CASE REPORTS

Sarcoidosis and chronic sensory neuropathy

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Summary
A patient with long-standing sensory peripheral neuropathy is described who finally presented with acute pulmonary sarcoidosis after a period of 18 years. Purely sensory neuropathy presumably due to sarcoidosis of this duration has not been reported before.

Introduction
Peripheral nerve involvement in sarcoidosis is uncommon, occurring in less than 2% of cases (Delaney, 1977). The neuropathy is usually entirely motor or mixed motor and sensory in type, characteristically it occurs in patients whose sarcoidosis is of recent onset (Silverstein, Feuer and Siltzbach, 1965) and the course of such disease is often transient. Pure sensory peripheral neuropathy in the absence of cranial nerve involvement is rare. The patient now described had long-standing sensory symptoms but finally presented with acute pulmonary disease after a period of 18 years.

Case report
A 58-year-old secretary, presented with a 2-week history of dyspnoea. She admitted to mild stiffness of both hands for several years and a periodic blistering rash on her hands and arms unrelated to exposure to sunlight or trauma. She also complained of numbness and paraesthesiae affecting her upper limbs. These had started 18 years previously as numbness of the middle 2 fingers of the left hand, progressing to involve the whole of the right and left hands over a period of 6 months, of sufficient severity to impair her writing and make knitting and typing almost impossible. The symptoms persisted almost unchanged for the next 6 years and then extended to involve both arms. She denied symptoms in her lower limbs or symptoms referable to the central nervous system. She had been treated for depression 6 years previously and on admission was taking phenelzine and chlorpromazine. Her alcohol consumption was low.

On examination she was overweight with late inspiratory crackles at both lung bases. She had no lymphadenopathy and examination of the heart and abdomen was normal. Examination of the nervous system revealed 'glove and stocking' loss of light touch sensation to wrists and mid-calf with peripheral blunting of pin-prick sensation. Vibration sense was absent to the elbows and both hips. Temperature sensation was normal. There was no other neurological abnormality.

Results of investigations were as follows: Hb 12.0 g/dl; no macrocytosis; WBC 7.9 x 10^9/l; ESR 20 mm in one hour; urea, electrolytes, blood sugar and liver function tests were all normal; Wasserman reaction negative; urinary porphyrins and porphobilinogen absent, blood protoporphyrin absent; serum B12 and red cell folate levels normal; serum and urinary calcium levels normal; serum immunoglobulins normal; Mantoux test 1/1000 negative. Examination of the CSF was normal, as were radiographs of the cervical spine. Chest radiograph showed irregular nodular shadowing throughout both lung fields. Pulmonary function tests showed a mild restrictive ventilatory defect. The results of nerve conduction and EMG investigations were as follows. Sensory studies: right ulnar sensory action potential (SAP) (fifth finger to wrist) absent; right median SAP (second finger to wrist), latency to peak 2.4 m/sec, amplitude 2.1 µV; right radial SAP, latency to peak 2.5 m/sec, amplitude 2.9 µV; right sural SAP absent. Motor studies: right ulnar motor conduction velocity (MCV), distance 27 cm, 55.1 m/sec. Stimulation at wrist, distal motor latency (DML) 2.2 m/sec, muscle action potential (MAP) 6.9 mV. Stimulation at elbow, proximal motor latency (PML) 7.1 m/sec, MAP 6.2 mV;
right median MCV (concentric needle electrode in abductor pollicis brevis), distance 20 cm, 52.6 m/sec. Stimulation at wrist, DML 2.9 m/sec, stimulation at elbow, PML 6.7 m/sec; right lateral popliteal (surface electrode over extensor digitorum brevis), distance 27.5 cm, MCV 45.8 m/sec. Stimulation at ankle, DML 4.2 m/sec, MAP 3.4 mV. Stimulation at knee, PML 10.2 m/sec, MAP 4.2 mV. Mixed nerve studies: right ulnar (wrist to elbow) nerve action potential, distance 26.5 cm, latency to peak 5.0 m/sec, amplitude 9.3 µV, velocity 53 m/sec. EMG studies: concentric needle electrode (CNE) in right abductor pollicis brevis, full pattern with some units to 6 mV. CNE in right abductor digiti minimi, no spontaneous activity, full pattern with units up to 5.0 mV. These results are consistent with a severe sensory neuropathy. Histological examination of a drill lung biopsy confirmed the presence of multiple, well formed, non-caseating granulomata with giant cells, compatible with sarcoidosis. Ziehl-Nielsen and Grocott stains were negative.

She was started on prednisolone 30 mg daily with rapid symptomatic improvement. At routine follow-up after 3 months she had no dyspnoea and her chest radiograph was clear. Her numbness and paraesthesiae had resolved and she was able to write and type without impairment. Nerve conduction studies at this stage showed no change. Cautious steroid reduction has resulted in slight recurrence of numbness in both hands. Her chest radiograph remains clear. Sensory action potentials are still absent after 10 months on systemic steroid therapy.

Discussion

This patient's chronic sensory neuropathy was only recognized when she presented with the typical pulmonary involvement of sarcoidosis, confirmed by lung biopsy. The rapid improvement in her symptoms following steroid therapy suggests sarcoidosis as the probable cause of her neuropathy and this dramatic, although partial, response to corticosteroids is typical (Matthews, 1965). Wells (1967) described 2 cases with chronic paraesthesiae, one of 3 years' duration, and drew attention to the phenomenon of a generalized systemic illness which flares up after years of local, seemingly benign, disease. However, such cases are rare and they have not otherwise been reported. The 18-year history of sensory neuropathy symptoms in the present patient is apparently unique.

The nature of peripheral nerve involvement in sarcoidosis is still unclear. It has been suggested that symptoms are of 'toxic' origin (Maycock et al., 1963) and the slowing of nerve conduction in some cases is of a severity usually ascribed to segmental demyelination, more suggestive of such a 'toxic' reaction. However, where sarcoid involves the cranial nerves or occurs elsewhere in the nervous system, granulomata are usually found (Jones Williams, 1967) and granulomatous involvement of peripheral nerves remains likely as the cause of peripheral neuropathy in sarcoidosis.

References


