

Fig. 4 Histology

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Chronic inflammatory demyelinating polyneuropathy associated with membranous glomerulonephritis: case report

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Summary A case of Chronic Inflammatory Demyelinating Polyneuropathy (CIDP) with Membranous Glomerulonephritis (MGN) is reported. This is the second case recorded in the literature and the article compares this case with the other reported case, including immunological implications. © 2000 Harcourt Publishers Ltd

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INTRODUCTION

Chronic Inflammatory Demyelinating Polyneuropathy (CIDP) describes an uncommon disorder of progressing or relapsing demyelinating peripheral neuropathies initially reported by Austin in 1958. The onset is usually insidious and maximal severity may

not be reached for months or even years. Dyck et al. (1975) defined the process physiologically as slowed nerve conduction velocities, disproportion between weakness and sensory loss, and an elevation of the CSF protein, notably gamma globulin. Rarely, CIDP occurs in association with solid tumours of the lung and gastrointestinal tract and has been described with immune mediated conditions including chronic active hepatitis, inflammatory bowel disease, Hodgkin's disease, monoclonal gammopathy and thyrotoxicosis.³ CIDP usually has a good response to immunosuppressive therapy.^{2–4}

Membranous glomerulonephritis (MGN) is characterised by irregular proteinaceous deposits containing IgG, along the outer aspect of the glomerular capillary wall.⁵ The capillary wall thickens with increased amounts of basement membrane-like material. Approximately 80% of cases present with overt nephrotic syndrome, while the remainder have isolated proteinuria. Associations include systemic lupus erythematosus, chronic infections, solid tumours of the lung and gastrointestinal system, and drugs such as penicillamine and captopril. Spontaneous remission occurs in approximately 20 to 40% of adults.

CASE HISTORY

The patient, an 81 year old female, was admitted for the investigation of progressive weakness of the lower limbs associated with lumbar back pain. The weakness had been evolving over a 3-month period and was associated with several falls. On examination, there was weakness of the proximal muscles, graded 3 to 3.5 out of 5. Reflexes were present, although reduced. There was decreased sensation to pin prick in the dermatome of L3 on the right-hand side.

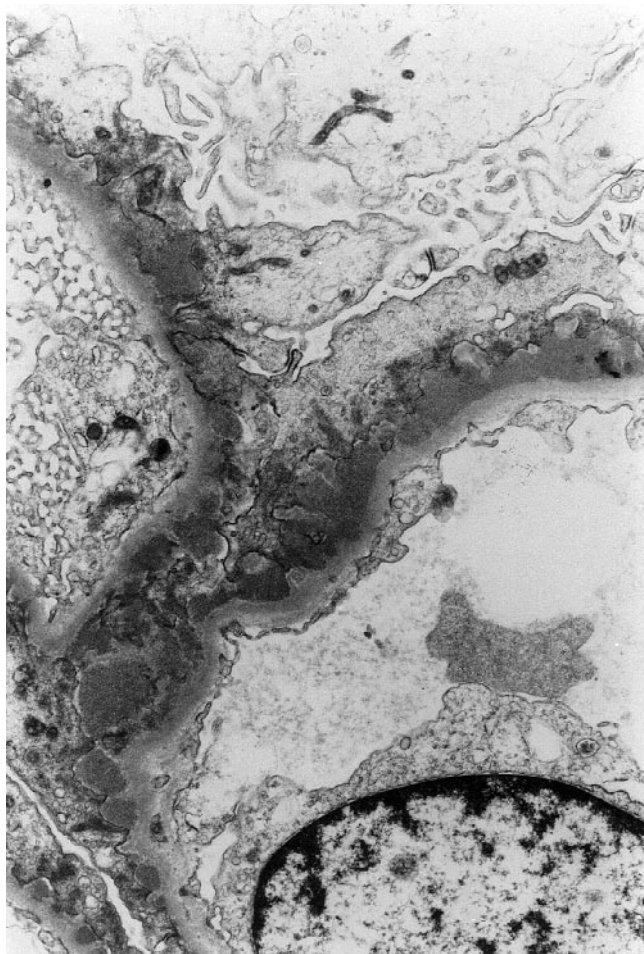


Fig. 1 Renal Biopsy. (Electron Microscope: Phillips CM-10).

Table 1 EMG Report

Nerve	Latency (ms)	Amplitude	Velocity(M/s)
Sensory action potentials			
Right Median		0 uV	
Right Ulnar		0 uV	
Motor Conduction			
Right Median	11.8	0.3 mV	
	16.4	0.4 mV	41
Right Ulnar	4.1	1.6 mV	
	14.3	2.3 mV	27
Right common peroneal		0 uV	
Right posterior tibial		0 uV	

Imaging of the spinal cord was undertaken. Lumbar myelogram showed severe canal stenosis at L4/5, with a mild disc bulge at L3/4. Cervical spine imaging showed narrowing of the C3/4, 5/6 and 6/7 disc spaces with associated prominent osteophytes. The patient had a decompressive laminectomy at the levels of L4/5 and T12/L1. Post laminectomy, the lumbar pain improved, although the weakness in the legs continued to be a problem and the patient suffered multiple falls. She was unable to cope with activities of daily living and was transferred to a rehabilitation unit.

Her leg weakness continued. She developed paraesthesias in the lower limbs and weakness of the upper limbs. On examination she had absent reflexes in both upper and lower limbs and loss of joint position sense which was more marked in the lower limbs. Fasciculations were noted in the right triceps muscle.

Treatment at this stage was complicated by a deep venous thrombosis with a suspected pulmonary embolus. However, the pleuritic chest pain was later proven to be due to several fractured ribs that she had sustained in a fall one week prior. She was treated with intravenous heparin.

CIDP was suspected and a lumbar puncture was performed. The total CSF protein was 1.2g/L units (ref range 0.15–0.45g/L). The results of nerve conduction studies are shown in Table 1. Sensory action potentials were absent. Distal motor latency was prolonged in the right median nerve, consistent with a demyelinating neuropathy.

Peripheral bilateral pitting oedema and hypoalbuminaemia (serum albumin was 29g/L, ref range 35–40) were features at this time and proteinuria was noted at 4+. Urinary protein loss was 3 g per day. Anti-nuclear antigen and complement assays were normal as was hepatitis B serology. A renal biopsy was performed and showed changes of MGN (Fig. 1) All capillary loops (15 glomeruli) showed diffuse thickening of the basement membranes with tubulo-interstitial structures well preserved. Immunofluorescence was positive for IgG and C3 (granular pattern) along capillary loops, and electron microscopy showed extensive subepithelial deposits along capillary loops with foot process effacement and early spike formation.

Treatment for the CIDP was commenced initially with 3 plasma exchanges followed by oral steroid therapy (prednisolone 60 mg daily) and azathioprine (100 mg daily). At the time of writing, 7 months after starting treatment, there was mild improvement in symptoms and signs and further plasmapheresis was being considered.

DISCUSSION

The patient's clinical abnormalities, nerve conduction studies and elevated CSF protein are consistent with the diagnosis of CIDP. A

sural nerve biopsy was not performed because of severe pedal oedema which might have impaired wound healing and increased the risk of infection.

The only other report of a patient with both CIDP and MGN is that of Kohli et al.³ The history was very similar to that of our patient and involved an 18 year old male who presented with lower limb weakness of 6 weeks duration. He was treated with prednisolone and showed significant improvement in power after 2 weeks, although proteinuria was unchanged. At 4 months follow-up, power had returned to normal, despite absent deep tendon reflexes and persistent proteinuria.

Our case, however, differs from that of Kohli et al. in that our patient is much older and had a poor response to immunosuppression. In a study of 92 cases of CIDP⁴ 73% of patients were not seriously incapacitated by their CIDP following immunosuppression and/or plasma exchange and were independent.

In a study of 72 adult patients⁶ with idiopathic MGN, 92% having proteinuria 3 g over 24 h or more were studied for the clinical evolution of the disease. At 10 years, 46% were in complete or partial remission. Ponticelli et al.⁷ described a treatment regimen for idiopathic MGN consisting of methylprednisolone and chlorambucil, and stated that this combination may protect renal function and increase the chance of remission of the nephrotic syndrome, as compared with a control group being treated with methylprednisolone alone.

The possibility that both CIDP and MGN could have occurred in the same patient at approximately the same time by chance has been considered, but is highly unlikely. Other possible pathogenic mechanisms of this scenario have also been considered, such as chronic antigenaemia or an infectious aetiology; these are certainly relevant to membranous disease, if not the neurological condition.

MGN at this age raises the suspicion of underlying malignancy, though none has been found so far in this patient. However, neoplasia does not usually occur in association with CIDP. A careful search for other possibilities, including a drug-related cause, has been investigated with no success.

There already exists an extensive list of immune mediated conditions that are observed with CIDP. This case and the case of Kohli³ strengthen the argument that CIDP has wider immunological associations than are currently considered.

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