inflammatory polyneuropathy have been described in which there was a combination of limb weakness and an internal and external ophthalmoplegia but in which limb ataxia was not a feature.\(^7\)

Al-Din et al\(^9\) reported a series of cases with clinical features similar to those of the Miller Fisher syndrome but which were considered to have brainstem encephalitis, some on the basis of computed tomography and postmortem examination. It was suggested that a spectrum might exist ranging between cases with pure brainstem involvement at one end, through mixed cases, to those with pure peripheral nerve involvement at the other. The present patient showed no evidence of CNS involvement apart from some delay in the SSEPs. The consensus opinion, however, is that the Miller Fisher syndrome represents a peripheral nerve disorder. Nevertheless, it is often difficult to demonstrate abnormalities of nerve conduction in the syndrome. Cases with relatively minor changes in motor nerve conduction and more obtrusive changes in sensory conduction have been documented.\(^10\) Those in the present case were modest but definite.

The mechanism of tremor in the Miller Fisher syndrome is unknown but a peripheral disturbance is generally assumed.\(^2\) It may involve a mismatch between afferent proprioceptive signals from muscle spindles and those from Golgi tendon organs.\(^11\) A central disturbance, related to a disruption of central cerebellar connections is not excluded, but lacks pathological substantiation.

Primary amyloidosis in a patient with myotonic dystrophy

Sir: Several immunoglobulin abnormalities have been described in myotonic dystrophy, a hereditary progressive muscle disorder transmitted as an autosomal dominant trait. These include hypogammaglobulinaemia,\(^1\) a shortened half-life of \(^131\)I-labelled magmoglobin,\(^2\) increased catabolism of IgG,\(^3\) and diminished antibody production to certain antigens.\(^4\) We describe the hitherto unreported occurrence of amyloidosis in a patient with myotonic dystrophy and hypogammaglobulinaemia.

A 47-year-old garage manager known to have myotonic dystrophy since 1947, was referred in September, 1981 with exertional left inframammary chest pain and paraesthesia in the left arm refractory to glyceryl trinitrate, metoprolol and nifedipine. On examination he had generalised muscle wasting with sternomastoid atrophy, frontal balding, testicular atrophy and early bilateral cataracts. Tendon reflexes were diminished, he had difficulty relaxing handgrip and myotonic contraction could be elicited by percussion of thenar eminences. His ECG, exercise ECG, echocardiograph and coronary angiogram were normal although left ventricular end diastolic pressure was 15 mm Hg. In January, 1982 he developed nephrotic syndrome. Investigations in-
cluded: creatinine clearance 90 ml/min; urinary protein 5G/24 h (non-selective) serum albumin, 20 g/l; ESR 108 mm in 1 h; ANF and anti double-stranded DNA, negative; ASO titre < 200 Todd units/ml; and serum complement normal. Serum IgA and IgM were normal but IgG was reduced (1.39 g/l). Plasma and urine electrophoresis and immunoelectrophoresis showed no monoclonal band. Renal biopsy showed a mesangial-proliferative glomerulonephritis with granular IgM and scanty deposits of C3, C1q and fibrin in the mesangium. Congo red stain showed amyloid in the glomeruli (fig). Amyloid fibrils were seen on electron microscopy.

Between January and November, 1982 he was readmitted four times with left in- framammary pain, oedema and increasing muscle weakness. Upper gastrointestinal endoscopy, oesophageal manometry, barium swallow and acid provocation test showed a hypotonic upper gastrointestinal tract. Most ECGs during chest pain were normal, a few showed ischaemia, and twice an arrhythmia was noted (supraventricular tachycardia and atrial fibrillation). In December, 1982 he died following the development of left hemiparesis and bronchopneumonia.

At necropsy the heart was slightly en- larged, stiff and waxy in texture; the major coronary arteries were normal. Lungs showed lower lobe pneumonia with oedema. Kidneys were slightly enlarged with pale, smooth cortices. Microscopy showed peri- vascular amyloid in the lungs, spleen, liver, oesophagus, stomach, duodenum, thyroid, aorta, testes and brain. In the kidney, amyloid was deposited in the glomeruli and larger blood vessels. Perivascular and intra- mural amyloid was found in small vessels in the myocardium but luminal narrowing was slight. The individual myocytes and capillar- ies were surrounded by amyloid and electron microscopy revealed muscle fibres being re- placed by amyloid fibrils. Unfortunately skeletal muscle was not examined. The cere- bral arteries were normal. A recent infarct in the right basal ganglia and insular cortex was attributed to hypotension preceding ad- mission. Immunoperoxidase staining of amyloid was positive for lambda light chains.

Although cardiac abnormalities inter- fer ing with conduction, and minor abnor- malities of cardiac muscle on electron microscopy, are well recognised features of myotonic dystrophy,5 6 we are unable to find any previous report of amyloidosis. The amyloidosis gave rise to ischaemic cardiac pain, as has been previously described,7 but this could not be diagnosed during life by

![Fig Renal biopsy specimen showing glomerular deposits of amyloid seen as spiky protrusions on the epithelial aspect of glomerular basement membrane. (Congo red viewed under polarised light x200).](http://jnnp.bmj.com/)

reported in patients without proteinuria in myotonic dystrophy.1 4 The relation be- tween the previously described immu- noglobulin abnormalities in myotonic dystrophy and primary amyloidosis in the present case is unclear.

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