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A case of severe motor axonal polyneuropathy with monoclonal gammopathy of undetermined significance

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Background

Polyneuropathy associated with monoclonal gammopathy of undetermined significance (MGUS-PNP) has a chronic and slowly progressive course. Monoclonal gammopathy of undetermined significance (MGUS) with severe motor axonal polyneuropathy is rare.

Case

A 73-year-old man with no past medical history presented to our hospital with 6-month history of muscle weakness and progressive muscle wasting of both hands. The neurologic examination revealed atrophy in both thenar muscles. The manual muscle testing grades were 3+/5 for both finger flexors and finger abductors. In addition, fasciculation in the upper extremity muscles was observed. A tongue atrophy and gait imbalance was not observed. The Hoffman and Babinski signs were negative. The sensory examination was intact. Electrodiagnostic study revealed low amplitudes on compound muscle action potentials (CMAP) from median and ulnar nerve, bilaterally. There was no evidence of conduction block in multiple nerves tested. The sensory nerve action potentials (SNAP) were normal. There were 2+ fibrillation potentials and positive sharp waves in the right abductor pollicis brevis, left flexor carpi radialis, and bilateral first dorsal interossei. Magnetic resonance imaging showed normal with mild degenerating features. The working diagnosis at this point was motor axonal polyneuropathy involving upper extremities. Further laboratory investigations revealed elevated IgGκ 270,48mg/L(normal, 3.30~19.40mg/L) in serum protein electrophoresis. The serum M-protein level of the patient was 0.7g/dL(normal, <3g/dl) and the clonal plasma cell population in the bone marrow was 1.5% (normal, <10%). These results support the diagnosis of MGUS. The increased levels of antibodies indicated an association between neuropathy and IgG-MGUS. 6 months later, the patient's symptoms progressed to dyspnea. In a repeat electrodiagnostic study, the results showed worsened low amplitudes on CMAPs of upper extremities. In addition, decreased CMAP amplitudes were also found in lower extremities that had normal findings in previous study. Most of distal latencies and nerve conduction velocities were normal. SNAPs were completely normal. Abnormal spontaneous activities in needle electromyography were observed in four extremities. The manual muscle testing grades of extremities remained 3+/5. Activities of daily living were needed maximal

assistance because the patient was bedridden status due to a ventilator applied. Plasmapheresis and low dose steroid treatment was administered. One month following the treatment, dyspnea was persistent and ventilator remained.

Conclusions

In our case, the patient who had muscle weakness and progressive muscle wasting of the limbs was diagnosed motor axonal polyneuropathy associated with IgG-MGUS. Polyneuropathy associated with MGUS as a chronic and slowly progressive course, but we report a case of IgG-MGUS polyneuropathy involving only pure motor neurons that relatively rapidly progressed.