Letters


Accepted 29 August 1987.

Motor entrapment neuropathies in the elderly

Sir: To our knowledge the frequency of entrapment neuropathies in the general population is unknown. From 1 June 1984 to 31 July 1985 we performed a survey on the neurological condition in the elderly in the Republic of San Marino, which is the smallest independent State in the world, located near the Adriatic Coast, within Italy. All people aged 67, 72, 77, 82 and 87 living in the Republic were invited to undergo a standardised neurological examination. Three hundred and ninety eight agreed to be examined out of a total sample of 498 but full examination was only possible in 396. People were not specifically asked about sensation disturbances of entrapment neuropathies. However, muscle wasting and loss of muscle strength were recorded during examination. Whenever examination neuropathy was suspected patients were further asked about their symptoms and a specific sensation examination was done. Diagnosis of entrapment neuropathy was based on neurological examination showing typical motor and sensory deficits. Whenever possible nerve conduction velocity (NCV) study was performed.

Among 396 people we found 7 (2%) with an entrapment neuropathy. Details of the patients are shown in the table.

In two patients NCV study was not done, but in these two the typical signs and symptoms enabled us to establish a diagnosis of entrapment neuropathy. The design of our study was such that only subjects with motor signs of entrapment neuropathy (with or without sensory disturbances) were considered abnormal. Therefore our figure may be considered indicative of the frequency of motor entrapment neuropathies in a general population older than 65 years.

Five patients had not previously sought neurological consultation. Their symptoms had been generically attributed to “arthritis”. This shows that entrapment neuropathies although not very frequent in the elderly, may be easily overlooked.

**Table** Features of motor entrapment neuropathies found among 396 people aged over 65 years

<table>
<thead>
<tr>
<th>Age (yr)</th>
<th>Sex</th>
<th>Nerve involved</th>
<th>Site of entrapment</th>
<th>Sensation deficit</th>
<th>NCV</th>
<th>Underlying factor</th>
</tr>
</thead>
<tbody>
<tr>
<td>67</td>
<td>♂</td>
<td>Right ulnar</td>
<td>Elbow</td>
<td>Yes</td>
<td>No</td>
<td>Elbow fracture aged 45 years</td>
</tr>
<tr>
<td>67</td>
<td>♂</td>
<td>Right ulnar</td>
<td>Wrist</td>
<td>Doubt</td>
<td>Yes</td>
<td>Articular blockade</td>
</tr>
<tr>
<td>72</td>
<td>♂</td>
<td>Left median</td>
<td>Wrist</td>
<td>Yes</td>
<td>No</td>
<td>Mild arthrosis</td>
</tr>
<tr>
<td>77</td>
<td>♂</td>
<td>Peroneal</td>
<td>Capitulum fibulae</td>
<td>Yes</td>
<td>Yes</td>
<td>Moderate arthrosis</td>
</tr>
<tr>
<td>77</td>
<td>♂</td>
<td>Bilateral median</td>
<td>Wrist</td>
<td>Yes</td>
<td>Yes</td>
<td>Mild arthrosis</td>
</tr>
<tr>
<td>77</td>
<td>♂</td>
<td>Right median</td>
<td>Wrist</td>
<td>Yes</td>
<td>Yes</td>
<td>Moderate arthrosis</td>
</tr>
<tr>
<td>77</td>
<td>♂</td>
<td>Right ulnar</td>
<td>Elbow</td>
<td>Yes</td>
<td>Yes</td>
<td>Moderate arthrosis</td>
</tr>
</tbody>
</table>

NCV, nerve conduction velocity.

Accepted 22 September 1987.

Relapsing dermatomyositis associated with sarcoidosis

Sir: Two cases of dermatomyositis in association with sarcoidosis have been described, both in Japanese patients. We report a further case in an Anglo-Japanese man, his illness running a relapsing course over a period of 6 years.

In October 1979 a 56 year old Anglo-Japanese accountant presented with a 6 month history of pain, stiffness and parasthesiae in both hands, the symptoms tending to be worse at night but also aggravated by heavy work. He also had swelling of the knuckles of both hands, and an irritable skin rash. He had lost 8 kg in weight. There was a past history of duodenal ulcer.

On examination he was a thin man (weight 57.5 kg). He had an erythematous scaling rash of his lower legs, arms, forehead and back, periorbital oedema, and bilateral axillary lymphadenopathy. Examination of his chest, heart and abdomen was normal. There was no muscle weakness or wasting, though he did have some muscle tenderness. Reflexes were normal and sensation was intact. Full blood count, bone marrow aspirate and trephine biopsy, barium enema, barium meal and intravenous pyelogram were normal. RA latex and ANA were negative. Erythrocyte sedimentation rate was 17 mm in 1 hour. Creatine kinase was elevated at 384 IU/l (normal less than 240 IU/l). A chest radiograph showed bilateral apical pleural thickening with a little localised calcification suggestive of previous pulmonary tuberculosis. There was also some interstitial shadowing at both bases, though this resolved after 1 month. Pulmonary function testing indicated a minor restrictive impairment and reduced carbon monoxide diffusion. Lymph node biopsy showed benign reactive changes with follicular hyperplasia, sinus histiocytes and numerous aggregations of histiocytes in the interfollicular cortex and medulla.

A diagnosis of dermatomyositis was made. His symptoms fluctuated over the next 5 years. The CK was persistently mildly elevated until the most recent relapse. From 1980-1982 he had only mild rash and muscle stiffness. In early 1982 he was given a 3 week course of steroids with resolution of his symptoms. Six months later he had a recurrence of skin rash and developed parasthesiae which resolved without treatment in 2 months.

In November 1983 he complained of a 2 month history of general malaise and muscle tenderness and was found to have mild proximal weakness (MRC grade 4–5). This resolved without treatment over the next 3
months. In February 1984 he began to lose weight and developed rectal bleeding. He was found to have a tumour of the distal transverse colon, for which he underwent extended transverse colectomy. Histology showed a moderately well-differentiated adenocarcinoma (Duke's B). He regained his weight following surgery and remained well over the next year, when the rash recurred. It settled after 1 month without treatment, but he again lost weight and noticed increasing weakness and muscular pains. He also complained for the first time of difficulty in swallowing with a tendency to choke on fluids. Examination showed a cachectic man with generalised muscle wasting. There was a global weakness of the arms (MRC grade 3–4), worse on the right, and mild proximal weakness of the right leg. Tendon reflexes were brisk and symmetrical with bilateral flexor plantar responses. Sensation was intact, and general examination was normal.

Full blood count, urea and electrolytes, blood glucose, serum calcium, alkaline phosphatase, albumin and serum thyroxine were normal. Erythrocyte sedimentation rate was 40 mm in 1 hour. Creatine kinase was elevated at 2464 IU/l, with a CK-MB of 204 IU/l (normal less than 16 IU/l). A chest radiograph was unchanged from that of 1979. Mantoux was positive at 1 in 1000. Kveim test was positive. Serum angiotensin converting enzyme was normal. Barium meal showed an aperistaltic oesophagus and hypotonic stomach with gross gastrointestinal reflux. Barium enema, abdominal ultrasound and isotope bone scan showed no evidence of recurrent neoplasm.

Nerve conduction studies were consistent with a mild sensorimotor peripheral neuropathy. Concentric needle electromyography showed prominent fibrillation potentials and polyphasic motor unit action potentials of low amplitude with a nearly normal interference pattern consistent with active polymyositis.

A muscle biopsy specimen showed many focal areas of inflammation and necrosis, with a large number of regenerating fibres, and very prominent areas of granuloma formation with the presence of Langerhans giant cells without caseation or demonstrable acid fast bacilli (fig).

A diagnosis of sarcoid polymyositis was made and the patient was started on prednisolone 40 mg daily together with isoniazid because of the evidence of previous tuberculosis. Over the next 4 weeks he made a marked symptomatic improvement and the CK fell to 130 IU/l. The rare association of dermatomyositis with sarcoidosis has been reported in two Japanese patients.\(^1\) The case reported by Itoh\(^1\) had a typical rash of dermatomyositis with muscle weakness but a normal CK. Active polymyositis with elevated CK may also occur.\(^3\)

In the present report the patient had a relapsing illness over 6 years including three distinct episodes of muscle pain and weakness, two of which were associated with skin rash. The CK was elevated during these episodes. In the first attack the principal problem was pain and stiffness, the CK being only slightly elevated, and the patient responded to a 3 week course of steroids. The second attack involved mild proximal muscle weakness and settled spontaneously after 3 months. The third episode, which again responded to steroids, was associated with mild muscle pain, considerable proximal muscle weakness and a markedly elevated CK.

Symptomatic muscle weakness is uncommon in sarcoidosis although up to 50% of muscle biopsies in asymptomatic patients may reveal granulomata.\(^4\)\(^5\) Muscle involvement in sarcoidosis has been divided into an asymptomatic form with granuloma, a nodular form which may be palpable, and an acute or chronic form associated with muscle weakness.\(^6\)\(^7\)

Symptomatic granulomatous myositis has been described not only in sarcoidosis but also in Crohn's disease,\(^8\) miliary tuberculosis, collagen vascular disease, possibly mixed connective tissue disease\(^9\) and in association with thymoma.\(^10\) The clinical picture in sarcoid myositis may be difficult to differentiate from subacute polymyositis particularly when there are no other clinical features of sarcoidosis and the diagnosis may only be made by muscle biopsy.

The case we describe here is complicated both by the radiological evidence of previous tuberculosis infection and by the occurrence of colonic carcinoma. Involvement of striated muscle by a primary tuberculous infection has been rarely observed.\(^11\) The positive tuberculin test is compatible with a diagnosis of sarcoidosis.\(^12\) The histological changes occurring in myositis associated with carcinoma are those of widespread muscle fibre degeneration, infiltrates of inflammatory cells, regeneration from surviving fibres, and fibrosis. Although localised tuberculoid granulomas have been described in lymph nodes in association with carcinoma\(^13\) there are no published reports of granulomatous myopathy in this context. Sarcoidosis would therefore seem to be the most likely cause for this patient’s granulomatous myositis. This is supported by the positive Kveim test, and the normal serum ACE level does not exclude this diagnosis. Tanaka \etal\(^14\) have reported the association of mixed connective tissue disease with a granulomatous polymyositis but the diagnosis of sarcoidosis was not completely excluded in their case. Active tuberculosis may be rarely associated with granulomas in muscle. There was no evidence that our patient had active tuberculosis. All of the cases of dermatomyositis

![Fig](http://jnnp.bmj.com/ on April 20, 2017 - Published by group.bmj.com)
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Associated with sarcoidosis have been in Japanese subjects suggesting the possibility of a genetic predisposition.

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References


Membranous glomerulonephritis and Grover disease with diverse neurological abnormalities: an immunological disorder due to poliovirus infection?

SIR: Guillain-Barré syndrome associated with the nephritic syndrome was reported and the immunological pathogenesis was suspected in the early 1970s.1,2 We here report a case of membranous glomerulonephritis and transient acantholytic dermatosis (Grover disease) with unique neurological abnormalities. This appears to be the first reported case of such a combination of disorders.

A 32 year old Japanese man was well until July 1984 when he gradually developed a low grade fever of 37–38°C, general malaise, occipital headache, and severe dysaesthesia in the lower extremities. Neurological examination on admission in late August revealed signs of meningeal irritation, diminished deep tendon reflexes, hypeaesthesia in the distal parts of the extremities, positive nerve stretch signs of the sciatic and femoral nerves, and mild gait ataxia. Signs of cerebellar dysfunction such as gaze nystagmus, scanning speech, and limb ataxia appeared in late September. All of the neurological abnormalities progressed gradually. In early October small vesicular papules appeared on the face, trunk, and proximal parts of the extremities. Past history was negative except that he had had the trivalent oral poliovirus vaccine at 12 years of age. His 2 year old daughter had this vaccination in the autumn of 1983.

On admission he had the nephritic syndrome with proteinuria ranging from 2 to 4 g/day. At the height of the illness, ESR was 88 mm/hour, and CSF contained 30 lymphocytes/µl, 250 mg/dl protein, and 29 mg/dl IgG. Peripheral nerve conduction velocity and short latency sensory evoked potential studies indicated abnormalities in the spinal nerve roots and peripheral nerves. Although complements level was low, the immune complex, cryoglobulin, LE test, anti-nuclear antibody, anti-DNA antibody, anti-RNP antibody, and anti-Sm antibody were repeatedly negative in the serum. There was marked elevation of the poliovirus type 2 antibody titre in the serum: neutralisation test (NT) was 1:2048 and complement fixation test (CF) was 1:16 in early September. Other viruses showed no remarkable elevation of the antibody titres.

The diagnosis of stage 2 membranous glomerulonephritis was made by kidney biopsy, which revealed electron dense deposits along the subepithelial processes of the basement membrane. Skin biopsy showed intra-epidermal vesicles with acantholytic changes, with suprabasal clefs at acro- syringium. Direct immunofluorescence revealed deposition of IgG at the dermo-epidermal junction. Muscle biopsy of the gastrocnemius showed a mild degree of mononuclear cell infiltration of small blood vessels in the perimysium, a finding of non- specific vasculitis.

Prednisolone 60 mg/day was started in mid October, and all of the abnormalities subsided rapidly. The skin lesions were diagnosed as Grover disease from the clinical course and the biopsy findings retrospectively. The poliovirus type 2 antibody titre declined gradually; NT was 1:128 and CF was <1:4 in November.

Membranous glomerulonephritis is a well known immune complex disease. Grover disease is a rare vesicular skin disease first described in 1970,3 and although the aetiology is still unknown, immunological disturbance has been suspected to play an important role.4 This case is unique in that the hitherto undescribed neurological abnormalities developed almost simultaneously with these disorders, and in that all responded dramatically to prednisolone. The neurological abnormalities of this case can be summarised as involving the leptomeninges, cerebellum, and spinal nerve roots and peripheral nerves, indicating a dissemination of the lesions throughout the nervous system. Under these circumstances it would be reasonable to speculate that the neurological abnormalities were also due to the same immunological disturbance that caused both the renal and dermatological abnormalities, although the reason is unclear why the neurological abnormalities preceded the others.

It is of particular interest in this case that the level of the CSF protein fluctuated in parallel with that of the urinary protein. The anatomical and physiological similarity of the choroid plexus and renal glomerulus has been pointed out by several workers,5–6 and the immune complex deposition in the choroid plexus has been observed in such immunological disorders as systemic lupus erythematosus, acute serum sickness, and experimental allergic encephalomyelitis.5,7

Although the precise mechanism of the elevation of CSF protein cannot be determined, choroid plexus damage due to the immunological disturbance mentioned above may have contributed in this case. The primary cause of this immunological disturbance could not be determined: the antigen was not identified. The antibody titre of the poliovirus type 2, however, was remarkably elevated in the early phase of the illness, and declined gradually thereafter. Although this might represent only a coin-

Accepted 29 August 1987
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*J Neurol Neurosurg Psychiatry* 1988 51: 311-313
doi: 10.1136/jnnp.51.2.311-a

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