Post poliomyelitis syndrome: A rare sequel of acute poliomyelitis

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Abstract

Post poliomyelitis syndrome (PPS) is a rare sequel of acute poliomyelitis, usually seen 30-40 years after an acute episode. It is characterized by new muscle weakness seen in survivors of acute poliomyelitis. We describe a rare case of a 50 year old man; with a previous history of poliomyelitis in right lower limb who now presented with complaints of progressive left lower limb weakness for past two years. The diagnosis was made on the basis of clinical suspicion and EMG findings. PPS is not a well recognized disease in Pakistan and due to the lack of documentation; its true prevalence is not known. Though, over the years, cases of Poliomyelitis have decreased worldwide, however, PPS still remains a constant challenge for the physicians. This report highlights the impact of the disease on the quality of life of patients suffering from PPS and emphasis on the need for new therapeutic approach.

Keywords: Post poliomyelitis syndrome, Muscle weakness, Pakistan.

Introduction

PPS is a delayed development of neuromuscular syndrome in patients with a history of acute poliomyelitis.[1] PPS usually occurs 30-40 years after an acute poliomyelitis attack.[2] With the advent of immunization, cases of acute anterior poliomyelitis (AAP) have drastically declined worldwide. However, PPS is still a constant threat to about 25 - 28% of patients who have already suffered from acute poliomyelitis attack in the past.[3]

PPS is a slowly progressive disease that leads to muscle deterioration from 7% in four years to 15% in eight years. Acute Poliomyelitis may have been eradicated from the Western world but the fact that it still persists in the developing countries means that PPS will tend to be a challenge for physicians for decades.[4] There is limited data on PPS and a Pub-med search did not reveal any previous case of PPS published from Pakistan.

Case Report

We report the case of a 50 years old man, with no known co-morbids. He was normal at birth but by the age of 6, he developed right sided lower limb weakness, which led to the diagnosis of Acute Poliomyelitis. He was started on rehabilitation and had a gradual recovery with residual deformity in his right foot. He has had a healthy active life since and was able to walk with "slight unnoticeable limp". For about four decades he had been clinically stable. However, at the age of 48, he developed left sided lower limb weakness, generalized aches and pains and muscle cramps. These symptoms have been progressively worsening for past 2 years. Initially he only experienced weakness while climbing stairs; however, now he complained of difficulty on walking. These complaints had limited his day to day activity. He had no history of trauma or injury prior to this weakness. His other limbs were unaffected and there was no sensory impairment. There was no medical history of diabetes mellitus, thyroid disease, and use of statins or alcohol dependence.

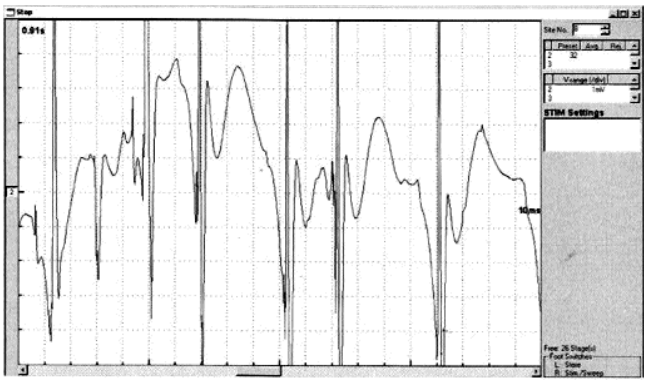

Figure: EMG of left sided Quadriceps muscle showing giant motor unit potential.

On examination, he had left quadriceps atrophy with compensated left calf hypertrophy. His right thigh was mildly atrophic with right calf hypertrophy and foot deformity, which according to the patient has been static since his childhood. Fasciculations were seen in the left quadriceps muscles. Superficial touch, pain and temperature sensations were normal. Deep tendon reflexes of lower limbs were diminished bilaterally.

Haematological investigations were all within normal range except raised levels of serum creatine kinase (807 U/L). MRI spine did not reveal any significant neurological

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Table-1: EMG shows abnormal study suggestive of moderate anterior horn cell disease affecting the lower limbs more than the upper limbs. In the clinical context of a remote post polio deformity of right leg, these findings could be due to post-polio syndrome.

Muscle Tested	Insertional Activity	Spontaneous Activity			Motor Unit			Recruitment	Interference
	↑↓	Fibs	PSW	Misc	Amp	Dur	Poly		
(L) Tibialis Anterior	-	1+	1+	Fasic	↑↑	↑	0	Mod-Severe ↓	Mod-Severe ↓
(L) Gastrocenemius	-	0	0	Fasic	↑↑	↑	0	Mod-Severe ↓	Mod-Severe ↓
(L) Vastus Lateralis	-	1+	0	0	N	↑	0	Mod-Severe ↓	Mod-Severe ↓
(L)Quadriceps Femoris	-	0	0	0	↑↑	↑	0	Mod-Severe ↓	Mod-Severe ↓
(L) Abductor Pollicis Brevis	-	0	1+	Fasic	N	N	0	N	Full
(L) Flexor Digitorum Indicis	-	0	0	0	N	N	0	N	Full
(L) Biceps	-	0	0	0	N/↑	N	0	N	Full
(L) Triceps	-	0	0	0	N/↑	N	0	N	Full
(L) Gluteus Medius	-	0	0	0	↑↑	↑	0	Mod-Severe ↓	Mod-Severe ↓
(L) Lumbar P/S (L)	-	0	1+	0					
(L) Lumbar P/S (M)	-	0	1+	0					
(L) Lumbar P/S (U)	-	1+	0	0					
(R) Lumbar P/S (L)	-	0	1+	0					
(R) Lumbar P/S (M)	-	0	1+	0					
(R) Lumbar P/S (U)	-	0	1+	0					

Table-2: Diagnostic Criteria of PPS.

Diagnostic Criteria
1. Previous history of poliomyelitis.
2. Partial or complete recovery after acute episode.
3. At least 15 years of clinical stability after recovery from acute illness.
4. Abrupt or gradual onset of new weakness, muscle fatigue or generalized fatigue.
5. Exclusion of other conditions with similar manifestations.

compression of the cord or nerve roots. Nerve Conduction Studies (NCS) were within normal limits. Electromyography (EMG) showed active denervation in L2 to L4 myotomes and fasciculations (Figure-1), comparable with PPS (Table-1).

The patient was counseled regarding the nature, clinical course and prognosis of the disease. He was advised rest and life style modifications including avoidance of excessive exertion, healthy diet and cessation of cigarette smoking. He was started on low intensity exercises and he was prescribed Capsule Vitamin E 400 mg OD, Tablet Amantidine 50 mg BD, Co-enzyme Q-10 50 mg BD and Tablet Acyclofenic Na 100 mg TDS PRN for pain. The patient has not come for follow up so far.

Discussion

PPS is difficult to diagnosis since the symptoms of presentation are usually vague and non specific. It is the diagnosis of exclusion, made mainly from patient's history and high index of suspicion from the physician. Table-2 shows the current diagnostic criteria of PPS proposed by Halstead in 1985, which were used for diagnosis in our patient.[5]

The exact cause and mechanism of PPS is still unknown. However, many theories have been proposed.[6] According to the most established hypothesis, the excessive metabolic stress on the remaining motor neurons over a period of time, results in their early degeneration and damage to axonal terminals.[7] This theory is supported by electromyography and muscle biopsy studies.

Haematological investigations in patients with PPS are usually normal, except for raised serum creatine kinase levels. Increase in creatine kinase level is due to the damage to muscle fibers and overuse of muscles and indicates an acute illness.[8] Electromyography (EMG) is a very useful investigation in patients with PPS. Ongoing denervation and fasciculations seen on EMG (Figure-1), similar to that present in our patient, indicates an acute disease process. It can also help to identify and exclude other conditions like radiculopathy, neuropathy and myopathy.[9] However, it cannot alone differentiate between PPS and asymptomatic patients with previous poliomyelitis. Imaging techniques can also be helpful to exclude other spinal conditions like spinal stenosis, neoplasm and spondylosis.

Several risk factors have been identified for PPS. Female gender, weight gain, severity of the disease at onset and greater amount of physical activity has been associated with PPS. Aging process, with gradual loss of neurons after age of sixty, may be another contributing factor.[10] It is postulated that life style modifications such as healthy diet, exercise, adequate sleep and weight control could help delay the progression of the disease but no study has proven this hypothesis so far. According to a survey

conducted in 2014, there was a significant increase in orthopaedic symptoms (p <0.001) and loss of functionality in upper limbs (p= 0.004) seen in patients suffering from PPS.[11]

Various trials have been conducted to study the role of pharmacological interventions on patients with PPS, but have found to be of no added advantage.[12] According to one randomized double blinded clinical trial, those patients with PPS who received intravenous immunoglobulin (IV-IgG) had increase in muscle strength compared to the control group. However, the quality of life did not vary between the two groups.[13] Likewise another trial has been conducted on the use of pyridostigmine in improving quality of life of patients with PPS. However no significant result has yet been found.[14] A case control study was done in 2005 to study the effects of lamotrigine in patients with PPS, was found to be ineffective in providing symptomatic relief.[15] It had been widely believed amongst physicians that Co-enzyme Q10 supplements would help muscle functions in patients with PPS, but trials so far have been negative.[16]

There is no specific therapy for the disease. Therapeutic approaches are symptomatic; usually include physiotherapy, excessive reassurance and life style modifications. The goal of rehabilitation in patients with PPS is to optimize function according to their disability. The key to the treatment is to maintain an equilibrium between activity and rest and hence, prevent excess muscle use and the deterioration that comes with it.[17]

Conclusion

PPS is a rare neurological disorder seen as a delayed sequel of acute poliomyelitis. Diagnosis requires the presence of certain clinical criteria's and to rule out similar conditions. The exact mechanism and nature of the disease is yet unknown and hence further studies need to be conducted in order to observe the behavior of neurons years after an acute poliomyelitis attack. Any new weakness in a patient previously affected from poliomyelitis, should raise the suspicion of PPS and early visit to a neurologist along with EMG findings can enable early diagnosis. So far, there are no treatment options that could reverse the effects of PPS. Current treatment options only focus on managing the symptoms of the patients suffering from PPS. Once, the mechanism of the disease is established, ways to prevent the development of PPS could be determined and better treatment techniques could be introduced. PPS is a constant threat to Poliomyelitis survivors and hence further research should be done to fight this threat.

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