Case Report

# Post-poliomyelitis Syndrome: Case Report and Review of the Literature

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## Abstract

**Introduction: Postpoliomyelitis syndrome (PPS) refers to the new neuromuscular symptoms that occur in patients years after their acute poliomyelitis has stabilised. PPS cases seen now are probably related to the poliomyelitis epidemics of the 1940s and 1950s. Clinical Picture and Investigation: A 57-year-old Chinese man with a history of poliomyelitis affecting both lower limbs presented with left upper limb weakness. Physical examination revealed atrophy of his left upper limb muscles. There were fasciculations in the biceps and brachioradialis muscles. Electromyography revealed ongoing denervation neurogenic units in the C5 to TI myotomes comparable with PPS. Conclusion: The aim of this paper is to review the present situation and to give a short summary of PPS, which can be difficult to diagnose because the symptoms of presentation are usually non-specific. We describe a case of PPS and review the current literature.**


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**Key words: New neuromuscular symptoms, Progressive postpoliomyelitis muscular atrophy**

## Introduction

Postpoliomyelitis syndrome (PPS) is characterised by the delayed appearance of new neuromuscular symptoms in patients many years after their acute poliomyelitis paralysis. PPS occurs 30 to 40 years after an acute poliomyelitis attack and is observed in approximately 25% to 28% of patients.[1] It is still unclear, at this point in time, if the occurrence of PPS increases with age. The PPS cases seen are probably related to the poliomyelitis epidemics of the 1940s and 1950s.[2]

## Case Report

A 57-year-old Chinese man with bilateral lower limb amyotrophy attributed to poliomyelitis sequelae presented with left upper limb weakness, which had first occurred 6 months ago. He had a history of poliomyelitis which affected both his lower limbs when he was 4 years old. He had contracted the disease in 1951, during the last epidemic outbreak in Singapore, just before the introduction of the polio vaccine. He had experienced progressive but partial recovery over time. During that acute attack, he had not felt any conspicuous weakness in his upper limbs. He had been well until 6 months ago, when he complained of progressive left upper limb weakness, with no sensory impairment. There was no associated trauma or injury to the upper limb or neck. His right upper limb was not affected. Physical examination revealed atrophy of his left upper limb muscles. There were fasciculations in the biceps and brachioradialis muscles. The tone was generally hypotonic. Further examination revealed weakness from myotome C6 to TI. Sensitivity to soft touch, temperature and pain was intact. Deep tendon reflexes were hyporeflexia in bilateral lower limbs and left upper limb. Electromyography revealed ongoing denervation neurogenic units in the C5 to TI myotomes comparable with PPS. Magnetic resonance imaging (MRI) of the cervical spine did not reveal any neurological compression of the cord or nerve roots. 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 (VDRL), full blood count and serum electrolytes were normal.

## Discussion

The current diagnostic criteria for PPS were first described by Mulder et al in 1972.[3] These criteria are 1) a prior

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episode of poliomyelitis with residual motor neuron loss; 2) a period of at least 15 years of neurological and functional stability after recovery from the acute illness; 3) the gradual or, rarely, abrupt onset of new weakness or abnormal muscle fatigue, and generalised fatigue; and 4) the exclusion of other conditions that could cause similar manifestations. When no alternative explanation can be found, this late-onset weakness is referred to as PPS. These new symptoms may lead to problems in activities of daily living such as walking, climbing stairs and dressing.[4]

The risks for the development of PPS were noted in patients who presented with significant paralysis of that limb at the time of acute illness. Patients with weakness in their legs were twice as likely to complain of new problems compared to patients with weakness in their arms. Interestingly, patients' presenting age and time duration since their poliomyelitis attack were not factors in determining their current PPS complaints. Elevated serum creatine kinase levels were present only in those who presented with new complaints.[5] Klingman et al[6] noted that patients who developed PPS had a history of more widespread paralysis during the acute illness, but they also had greater functional recovery. They were also less disabled, and reported higher levels of recent activity. It was felt that the degree of functional recovery might be an even more important predictor of PPS than the others.

New muscle weakness is the most significant neurological problem.[7] This new muscle weakness usually progresses at a slow rate, is asymmetrical, and can be of a proximal, distal or patchy distribution. It 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. More than 50% of the muscle's anterior horn cell population death must occur, before clinical weakness can be detected. EMG studies indicate that many of the clinically unaffected muscles were involved subclinically during the acute episode of disease presentation.

In addition to generalised weakness in the body, leg and arm, it may also present as respiratory insufficiency, bulbar muscular dysfunction (dysphagia, dysarthria and aphonia) and sleep apnoea. Respiratory problems are present in a minority, who will require special attention and intervention.

The aetiology and mechanism of PPS is still unknown, causing much anxiety among poliomyelitis survivors. The most established hypothesis at the moment is an excessive metabolic stress on the remaining motor neurons over many years, resulting in the attrition and premature degeneration of surviving motor neurons with the loss of axonal terminals.[8] This theory is supported by EMG and muscle biopsy studies. Another hypothesis concerns the persistence of poliovirus in the central nervous system. Sharief et al[9] demonstrated the presence of poliovirus immunoglobin M (IgM) antibodies and poliovirus-sensitised cells in the cerebrospinal fluid (CSF) of PPS patients. Conclusive evidence of the poliovirus from the PPS patient's CSF has, however, not been found.

Blood test results are usually normal except for occasionally raised serum creatine kinase levels in patients, which may suggest that muscle fibres are damaged, and is an indication of muscle overuse.[10,11] EMG studies can identify changes consistent with previous poliomyelitis, but cannot separate those with PPS from asymptomatic patients with previous poliomyelitis. EMG and nerve conduction studies are, however, useful for identifying and excluding other diseases (radiculopathy, neuropathy and myopathy). Imaging studies (e.g., computed tomography, MRI) may be needed to exclude spinal conditions, such as spondylosis, spinal stenosis and neoplasms.

Due to the lack of definite aetiology, treatment is currently focused on symptomatic relief, including reassurance of limited neurological impairment. Medications, most of which address fatigue, have been used with limited success in PPS patients. Proper rehabilitation programmes to maintain the patient's independent functional status and regular exercises which do not place excessive stress on the joints are the mainstay of treatment. The perceived fatigue and decreased endurance found in patients with PPS has no clear explanation. Generalised fatigue is best treated with lifestyle adjustments, including reduced physical activities, a weight loss programme, and the use of technical devices for mobility (walking sticks, wheel chair, orthoses). Non-fatiguing exercise has been found to be useful for patients with muscular weakness caused by PPS.[12-14] Excessive conditioning exercises may lead to further loss of function, instead of improvement. Studies on non-fatiguing exercises have demonstrated increased muscle strength and none have shown significant laboratory evidence of muscle overuse (e.g., increase in serum creatine kinase level, EMG evidence of muscle injury or biopsy revealing evidence of muscle damage). These carefully paced exercise programmes emphasise the prevention of muscle overuse. In order for muscle recovery to take place, the patient should rest in between bouts of short interval exercises.

## Conclusion

PPS is difficult to diagnose since the symptoms of presentation are usually non-specific. Diagnosis is made mainly from the patient's history coupled with a high index of suspicion from the physicians. Although the cause of the disease is still unknown, PPS is likely due to the premature degeneration of surviving motor neurons. There is no specific therapy. Therapeutic approaches are symptomatic and include physiotherapy exercises, reassurance to the

patient regarding their condition and modification of lifestyle to combat fatigue.

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