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A CASE OF CHRONIC INFLAMMATORY DEMYELINATING POLYRADICULONEUROPATHY ASSOCIATED WITH SYSTEMIC SCLEROSIS SUCCESSFULLY TREATED WITH RITUXIMAB

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Abstract

Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) is a rare subtype of peripheral neuropathy and it may be accompanied by connective tissue disease (CTD). Although there are very few case reports of CIDP associated with CTD, to our knowledge, no case related to systemic sclerosis has been reported in the literature. A 50-year-old male patient who had been followed up by the neurology clinic with a diagnosis of CIDP for 2 years was referred to us with newer onset skin stiffness, Raynaud's phenomenon, swelling and pain in the joints of the fingers. With a detailed evaluation, the patient was diagnosed with systemic sclerosis and the CIDP was considered to be associated with systemic sclerosis. The patient's neurological findings improved with rituximab treatment in addition to corticosteroid.

Keywords: Chronic inflammatory demyelinating polyradiculoneuropathy, systemic sclerosis, rituximab

INTRODUCTION

Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) is a rare and acquired immune-mediated neuropathy and the incidence of CIDP is estimated to be 1 in 100,000 persons (1,2). Generally, it is manifested by proximal and distal symmetrical weakness accompanied by sensory complaints with a progressive course for more than 8 weeks (2). The diagnosis of CIDP is based on clinical findings and signs of demyelinating changes in electro-diagnostic tests (2). The cause of CIDP is an autoimmune process consisting of humoral and cellular immunity, with an unknown trigger in most cases (3). CIDP can be observed during many connective tissue diseases, particularly in systemic lupus erythematosus (SLE) and Sjogren syndrome before or after the onset of symptoms (3).

Systemic sclerosis is an autoimmune connective tissue disease leading to fibrosis in the skin, internal organs, and vessels (4,5). The pathogenesis of the disease consists of vasculopathy, immune system activation, and diffuse fibrosis (5). The diagnosis of the disease is generally based on clinical findings, however systemic sclerosis-associated antibodies and nailfold capillaroscopy also support the diagnosis (5). Peripheral neuropathy associated with systemic sclerosis is a known manifestation, but CIDP has not previously been reported during systemic sclerosis. Here, we report a case of systemic sclerosis who presented with CIDP.

CASE REPORT

A 50-year-old male patient was admitted with the complaints of weakness and numbness in both the legs and arms, inability

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to walk without support, and joint pain. The patient had worked in the manufacturing of glass bottles with occupational silica exposure for many years. Two years ago, he started to have difficulty in swallowing, sitting, and climbing stairs, and inability to walk without support. Patient was evaluated by the neurology department and no pathology was detected in the lumbar puncture examination. Electromyography (EMG) revealed mixed type sensorimotor polyneuropathy and he was diagnosed with chronic inflammatory demyelinating polyneuropathy. Intravenous immunoglobulin (IVIG) therapy of 2 gr/gr monthly was started. The patient was followed up for 2 years in the neurology clinic with monthly IVIG treatment. Then, he was referred to us with the skin stiffness, Raynaud's phenomenon, swelling and pain in the joints of the fingers started a few months ago. On his physical examination, he could not stand without support and could not move alone. Muscle strength was 4/5 in both the upper limbs and 3/5 in the lower limb. Raynaud's phenomenon on the hands and feet, telangiectasia on the face, stiffness and redness of the skin of the hands, sclerodactyly, bilateral arthritis of metacarpophalangeal joints, proximal interphalangeal joints and wrists were detected. Antinuclear antibody and anti-Scl-70 antibody were positive. Computed tomography of the thorax revealed ground glass opacity compatible with non-specific interstitial pneumonia. The control EMG findings were compatible CIDP. The patient was diagnosed with CIDP associated with systemic sclerosis and methylprednisolone at 8 mg/day and 1 course of rituximab (1,000 mg 2 doses, 15 days apart) were given as treatment. At the control examination of the patient after 2 months, he could walk alone without any support, muscle strength was found to be 5/5 in both the upper and lower extremities, and no arthritis was found.

DISCUSSION

CIDP is formed as a because of the influence of environmental and genetic triggers (6). Despite the paucity of cohort studies in the literature, infection is the main trigger for developing CIDP (6). CIDP can be observed during many bacterial and viral diseases. Besides, the polyautoimmunity phenomenon is also a concept that should be mentioned in the cause of CIDP, suggesting an increased risk of developing another autoimmune diseases in a patient with an autoimmune disease. It is common for patients diagnosed with CIDP to be accompanied by an autoimmune disease at the time of diagnosis or at later stages during the disease. In the literature, many cases of CIDP associated with various autoimmune diseases, especially with multiple sclerosis, myasthenia gravis, SLE, Sjögren's syndrome, rheumatoid arthritis,

Hashimoto's thyroiditis, Graves' disease, type 1 diabetes mellitus, vitiligo, primary biliary cholangitis, and autoimmune hepatitis, have been reported (6). The neurologic involvement in systemic sclerosis contains the central and peripheral nervous system, but the central nervous system involvement is rare. Peripheral neuropathy is not rare in patients with systemic sclerosis with a prevalence of 27.37% (7). In peripheral nervous system, cranial, peripheral, cutaneous, autonomic, and entrapment neuropathies can be detected (7,8). During systemic sclerosis, trigeminal neuralgia, peripheral sensorimotor polyneuropathy, carpal tunnel syndrome are often observed (8). However, when we look at the literature, no case of CIDP associated with systemic sclerosis has been reported to date. There are options such as glucocorticoids, IVIG, rituximab, plasmapheresis for treating CIDP (9). If there is an underlying cause or disease, treatment should be administered for it as i ur case.

CONCLUSION

CIDP may be one of the rare neurological involvement patterns of systemic sclerosis and the underlying systemic sclerosis should be considered in both the evaluation and management of the patient to control findings of CIDP effectively.

Ethics

Informed Consent: We written informed consent for publication of clinical details was obtained from the patient. No ethics board approval was required for this case report.

Peer-review: Externally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: M.K., C.B., Concept: C.B., Design: C.B., Data Collection or Processing: M.K., C.B., Analysis or Interpretation: C.B., M.K., Literature Search: M.K., R.D., C.B., Writing: M.K., R.D., C.B.

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