though, several points in the historical evolution of the concepts elaborated by Professor McDonald that bear modification. Dawson, while among the earliest to related the topography of plaques to the vascular tree, did not in fact implicate a relationship with venous territories. He stated “that in the course of this study several small areas have been followed up, serially, throughout their whole extent, and I have come to the conviction that the changes appear, but do not coincide with the area of distribution of the arteries” (p. 619).

The dominant description of the vascular supply of the central nervous system at that time was that of Kadyi, and it was not until the work of Herren and Alexander in 1939, that the venous drainage of the spinal cord was well defined. With this, it became possible to show that the topography of multiple sclerosis plaques in the cord was indeed that of the venous drainage, and that the earliest lesions seemed to be perivenular (p. 163). With further assessment it became clear that cerebral plaques also were not only perivascular but also perivenular (p. 151).

The great periventricular plaques in the walls of the lateral ventricles so commonly seen at necropsy and also in MRI are in fact the confluence of perivenular plaques, with or without perivenous extensions into the central white matter (Dawson’s “fingers”). This was demonstrated by reconstruction of such plaques, as illustrated in fig. 13 (p. 80). Periphlebitis retinae (“venous sheathing”) is a similar process on the retina, with lymphocytic infiltrations about the vessel (figs 17, 18; p. 154). Of course one can not speak of “plaques” without demyelination or astrocytosis, but the essential pathology seems quite similar. Tine Engell, found periphlebitis retinae in 15% of 135 patients hospitalised for multiple sclerosis versus 5% of 168 multiple sclerosis patients examined at a rehabilitation centre. Of 37 patients evaluated during exacerbation or rapid progression, periphlebitis retinae was present in 16 (43%). I would suggest then that one might reconsider the last sentence of my work on cerebral plaques: “From the point of view of pathological anatomy, multiple sclerosis is a condition of “periphlebitis cerebrospinalis et retinalis”, (p. 159).

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3 Kadyi H. Über die Blutgefäße des menschlichen Rückenmarkes. Lemberg 1889.

Matters arising

Thoracic cord compression from metastatic prostate carcinoma with Lhermitte’s “sign”

Sir: I was interested to read the report by Baldwin and Chadwick in which a patient with a cavernous hemangioma producing complete spinal block at the fifth thoracic level experienced Lhermitte’s “sign”. I thought the authors might be interested in a report of similar symptoms in another patient with thoracic spinal block, although flexion of the neck in this case produced a sensory disturbance radiating into the anterior thighs. The description of the patient’s symptomatology is rendered vividly as the author (a neurosurgeon) described the symptoms experienced at the time he developed spinal cord compression from prostate carcinoma. Interestingly, the level of spinal block was also at the fifth thoracic level. His symptoms, as with their patient, disappeared after surgical decompression.

The mechanism of the particular sensory disturbance experienced upon neck flexion by both of these patients remains speculative. I concur with Baldwin and Chadwick that, at least in some patients, Lhermitte’s “sign” should direct investigation to possible pathology in the thoracic as well as cervical spine region.

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References

Notices

International Society for the Study of Brain Edema. The Triennial Meeting will be held 7–10 October 1987 in Baltimore, USA. Information may be obtained from: Program Coordinator, Office of Continuing Education, The Johns Hopkins Medical Institutions, Turner 22, 720 Rutland Avenue, Baltimore, Maryland 21205, USA.

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 50, CH-1211 Geneva 8, Switzerland.
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