

Case report:

A 60-year-old man with isaacs mertens-syndrome a rare entity

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Abstract

Isaacs-mertens syndrome also known as neuromyotonia is a rare neuromuscular disorder caused by hyperexcitability and continuous firing of axons of the nerve that innervate the muscle fibers. It is an immune-mediated disorder with elevated antibody level against voltage gated potassium channels (VGKC). Symptoms include progressive muscle weakness, continuously twitching muscles (myokymia), cramping, increased sweating and delayed muscle relaxation. This is a case report of a 60-year-old man who presented with pain and weakness of right lower limb for 3 months, which gradually became worse to involve both the lower and upper limbs. NCS revealed to have motor neuropathy and EMG showed fibrillations and fasciculations with positive sharp waves. After which he was diagnosed with Isaacs-mertens syndrome and was followed up for 8 weeks with medications and physiotherapy management.

Keywords : Isaacs-mertens syndrome; neuromyotonia; muscle twitching; massage and breathing exercise

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Introduction

Isaacs – Mertens Syndrome also referred as acquired neuromyotonia, is an autoimmune disease caused by hyper-excitability of the motor nerves and activation of muscle fibres owing to autoantibodies directed against the VGKCs on peripheral nerves.¹Jacob Isaac Merten first delineated it in 1961.²Clinically it is characterized by continuous, spontaneous, widespread muscle twitching, muscle stiffness, cramps, pseudo myotonia, pseudotetany and at times muscle hypertrophy, mild muscular weakness, parasthesias and numbness that gradually develops between 15 to 60 years.³ The excessive and spontaneous muscle activity persists throughout sleep, attributed to hyperexcitability of terminal parts of motor nerve fiber, possibly as a result of a partial loss of motor innervation and compensatory collateral sprouting of surviving axons measured as high-frequency (up to 300-Hz) repetitive discharges

of varying waveforms in EMG.Differential diagnosis includes hereditary neuromyotonia or myokymia (with/without episodic ataxia) syndromes, cramp-fasciculation syndrome, motor neuron diseases (progressive spinal muscle atrophy, neuropathy, amyotrophic lateral sclerosis (ALS) and tetanus.⁴

Case Description

This case study was carried out in Physiotherapy Out Patient Department, ACS Medical College & Hospital, Vellapanchavadi after the approval of Institution's Review Board

History

A 60-year-old man who was shopkeeper by profession came to our department with complaints of weakness in both the legs and left hand with frequent cramps while walking. The symptoms began 9 months prior to the first day of assessment. Initially he took some Ayurveda medicines for his

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symptoms for 2 months but had no relief; instead, his symptoms began to worsen and became more dependent in his activities of daily living. Initially, doctors thought it as Amyotrophic lateral sclerosis, but electrophysiological studies and immunological studies confirmed the presence of Isaacs-Mertens Syndrome and was under immune-therapy and was then referred to the physiotherapy department for further management.

Examination

While observing, the patient used his right hand to move in the bed and his lower limbs felt to be flaccid. Bilateral triceps, biceps and brachioradialis jerk were diminished and knee and ankle jerk were absent. The muscle power was 4 in his right upper limb, 3 in few muscles and 2 in few muscles of left upper limb, 0 in right lower limb and 1 in left lower limb on manual muscle testing (MMT). There was an altered sensation in the lower limb when compared to upper limb. Chest expansion was normal and single breath count was 38. Barthel index score was 50/100. Visual analogue fatigue scale (VAFS) score was 7. MRI brain, CSF examination was normal. EMG studies suggested denervation of muscles with spontaneous fibrillation and fasciculation, and NCS suggested motor neuropathy. Radioimmunoassay suggested presence of antibodies against VGKC.

Intervention

There is no cure for Isaacs mertens syndrome. The role of physiotherapy in this case was to reduce the symptoms and to maintain the muscle properties, reduce cramp pain and maintain respiratory functions. Active assisted movements to all the joints was given without causing fatigue to the patient once daily for 8 weeks. 10 minutes of massage (effleurage and deep muscle massage) was given on each calf muscle and 3x30 seconds of calf stretching on each leg, with a period of rest of 30 seconds for 2 days/week for 8 weeks to relax the calf muscles and minimize the effects of hypertrophy. Patient was encouraged to perform Breathing exercises for 2 times a day for 8 weeks daily to maintain the respiratory function.

Discussion

Due to the rare occurrence and unknown aetiology of Isaacs Syndrome, the patients have been managed pharmacologically to reduce their symptoms till date. This case report portrays that physiotherapy based

programs may likewise be powerful in maintaining the independence and reducing the impedances associated with muscle impairments in people with Isaac Mertens Syndrome.

The patient in this study presented with continuous muscle twitching with lack of muscle relaxation, triggering cramps and discomfort. After 8 weeks of intervention period there was no change in the muscle power and barthel score but the single breath count increased from 38 to 40 and VAFS reduced from 7 to 5. This finding was similar to the systematic review of *Ahmed A et al*, which stated that strengthening and aerobic exercise programs in subjects with muscular problems has benefits in body function, activities and participation (according to ICF), as respiratory training in patients with myasthenia gravis.⁵

However, only exercise-based programs (strengthening or aerobic) and respiratory component were described in earlier studies, not verifying the effects of passive interventions based on stretching, massage and heat. Researchers have also denoted that massage has a decrease or inhibitory influence on alpha motoneuron excitability by investigating the H-Reflex of muscles.⁶ Using active approaches, like exercise, to reinforce muscle strength will increase the fatigability of the patient⁷, hence passive approaches were chosen in this study to reduce the hyper-excitability of the motor fibres.

Conclusion

Thus, it is concluded from this study that physiotherapy intervention can be used as an adjunct to pharmacological methods in treating patients with Isaacs Mertens syndrome to increase their life expectancy by reducing the symptoms and preventing further complications.

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Authors' contribution:

Data gathering and idea owner of this study: ..G.THARANI

Study design: ...K.KAMATCHI,G.YUVARANI

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Writing and submitting manuscript:G.THARANI

Editing and approval of final draft: ...G.THARANI

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