CASE REPORT

AMSAN Variant of Guillain Barre Syndrome Progressing to Chronic Inflammatory Demyelinating Polyneuropathy in a Patient with Marfan's Syndrome and Pulmonary Tuberculosis

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Summary

We report a 40-year old man who has Marfan's syndrome and was recently diagnosed to have pulmonary tuberculosis when he presented with chronic cough. He was admitted with bilateral lower limb weakness which was ascending in nature. He eventually required ventilation. It was initially thought to be isoniazid-neuropathy. However stopping the drug did not improve the condition and the patient developed bilateral lower motor neuron 7th cranial nerve palsy. Nerve conduction, MRI and CSF studies were done to confirm a first case report of AMSAN variant progressing to CIDP in a patient with Marfan's syndrome and pulmonary tuberculosis.

Key Words: Guillain Barre syndrome, AMSAN, Marfan's syndrome, Tuberculosis

Introduction

A 40-year-old Malay man who has Marfan's syndrome, with aortic valve replacement done for severe aortic regurgitation, chronic renal failure secondary to long-standing hypertension and was on anti-tuberculosis therapy (isoniazid, rifampicin, pyrazinamide and ethambutol) for pulmonary tuberculosis, was admitted to the medical ward with progressive weakness of both lower limbs of one-day duration. It gradually ascended to the upper limbs, associated with numbness up to the face, dyspnoea and dysphagia. Physical examination revealed a lanky thin man with Marfanoid features, with nasal speech, floppy tetraparesis. Blood pressure was 136/60mmHg, pulse rate was 88 beats per minute and collapsing. Apex beat was displaced and there was an early diastolic murmur at the left sternal edge with no thrill and no Austin Flint murmur. Lungs were clear. Power was 0/5 of both shoulders and hip flexion-extension, 1/5 of both knee and elbow flexion-extension and there was generalized areflexia. There was patchy sensory loss in both upper and lower limbs, but no definite sensory level was detected. Lung function test indicated severe restrictive lung defect, with FEV1 / FVC being 0.62.

CSF examination revealed no leucocytes, normal protein level of 367mg/L, glucose level of 2.8mmol/L, staining with Indian Ink and TB PCR were negative. MRI of brain and spinal cord did not show any pathology.

Nerve conduction study (Figure 1) was in keeping with a demyelinating and axonal generalized sensory and motor neuropathy (of the right / and left median/ ulnar/ common peroneal/ tibial/sural nerves). This matches a

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diagnosis of acute motor sensory axonal neuropathy (AMSAN) variant.

Patient was given intravenous Immunoglobulin (IVIG) (0.4gm/kg/day) for 5 days. Plasmapharesis was initiated at day-21 for 3 consecutive sessions as there was no neurological improvement. His neurological condition did not show any improvement for the following 2 months. He subsequently developed ventilator-associated pneumonia, severe sepsis and died.

This is to illustrate a rare case report of CIDP AMSAN variant of Guillain Barre Syndrome occurring in a patient with Marfan's syndrome and pulmonary tuberculosis.

Discussion

Guillain Barre Syndrome (GBS) is a syndrome manifested by an acute inflammatory polyradiculoneuropathy with resultant weakness and reflex changes. It is considered to be a postinfectious immune mediated disease targeting peripheral nerves. There are different variants based on a clinical spectrum of symptoms and findings. Acute inflammatory demyelinating polyneuropathy (AIDP) is the most widely recognized form in western countries. This patient presented with AMSAN variant of GBS, which is a rapid and severe paralysis associated with delayed and poorer recovery. Patients with AMSAN in general do not respond to IVIG or plasmapheresis. Studies on nerve pathology in these patients show severe axonal degeneration of motor and sensory nerve fibres with little demyelination.

We ran a medline search which revealed no previous reported case of GBS with AMSAN variant occurring in Marfan's syndrome or pulmonary tuberculosis. The mortality rate of GBS ranges from 2-12%. Death usually occurs in ventilator dependent patients due to complications such as pneumonia, sepsis, ARDS or autonomic dysfunction. The differential diagnosis of isoniazid-neuropathy was considered very early on in this patient. However, we consider it highly unlikely as there was no neurological recovery upon drug cessation and there was subsequent development of a bilateral lower motor neuron lesion of the 7th cranial nerve. This left the most likely diagnosis to be GBS which was of AMSAN variant and which progressed to CIDP.

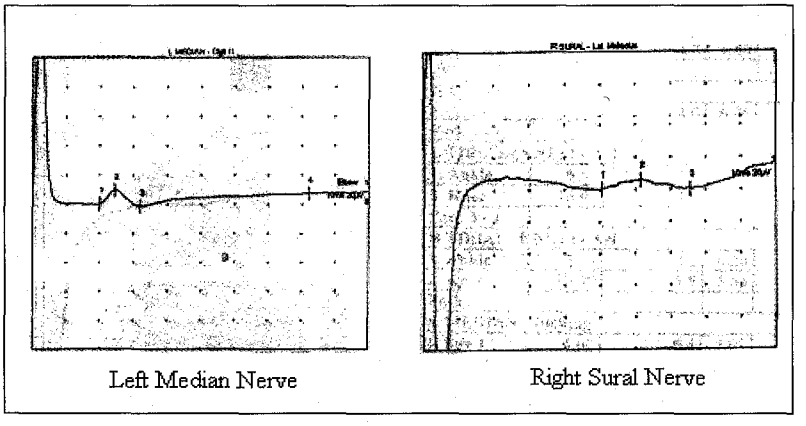

Fig. 1: Nerve Conduction Studies of left median and right sural nerves

Fig. 2: Magnetic resonance imaging (MRI) brain and spinal cord showed no obvious abnormality.

References

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