Intravenous Immunoglobulin Therapy in Postpoliomyelitis Syndrome

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Abstract

Postpoliomyelitis syndrome is a new neurological disorder seen in individuals who survive remote poliomyelitis. An adequate diagnosis is important for treatment and prognosis. The diagnosis of postpoliomyelitis syndrome might be difficult as the diagnosis mostly depends on the patient's subjective description of the symptoms. The diagnostic criteria primarily depend on new and progressive muscle weakness, muscle fatigue, general fatigue, and pain in the absence of other disorders which may be similar to this syndrome. Furthermore, the postpoliomyelitis syndrome patients have reached an age when concomitant diseases are common. The possible etiology is that this entity of paralysis can be exploited by slowly progressive distal neuronal degeneration. Interestingly, this patient had primary upper limb polio paralysis and then new slowly progressive asymmetrical, muscle weakness in the contralateral lower limb. Few clinical trials using human intravenous immunoglobulin (IVIG) and pyridostigmine gave controversy about a specific treatment. In this case, we tried an IVIG therapy, but the weakness was progressively worsening and there was more atrophy in the muscle bulk of the affected muscle group. However, supportive treatment is also important to be planned. The aim of this case report is to describe the complexity of the diagnosis and treatment of such cases of the postpoliomyelitis syndrome in comparison to relevant literature.

Keywords: Intravenous immunoglobulin in postpoliomyelitis, postpoliomyelitis syndrome, weakness

Introduction

Postpoliomyelitis syndrome is a neuromuscular syndrome, usually characterized by new insidious symptoms of increased muscle weakness, pain, and fatigue at least 15 years following the paralysis of poliomyelitis, with a mean interval roughly around 35 years.[1-3] Postpoliomyelitis syndrome might be difficult to diagnose depending on the patient's own subjectivity since the complexity of the postpoliomyelitis syndrome necessitates taking into consideration and excluding other concomitant disorders such as disc herniation, diabetes mellitus, trauma, and other neuropathic disorders.

The goal of this presentation was to show the complexity of the postpoliomyelitis syndrome diagnosis in exclusion of concomitant disorders and to report that the weakness could affect the lower limb in upper limb-prior polio patient and to use intravenous immunoglobulin (IVIG) to minimize neuronal degeneration.

Case Report

A 45-year-old male from Iraqi Kurdistan, a hospital-laboratory clerk, had polio at the age of 2 years, affecting the right upper limb. On April 2018, he noted to develop new insidious weakness of the left lower limb over 3 months with no sensory symptoms, also associated with asymmetry of the bulk of the thigh muscle. There was no history of trauma or injury or lumbar pain. His right lower limb was not affected. Physical examination revealed atrophy of his left pelvic and thigh muscle. There was fasciculation in the quadriceps and hamstring muscles. The tone was generally hypotonic. Further examination revealed the weakness of Medical Research Council (MRC) grade four out of five. The sensory function to pinprick sensation was intact. The examination of deep tendon reflexes disclosed reduced responses in the right upper limb and the left lower limb. Electromyography (EMG) revealed ongoing denervation neurogenic units in the left lumbosacral roots. Magnetic resonance imaging (MRI) of the lumbosacral spines did not reveal any neurological compression to the spinal cord or to the nerve roots, but MRI of the pelvis

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shows atrophy of the left pelvic muscle group. There was no medical history of diabetes mellitus, thyroid disease, or alcohol dependence. Blood investigations of free thyroxine level, thyroid-stimulating hormone, venereal disease research laboratory, full blood count, and serum electrolytes were normal.

Discussion

A careful evaluation to diagnose postpoliomyelitis syndrome is mandatory since other concomitant disorders may mimic postpoliomyelitis syndrome. Hence, correct analysis of clinical features and then relevant investigatory tools were applied in this patient to apply a proper treatment. The United States National Institute of Neurological Disorders and Stroke lists the plausible criteria for the diagnosis of postpoliomyelitis syndrome as shown in Table 1.[4]

Patients with polio weakness in their legs were twice as likely to complain of the postpoliomyelitis syndrome compared to patients with weakness in their arms. The postpoliomyelitis syndrome can occur in muscles previously affected or clinically unaffected during the acute attack but is more likely to occur in the muscles that were originally affected. Interestingly, this patient had upper limb polio paralysis and then new slowly progressive asymmetrical, muscle weakness in the contralateral lower limb.[3,5]

Certain risk factors for the development of the postpoliomyelitis syndrome have been identified, including the degree of severity of acute polio paralysis, advanced age of onset of acute polio paralysis, the number of permanent deficits present after recovery, female gender, recent weight gain, and a greater amount of physical activity during stability.[6]

The exact etiology is unknown. Many theories have been proposed, including immunopathogenic mechanisms, neuronal aging, and viral reactivation, but the most accepted hypothesis would be an excessive metabolism demand on surviving giant-sized motor units, which in turn would be brought on by equally excessive use of muscles and then excessive metabolic stress on the remaining prematurely degenerated neurons, thus resulting in a reduction in axonal "sprouting" in muscle fibers.[7]

Table 1: Diagnostic criteria for Postpoliomyelitis syndrome

NINDS criteria for postpoliomyelitis syndrome
1. A history of paralytic poliomyelitis as confirmed by history, neurologic examination, and electromyography
2. A period of partial or complete functional recovery, followed by a stable period of at least 15 years
3. The gradual or sudden onset of progressive and persistent new muscle weakness or decreased endurance, with or without generalized fatigue, muscle atrophy, or muscle and joint pain
4. The symptoms should persist for at least 1 year
5. Exclusion of other causes

NINDS: National Institute of Neurological Disorders and Stroke

Electromyogram studies indicate that >50% of the muscle's anterior horn cell population death must occur before clinical weakness can be detected.[3,8]

The evidence of poliovirus in cerebrospinal fluid in patients with the postpoliomyelitis syndrome has been encountered. Sharief *et al*.'s study show the presence of only poliovirus immunoglobulin M (IgM) antibodies and they failed to find poliovirus in those patients.[9] Although the cerebrospinal fluid analysis for this patient was normal, unfortunately, the laboratory tool for poliovirus IgM is not available in our hospital.

Apart from physiotherapy and analgesia, up to date, there is no specific medical treatment for the postpoliomyelitis syndrome, even though few clinical trials used human IVIG.[10] Other trials which used pyridostigmine have been conducted, although the controversy was the dominated issue.[11]

Farbu *et al*.'s pilot study suggested that IVIG could be a therapeutic option. Patients receiving an IVIG reported a significant improvement in pain during the first 3 months, but no change was noted for subjective fatigue and muscle strength.[12]

There is a controversy over the appropriate dose of IVIG and therapeutic interval.[13] In this case we tried to show the beneficial therapeutic effect of human IVIG in a dose of 2 g/kg of body weight (total dose of 130 g divided over 5 consecutive days). The results noted 3 months later that muscle atrophy was progressively worsening in the affected muscles, and the strength of the muscle was Grade 3 MRC scale. Furthermore, EMG finding was approximately the same finding as it was shown before IVIG treatment. The only positive effect experienced by the patient was no more complaint of pain in his left lower limb. Hence, unfortunately, this line of immunotherapy is not promising. Similarly, one of a meta-analysis study stated that IVIG therapy to patients with postpolio syndrome (PPS) was unlikely to produce significant improvements in pain, fatigue, or muscle strength. Thus, based on randomized controlled trials, routinely administering IVIG to patients with PPS is not recommended as a standard treatment in PPS.[13,14]

Conclusion

Although there is no specific therapy, IVIG failed to give hope to treat the postpoliomyelitis syndrome. Hence, the goal of treatment is directed to physiotherapy, reassurance, and modification of lifestyle. In this case, the primary polio paralysis affected the upper limb, but the weakness of the postpoliomyelitis syndrome affected the lower limb.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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