

Clinical Image

Spontaneous Remission of Isaacs' Syndrome

Uslu et al. Isaacs' Syndrome

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A 18-year-old man presented with generalized body ache, muscle cramps, weight loss and continuous, painful muscle twitching in his lower extremities. His symptoms aggravated with physical exertion. Increased sweating occurred on his limbs and trunk. His medical history was notable for lichen planus which frequently accompanied by autoimmune diseases. He did not smoke or abuse illicit drugs. Family medical history was not significant. On examination, there were fasciculations of his lower extremity muscles (video 1). Consciousness, mental state, tone, power, reflexes, sensory, cerebellar signs and cranial nerve exam were all normal. Laboratory investigations of the patient revealed elevated muscle enzymes: creatine phosphokinase 922 U/L [0–171 U/L], aspartate aminotransferase 85 U/L [0–50 U/L], alanine aminotransferase 157 U/L (0–50 U/L). Serum electrolytes, thyroid hormone, calcium, phosphate, vitamin D were normal. Anti nuclear, antineutrophil cytoplasmic and extractable nuclear antibodies were negative. Muscle magnetic resonance imaging (MRI) showed no evidence of myositis. Cranial MRI was normal.

Normal electrophysiological findings were seen in nerve conduction studies. A decremental response to repetitive nerve stimulation was detected. Needle EMG examination revealed spontaneous motor unit potential transitions and fasciculations in gastrocnemius muscles. Serum anti-voltage-gated potassium channel (VGKC) antibodies were 144 pmol/L (N < 85 pmol/L). Computerized tomography scan of the chest and abdomen did not show any evidence of malignancy. Based on the clinical, electrophysiological findings and antibody positivity, a diagnosis of Isaacs' syndrome was made. While further investigations were in progress, he recovered spontaneously without treatment.

Isaac's syndrome is a rare neuromuscular disorder characterized by hyperexcitability of the peripheral nerves and continuous activation of muscle fibres due to auto-antibodies directed against the VGKCs [1]. If muscle pain and cramps, myokymia, pseudohypertrophy, excessive sweating, delay in muscle relaxation after contraction, elevation of creatine kinase and neuromyotonic discharges are seen in a patient; Isaacs Syndrome should be considered.

Symptomatic treatments (anticonvulsants) or immunosuppressive drugs (oral steroids with or without steroid sparing agents such as azathioprine), plasma exchange or intravenous immunoglobulin infusion can be initiated according to the severity of the disease [2]. And also, spontaneous remission could be observed, finding consistent with autoimmunity [3].

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