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Short communication

Relapsing Guillain Barré Syndrome and nephrotic syndrome secondary to focal segmental glomerulosclerosis

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Abstract

A 49-year-old man developed simultaneously a Guillain Barré Syndrome (GBS) and a nephrotic syndrome (NS). The patient relapsed twice, despite treatment with intravenous immunoglobulins (IVIg) after a full or partial recovery, and became resistant to IVIg. Renal biopsy revealed focal segmental glomerulosclerosis (FSGS). He responded to plasmapheresis and corticosteroids with simultaneous recovery of his GBS and NS, suggesting a common pathogenesis of the two conditions.

Keywords: Guillain Barréx Syndrome; Nephrotic syndrome; Focal segmental glomerulosclerosis; Relapse; IVIg

1. Introduction

The association of Guillain Barré Syndrome (GBS) and renal abnormalities was reported as early as 1918, when Bradford et al. reviewed 30 cases of acute progressive inflammatory polyradiculoneuropathy in an attempt to provide a uniform classification of this disorder [1]. The spectrum of renal manifestations in GBS ranged from asymptomatic proteinuria to overt nephrotic syndrome (NS) [2–4]. Most cases are related to membranous glomerulonephritis, although minimal changes disease, focal interstitial nephritis, and immune complex glomerulonephritis have been reported [5,6].

We describe a case of relapsing GBS with NS related to a focal segmental glomerulosclerosis (FSGS). We will present a review of pertinent literature and discuss the pathogenesis of the association between the two conditions. To the best of our knowledge, this is the first case report of simultaneous relapsing GBS and NS with FSGS.

2. Methods: case report

2.1. Results

2 weeks before his hospitalization, a previously healthy 49-year-old man developed an ascending paresthesias, weakness, and swelling of his feet that progressively spread to his hands. Approximately 5 days prior to admission, the patient reported worsening weakness to the point that he was unable to walk without help. He also noticed loss of hand dexterity. He denied any falls, swallowing difficulties, diplopia, or urinary symptoms. The patient received travel immunization before going to Africa 6 months prior to admission. He had no blood transfusions. Physical examination demonstrated a blood pressure 206/119, weakness in his lower extremities, predominantly in the distal muscles (Medical Research Council 4/5), and absence of deep tendon reflexes except for the right biceps. Decreased vibration in the lower extremities and reduced pinprick sensation in

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both hands and feet were noted. Laboratory tests showed urea 20 mg/dl (normal 8–25 mg/dl), creatinine 1.4 mg/dl (normal 0.6–1.5 mg/dl) creatinine clearance 72 ml/min, proteinuria (10 g/24 h), and a low serum albumin of 2.3 mg/l (normal 3.1–4.3 mg/l). Immunological work up showed normal complement levels, negative anti-nuclear antibody, anti-neutrophil cytoplasm antibody, anti-neuronal antibody, cryoprotein, and rheumatoid factor. Serum protein electrophoresis and immunofixation revealed moderate diffuse decrease in gamma globulins with IgG of 319 mg/dl (normal 614–1295 mg/dl) and a very low concentration band in the slow gamma region identified as IgG kappa M component. Bence–Jones protein was not detected in the urine. The following serologies were normal: *Campylobacter jejuni*, hepatitis B and C, cytomegalovirus, Epstein–Barr virus, Lyme, and Venereal Disease Research Laboratory. Thyroid stimulating hormone and vitamin B12 levels were normal. Stool study was negative for enteric pathogens. CSF studies were normal. Nerve conduction studies showed small and markedly dispersed tibial motor responses, prolonged left peroneal distal motor latency (8 ms) with a normal amplitude motor response, and absent right H reflex. Needle EMG showed neurogenic pattern of recruitment with normal motor unit potentials in several muscles of the right lower extremity. These findings were consistent with a primary demyelinating polyneuropathy.

The patient received IVIG 0.4 g/kg/day for 5 days, and his strength improved dramatically. There was mild residual weakness in his feet, but he was able to walk without difficulty at the time of discharge. His blood pressure was reduced with ACE inhibitors. His renal function improved with decreased proteinuria to 4.9 g/24 h at 9 days after admission. He was stable for 10 days after his discharge. Then he reported recurrent tingling in his toes and fingers that progressed over one week and was followed by weakness. On the day of his second hospital admission, which was approximately 3 weeks after the first one, he had difficulty walking more than a few steps. Neurological examination demonstrated bilateral weakness (4/5 in upper extremities and 3/5 to 4/5 in the lower extremities), generalized areflexia, and decreased sensation to vibration, pinprick, and light touch in the lower extremities. A second electromyographic study (EMG), performed approximately 4 weeks after his first study, showed more dispersion of the tibial motor responses and further prolongation of the tibial F waves, bordering on the demyelinating range. His renal function demonstrated urea 16 mg/dl (normal 8–25 mg/dl), creatinine 1.4 mg/dl (normal 0.6–1.5 mg/dl) creatinine clearance 70 ml/min, proteinuria (5.2 g/24 h), and his albumin was 1.9 g/dl (normal 3.1–4.3 mg/l). He was treated again with 5 days of IVIg. His strength improved significantly, but unlike during his first admission he presented with residual weakness in his feet. At the time of discharge, he was able to walk with a cane, and his renal function was stable. Approximately 5 weeks after his first admission, he was admitted for a third time because of progressive numbness and weakness of his legs. On the day of admission, he was not able to walk. Neurological examination demonstrated normal strength in his upper extremities and marked weakness of his lower extremities 3/5 to 4/5. His third EMG, performed ten days after the second EMG, showed substantial interval progression of the nerve conduction abnormalities; the right median F latencies were prolonged in the demyelinating range, and the tibial and peroneal F waves were no longer present. He did not improve with 5 days of IVIg. His weakness progressed to 2/5 in the lower extremities and 4/5 in the left deltoid. He also developed severe urinary retention requiring an indwelling catheter. His renal function urea 22 mg/dl (normal 8–25 mg/dl), creatinine 1.7 mg/dl (normal 0.6–1.5 mg/dl) creatinine clearance 56 ml/min, proteinuria (10.1 g/24 h), and his albumin was 1.6 g/dl (normal 3.1–4.3 mg/l).

A renal biopsy performed 5 weeks following his initial admission consisted of 2 cores and contained 12 glomeruli, of which two were globally sclerosed. Some of the remaining glomeruli were hypertrophied. In two glomeruli, focal segmental glomerulosclerosis was present with foam cells (Fig. 1). The proximal tubules showed prominent reabsorption droplets with rare focal acute tubular injury and with an occasional mitotic figure. Occasional red cells were noted within tubular lumens. Focal interstitial mononuclear cell infiltrate and rare eosinophils were noted. Interstitial fibrosis and tubular atrophy occupied about 15% of the cortex. Arterioles showed hyalinosis. Arteries were unremarkable. Immunofluorescence demonstrated IgG, IgM, IgA and C3 deposition in the glomerular basal membrane and mesangium. Electron microscopy of 2 glomeruli showed widespread podocyte foot process effacement. These features strongly support the diagnosis of focal segmental glomerulosclerosis.

The patient was started 60 mg/day of prednisone, and he received plasmapheresis using albumin in return (twice a week for 2 weeks). His muscle strength improved dramatically, and his urinary retention resolved. He was discharged on a gradual tapering dose of prednisone, and he continued on plasmapheresis (once a week for the next 5 weeks after his discharge). He fully recovered from his neurological symptoms and NS after approximately 8 weeks of his discharge. At that time, his renal function demonstrated urea 22 mg/l, creatinine 1.2 mg/l, creatinine clearance 84 ml/min, and proteinuria 1.2 g/24 h. Over more than two years of follow up, the patient did not have any weakness of sensory loss. However, he required low dose prednisone (5 mg every third day). Approximately 18 months after symptoms onset, his albumin and total protein levels were normal at 4.4 g/l and 7 g/l, respectively. Approximately 19 months after symptoms onset, his urea was 14 mg/l, creatinine 1.4 mg/l, and creatinine clearance 70 ml/min. His proteinuria was not performed 19 months after symptoms onset, but had decreased to 0.43 g/24 h by roughly 8 months after symptoms onset. Attempts to further reduce the prednisone dose were followed by numbness of his feet.