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Case Report

A case of post-polio syndrome in a 56 years old male who suffered from a paralytic poliomyelitis at the age of 10 years

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Abstract

Introduction: Polio survivors can develop a new onset or worsening of neuromascular functions several years after regaining neurofunctional stability from acute poliomyelitis, a syndrome known as post-polio syndrome (PPS). Diagnosis of this syndrome is usually based on a typical medical history, distinctive electromyographic findings and exclusion of other similarly presenting illnesses.

Case presentation: We report a case of PPS in a 56-year-old male of African descent who had the initial acute onset of poliomyelitis at the age of 10 years, now presenting with a new onset of muscular weakness and atrophy. He had experienced progressive but partial recovery over the years and had attained fairly stable neuromascular functions until 18 months ago. Physical examination revealed global weakness and atrophy of all four limbs with a general hypotonic tone. Sensory findings and proprioception were normal. There was a hyporeflexia during deep tendon reflex assessment and superficial reflexes were absent. No fasciculation was observed even after provocation in both upper and lower limbs. Serum creatinine kinase was within a normal range (55 mmol/L). Magnetic resonance imaging of the spine and a muscle biopsy ruled out neurological compression and inflammatory myopathies respectively. A nerve conduction study was conclusive for a severe motor axonal neuropathy. Electromyogram showed ongoing denervation bilaterally in the myotomes C6 to T1 and L2 to S1 in the upper and lower limbs respectively which confirmed the diagnosis of PPS.

Conclusions: Polio survivors presenting with a new onset or worsening of neuromuscular functions several years after the acute onset of poliomyelitis should raise an index of suspicion for a diagnosis of post-polio syndrome and should undergo a comprehensive investigation and management.

Key Words: Post-polio syndrome; poliomyelitis; neuromascular weakness; electromyogram; case report

Introduction

Post-polio syndrome (PPS) is characterized by a new or increased muscular fatigue, weakness, pain, and atrophy at least 15 years after regaining neurological and functional stability from acute polio. Depending on the population studied and/or diagnostic criteria used, the prevalence of PPS ranges from 15%-80% and the peak onset is about 30 years after the acute polio onset [1]. Several theories are implicated in the pathophysiology of PPS including; degeneration of the surviving motor nerve cells due to an overexertion phenomenon, reactivation of a persistent latent virus, infection by a different enterovirus from the one initially responsible for polio and neuronal aging [2].

Diagnosis of PPS is usually based on a typical medical history, electromyographic pattern and exclusion of other diseases with similar presentation including multiple sclerosis, amyotrophic lateral sclerosis, and polymyositis. In the management of PPS, pharmacotherapy trials involving; acetylcholinesterase inhibitors, steroids, amantadine, modafinil, lamotrigine, coenzyme Q_2 and intravenous immunoglobulin have shown no therapeutic benefit in improving fatigue, muscular strength or quality of life [1]. On the other hand, physical, occupational and speech therapies have been shown to be safe and effective in reducing further decline of muscle strength and improving symptoms of fatigue, weakness and pain [1, 2]. We report a case of PPS in a 56 years old male of African

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descent who presented with a new onset of muscular weakness and atrophy 46 years after the initial acute onset of poliomyelitis.

Case Presentation

A 56-year-old male of African descent presented to us with progressive weakness and wasting of upper and lower limbs for 18 months. He missed the opportunity for polio vaccination during childhood and had a 46-year history of a weak and atrophic left lower limb secondary to poliomyelitis suffered at the age of 10 years. He had experienced progressive but partial recovery over the years and had attained fairly stable neuromascular functions until 18 months ago. He was diagnosed to have type-2 diabetes mellitus 5 years ago and despite a self-reported good adherence to metiformin and glibenclamide for the entire duration, his blood sugar records show a poor glycemic control pattern.

About 18 months ago, he progressively developed a new onset of muscle weakness in all four limbs with subsequent wasting. The muscular weakness was of gradual onset began in the lower limbs then progressed to involve both upper limbs. The weakness initially affected the proximal group of muscles then progressed to affect the distal ones. He started experiencing difficulties in walking long distances, reaching for far objects, lifting heavy objects, standing from squatting position and climbing followed by descending stairs. He denied any history of trauma or injury, pain, fever, heat or cold intolerance, swelling of the anterior neck, alcohol dependence or familial muscular disease. He has been experiencing intermittent episodes of chest tightness with difficult in coughing and has lost about 11 kilograms during this period. He has a positive history of diabetic complications including poor vision, numbness, tingling and burning sensation of upper and lower limbs, and erectile dysfunction with a reduced sexual desire. Throughout, he has had a normal appetite, micturition and bowel habits, swallowing, and speech. He has been bedridden for the past 8 months.

The patient had stable vitals (BP 122/73 mmHg, HR 81 beats/min, RR 21 breaths/min, temperature 36.9°C and oxygen saturation 97% on room air). Physical examination revealed global weakness and atrophy of all four limbs with a general hypotonic tone. Sensory findings and proprioception were normal. There was a hyporeflexia during deep tendon reflex assessment and superficial reflexes were absent. No fasciculation were observed even after provocation in both upper and lower limbs. All cranial nerves were intact. Eye examination revealed bilateral cataract and a funduscopic examination showed evidence of neovascularization, a feature of proliferative diabetic retinopathy. Musculoskeletal system revealed a winging of scapulae, kyphoscoliosis and a left foot drop. There was a decrease in chest movement bilaterally with the use of accessory muscles during respiration. He had a scaphoid abdomen otherwise normal abdominal examination.

Full blood count, electrolyte panel, VDRL for syphilis and renal, liver and thyroid function tests were normal. His random blood glucose was 16.4 mmol/L and his glycated hemogloblin was 8.9%. Serum creatinine kinase was within a normal range (55 mmol/L). Magnetic resonance imaging (MRI) of the spine and a muscle biopsy ruled out a neurological compression and inflammatory myopathies respectively. Nerve conduction studies was conclusive for a severe motor axonal neuropathy. Electromyogram (EMG) showed ongoing denervation bilaterally in the myotomes C6 to T1 and L2 to S1 in the upper and lower limbs respectively which confirmed the diagnosis of PPS.

Discussion

The incidence of post-polio syndrome is unknown and its etiology remains unclear. Nevertheless, the new onset of muscular weakness negatively affects the daily activities and significantly impairs the overall quality of life of polio survivors. Post-polio syndrome is a diagnosis of exclusion an attribute that makes its diagnosis difficult especially in resource-limited settings where either some investigations are unavailable or patients are unable to pay for certain investigations due to poverty.

Several factors have been associated with an increased risk of PPS including; a significant paralysis during the acute polio, greater functional recovery, higher levels of recent activity, and lower limbs involvement (i.e. doubled risk compared to upper limbs) [3, 4]. EMG studies are crucial in reaching a PPS diagnosis; however, separation of those with PPS versus asymptomatic patients who ever suffered from acute polio is not possible. Furthermore, EMG studies are useful in ruling out other similarly presenting conditions including radiculopathies, neuropathies and myopathies.

Owing to the lack of etiology identification, the treatment of PPS primarily focuses on symptomatic relief. Psychotherapy, physiotherapy and occupational therapy are the mainstay in the management of PPS. Patients might present with reduced respiratory functions as a result of weak respiratory muscles and/or chest deformities [5]. Moreover, bulbar symptoms including; dysphagia, voice weakening and vocal changes have also been reported [6].

Conclusions

In conclusion, polio survivors presenting with a new onset or worsening of neuromuscular functions several years after the acute onset of poliomyelitis should raise an index of suspicion for a diagnosis of post-polio syndrome and should undergo a comprehensive investigation and management.

Consent

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Abbreviations

EMG: electromyography;

MRI: magnetic resonance imaging;

PPS: post polio syndrome;

VDRL: venereal disease research laboratory.

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