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Anti-Jo-1 positive inclusion body myositis with a marked and sustained clinical improvement after oral prednisone

The three major categories of the idiopathic inflammatory myopathies are dermatomyositis, polymyositis, and inclusion body myositis.¹ The most important clinical feature distinguishing inclusion body myositis from dermatomyositis and polymyositis is the lack of responsiveness to immunosuppressive treatment.^{1,2} Yet, it is the experience of many clinicians that a small subgroup of patients with inclusion body myositis show at least a partial response to immunosuppressive treatment. There are no specific characteristics which can identify this subgroup. In this report we present an anti-Jo-1 positive patient with inclusion body myositis who showed a marked and sustained clinical improvement after treatment with oral prednisone.

A 74 year old man, with no relevant medical history, presented with a slowly progressive proximal muscle weakness of the lower limbs. On presentation there were no complaints of muscle weakness of the upper limbs, dysphagia, myalgia, arthralgia, or feelings of general malaise. He denied any sensory symptoms. His family history was negative for neuromuscular or rheumatic disorders and he did not use any myotoxic drugs.

Physical examination showed asymmetric proximal and distal muscle weakness (muscle strength in MRC grades: neck flexors 3, right and left triceps 4, right and left iliopsoas 4, right and left gluteus maximus 4, right quadriceps 4, left quadriceps 2.5, right and left hamstrings 4, right and left anterior tibial 2, left gastrocnemius 4, all other muscles 5), marked atrophy of the quadriceps muscles and low symmetric tendon reflexes. The muscles were not painful to palpation. All other aspects of the general and neurological examination were normal.

Laboratory investigations showed slightly increased concentrations of serum creatine kinase (260 U/l; normal <200 U/l). All other aspects of the routine laboratory investigation including erythrocyte sedimentation rate, lactate dehydrogenase, vitamins, thyroid function tests, and antinuclear factor were normal or negative. Serum was screened for the presence of myositis specific autoantibodies (MSAs) using immunoblotting, enzyme-linked immunosorbent assay (ELISA), and immunoprecipitation as previously described.³ Assays were positive for the anti-Jo-1 autoantibody.

Electromyography demonstrated fibrillation potentials with positive sharp waves, polyphasia, short duration small amplitude motor unit potentials, and several high amplitude motor unit potentials in proximal and distal muscles. Nerve conduction studies were normal.

Figure 1 Cumulative MRC scores (MRCsum) and dosage of oral prednisone (mg/day) over time. Cumulative MRC score is calculated by adding the MRC grades of the following muscle groups: neck flexors, neck extensors, biceps, triceps, forearm flexors, forearm extensors, iliopsoas, gluteus maximus, quadriceps, hamstrings, anterior tibia, and gastrocnemius.

Muscle biopsy of the right quadriceps muscle showed the presence of small endomysial inflammatory infiltrates, invasion of non-necrotic muscle fibres, basophilic rimmed vacuoles, increased number of muscle fibres containing internal nuclei, ragged fibres, atrophic muscle fibres, and positive staining of the sarcolemma for HLA-ABC. Electron microscopy demonstrated the presence of 15-18 nm tubulofilaments in the cytoplasm. The diagnosis of definite inclusion body myositis was made.²

Because the patient was in good general health and the degree of inflammation on muscle biopsy was rather extensive, the decision was made to start treatment with oral prednisone (60 mg once daily). Three months after the initiation of treatment a marked improvement of muscle strength was found (fig 1), serum creatine kinase had normalised (62 U/l), the anti-Jo-1 autoantibody was no longer detectable, and EMG demonstrated a significant improvement with less spontaneous activity and fewer short duration small amplitude motor unit potentials. Prednisone was slowly tapered and stopped 1 year after the initiation of treatment. Muscle strength remained stable (muscle strength in MRC grades 18 months after treatment initiation: right and left triceps 4, right and left quadriceps 4, right and left hamstrings 4, right and left anterior tibial 2, all other muscles 5).

What makes this particular case of great interest are the presence of the anti-Jo-1 autoantibody and the marked and sustained clinical improvement after treatment with oral prednisone. Although a few studies have reported mild improvement in inclusion body myositis with various immunomodulating agents, significant and sustained improvement remains extremely rare in inclusion body myositis.^{1,2}

The anti-Jo-1 autoantibody is the most prevalent MSA and is found in 25% of patients with dermatomyositis and patients with polymyositis.⁴ In patients with inclusion body myositis the antibody is hardly ever detected.^{4,5} Until now, only three patients with inclusion body myositis have been reported in whom the antibody was found.³ Unfortunately, the clinical picture of these patients was not described. The relative absence of Jo-1 in inclusion body myositis is seen as support for the hypothesis that the immune response in this disease differs from that in dermatomyositis and polymyositis.⁴ It has therefore been suggested that the anti-Jo-1 autoantibody can aid in the differential

diagnosis between the three entities by virtually excluding inclusion body myositis in cases of anti-Jo-1 positivity and therefore providing an additional argument for the start of immunosuppressive therapy.⁴

The clinical and electrophysiological improvement after immunosuppressive therapy in the presented patient, together with the presence of a disease specific autoantibody, suggests a prominent role of the inflammatory response. Although it is only based on one case history, this report raises the question whether the presence of an MSA can aid in the identification of patients with inclusion body myositis who might show a response to immunosuppressive therapy. It is not known how MSAs are generated and whether they represent an epiphenomenon or whether they are somehow involved in the pathogenesis of idiopathic inflammatory myopathies. Based on their specificity for myositis it does seem likely that they are the result of a yet unidentified immunological mechanism which is specific for idiopathic inflammatory myopathies and which is, directly or indirectly, linked to the occurrence of clinical myopathy. It can be hypothesised that the presence of an MSA in inclusion body myositis is the result of an identical immunological mechanism as in MSA positive patients with dermatomyositis or polymyositis and that thus immunosuppressive treatment would be of benefit, as in this patient. Additional studies are required to consider these questions.

In conclusion, the present data suggest that in a patient with an idiopathic inflammatory myopathy, even inclusion body myositis, and the presence of an MSA, immunosuppressive treatment should be started and continued for at least 3 months.

We thank WTM Vree Egberts, BAW de Jong, and E Nuy-Terwindt for their expert technical assistance. This work was supported by grant 93-1112 of the Prinses Beatrix Fonds and grant 940-37-009 of MW-NWO.

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Upholsterer's PIN

Posterior interosseous nerve (PIN) palsy has been recognised at least since 1905 when Guillain and Courtellemont described a case in an orchestral conductor. In 1899 Gowers described involvement of the muscles of the

forearm in radial nerve palsy but did not specifically note damage to its important branch, the PIN. During the first world war Tinel described the anatomy without mentioning the nerve by name and without discussion of causation. By the end of the second world war the PIN had been so named.¹

The PIN consists of one bundle of motor nerve fibres at the arcade of Fröhse (the supinator arch) but divides into two bundles near its point of exit from the supinator muscle. The recurrent superficial motor branch innervates the extensor digitorum, extensor digiti minimi, and extensor carpi ulnaris; the descending or deep motor branch innervates the abductor pollicis longus, extensor pollicis brevis, extensor pollicis longus, and extensor indicis.²

The syndrome is also known as the supinator syndrome. It is distinguished from a seventh cervical root syndrome in which the triceps is involved but the wrist extensors usually are spared.

Palsy of the PIN produces the characteristic syndrome of weakness of extension of the digits and thumb with preservation of extension of the wrist which, nevertheless, may deviate radially. Sensory function, supination, and brachioradialis are not affected. Pain is often felt lateral to the elbow.

Palsy of the PIN may be caused by acute trauma in gunshot wound or pressure from a space occupying lesion. Repetitive use of the forearm has caused PIN palsy in a bartender, violinist, swimmer, conductor, waiter, factory worker, and an embroiderer. It has also been reported with strenuous arm exercises, snow shovelling, in frisbee games and tennis playing, and after the prolonged carrying of an M60 machine gun. It has also been reported after cannulation of a forearm vein.³

We report a case of PIN palsy after repetitive movements of the forearm and we have named this variant "upholsterer's PIN".

A 47 year old right handed woman, an enthusiastic pianist, at an upholstery class one evening used both hands to compress springs and apply clips to the springs. During that evening she noted aching around the lower part of the right arm and on the next day found she could not extend the right thumb and fingers. She had not slept heavily on that side and had not noted trauma to the

elbow. She did not have cervical pain or numbness.

The region of the lateral epicondyle of the elbow was tender. Finger flexion and volar flexion at the wrist were strong as were flexion and extension of the elbow, supination, and pronation. Examination failed to show movement in the extensor digitorum and extensor pollicis longus.

An EMG showed changes typical of a PIN lesion—namely, active denervation in the extensor carpi radialis brevis, extensor digitorum communis, and extensor carpi ulnaris. The brachioradialis, biceps, and triceps brachii were normal as was sensory conduction in the superficial branch of the radial nerve. The active denervation noted in the extensor carpi radialis longus is an anomaly and suggests aberrant innervation or anatomical anomaly.

At exploration the PIN was degenerate from the origin of the superficial radial nerve to the branch of the brachioradialis although it did not seem compressed. An 8 to 10 mm segment of obvious abnormality within the PIN itself was resected and the nerve ends sutured together. The patient continued to recover function and did not require tendon transfer as her functional deficit substantially improved.

The histology of the excised segment showed very severe demyelination, presumed to be the primary pathology. Demyelination secondary to underlying axonal disease could not be excluded (figure 1).

The clinical features were typical of a PIN palsy. However, EMG studies showed active denervation in extensor carpi radialis longus and extensor carpi radialis brevis. As the extensor carpi radialis longus is innervated by the radial nerve before the origin of the PIN, it is possible to distinguish a PIN palsy from a more proximal radial nerve injury, which typically produces wrist drop and has also been reported after repetitive muscle contraction.⁴

We suggest that focal demyelination in the PIN and conduction block resulted from ischaemia due to direct pressure on the nerve at an area of anatomical constriction as a result of the repetitive action of the surrounding muscles.

Patients with PIN palsy sometimes report a history of forceful, repetitive rotation of the

forearm preceding the onset of symptoms. Because we do not know of a report of PIN palsy in association with the compressing of springs, we have named this palsy "Upholsterer's PIN".

We are extremely grateful to Dr Elias Ragi for the nerve conduction studies and to Dr R Simpson for supplying the photographs illustrating nerve demyelination.

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CORRESPONDENCE

Neurocysticercosis and epilepsy in developing countries

I have several comments to make about the article by Pal *et al*¹ on neurocysticercosis and epilepsy in developing countries.

The title should refer to "neurocysticercosis in India" because almost all examples deal with that country, despite the fact that, as the authors correctly state, it is a disease found in many developing countries. Furthermore, several statements made by the authors do not reflect the knowledge gathered in Mexico, which is summarised in this letter.

The authors indicate in the text that "Population control measures have not shown long term effectiveness" . . . "There is little epidemiological data on risk factors" . . . "Migration from the countryside and the risk of urban slums obviously influence the changing epidemiology" . . . "mass treatment with praziquantel may produce early benefit but larger term evaluation shows no lasting impact". Sarti *et al* have studied exhaustively risk factors² and evaluated control measures in Mexico; the results indicate that the main risk factor is the tapeworm carrier in the close environment, and that treatment of tapeworm carriers or health education of the community are useful control measures as evaluated at 6 and 42 months after their implementation in rural populations.^{3,4} Moreover, vaccines for swine cysticercosis are being successfully developed and evaluated.⁵

Multiple studies performed in various countries clearly show the usefulness of serological methods for diagnosis, including antigen detection, which Pal *et al*¹ indicate was recently developed, but there are several prior references including that of 1989 by Correa *et*

Figure 1 Nerve biopsy stained by osmium tetroxide shows demyelination with some residual myelin in globules.