

POST-POLIOMYELITIS SYNDROME

Case report

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ABSTRACT - The post-polio syndrome (PPS) is an entity characterized for an episode of muscular weakness and/or abnormal muscular fatigue in individuals that had presented acute polio years before. We report the case of PPS in a patient, 40 years, that thirty-five years after had had paralytic poliomyelitis, developed new symptoms of fatigue, muscular atrophy, dyspnea, difficulties in deambulation and muscular and joint pain. The electromyographic findings revealed injured neurons of the anterior horn of the marrow and reinnervation after muscular tests.

KEY WORDS: post-poliomyelitis syndrome, paralytic poliomyelitis, neuromuscular diseases.

Síndrome pós-polio: relato de caso

RESUMO - A síndrome pós-polio (SPP) é entidade caracterizada por um episódio de fraqueza muscular e/ou fadiga muscular anormal em indivíduos que apresentaram poliomielite aguda anos antes. Relatamos o caso de SPP em um paciente, 40 anos, que trinta e cinco anos após haver tido poliomielite paralítica, desenvolveu novos sintomas de fadiga, atrofia muscular, dispnéia, dificuldades na deambulação e dores articulares e musculares. Os achados eletromiográficos revelaram acometimento dos neurônios da ponta anterior da medula e reinervação após a realização de testes musculares.

PALAVRAS-CHAVE: síndrome pós-polio, poliomielite paralítica, doenças neuromusculares.

Several patients who presented with paralytical poliomyelitis developed, after many years of neurologic and functional stability, a progressive worsening of clinical symptoms^{1,2}, thus constituting a new syndrome comprising fatigue, muscular atrophy, and pain. It is called "post polio syndrome" (PPS)^{3,4}. Acute anterior poliomyelitis (AAP) is a viral disease⁵, which is characterized by headache, fever, pharyngitis, as well as signs of advanced meningeal irritation, followed by a lower motor neuron syndrome located to the spinal cord and consisting of an assymetric flaccid palsy of spinal muscles, and predominantly in the lower limbs^{6,7}. Following the advent of immunization, the overall incidence of AAP declined drastically worldwide, and a portion of patients who had previously developed polio during the 1940's and 50's are now currently presenting with the delayed, tardive effects of poliomyelitis¹. These delayed clinical

signs is probably related to the excess of physical activity during the phase of relative stability⁷.

CASE

A 40 years man, tax auditor, reported that at five years of age presented with clinical picture of AAP that subsequently resulted in upper right limb and lower left limb paresis, difficulty walking and in the carrying out of some basic daily activities. Normally returned to his activities and later enrolled in a regular exercise program. After three decades of clinical stability, he began referring weakness, muscle aches, and cramps in muscle groups not previously affected during his first bout of polio. In August of 2005, was diagnosed with PPS. The neurological examination showed assymetrical paresis in upper and lower limb as well as absent tendon reflex and except the flexor muscles of the hand. Myofasciculations and general twitching were also noted in the scapular region bilaterally, as well as in the braquial biceps, braquial triceps and quadriceps muscle groups. Figures 1 and 2 show the patient with marked right

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Fig 1. Marked right deltoid atrophy.



Fig 2. Atrophy in the left lower limb muscles.

Table 1. Degree of paresis in diverse muscle groups (MRC-upper limbs)

Muscles	Left side	Right side
C5 – Elbow flexors	5	4
C6 – Wrist extensors	5	5
C7 – Elbow extensors	3	2
C8 – Fingers flexors	5	5
T1 – Finger abductors	4	4

Table 2. Degree of paresis in diverse muscle groups (MRC-lower limbs).

Muscles	Left side	Right side
L2 – Hip flexors	3	4
L3 – Knee extensors	2	4
L4 – Ankle dorsiflexors	0	5
L5 – Long toe extensors	0	5
S1 – Ankle plantar flexors	0	5

deltoid atrophy, and in practically all of the left lower limb muscle groupings, respectively. Tables 1 and 2 show various degrees of paresis in diverse muscle groups⁹. An electroneuromyogram revealed decreased insertional activity in proximal muscles of the upper limbs as well as of the right scapular group. The recruitment pattern observed in affected muscles was of the incomplete type, thus indicating chronic reinnervation (old lesion). A magnetic resonance imaging (MRI) of the cervical spinal cord showed spinal (medullary) atrophy in the presence of a left postero lateral herniated disk at the C6/C7 level (Fig 3).

DISCUSSION

PPS is defined as the delayed development of a neuromuscular syndrome, at least 15 years following the stabilization of a previous bout of clinically evident poliomyelitis⁴. The mean interval between AAP and the first signs and symptoms of PPS is roughly around 35

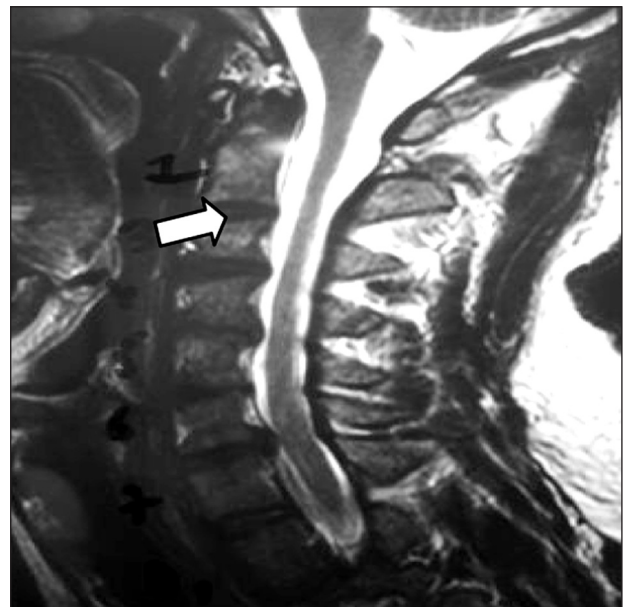


Fig 3. MRI (T2) of cervical cord: cervical spinal atrophy (arrow).

years⁹. It is a slowly progressive disease², usually with an insidious, subacute onset, and resulting in sometimes important restriction of daily activities¹⁰.

The precise etiology is unknown, although innumerable theories have been proposed, including immunopathogenic mechanisms, neuronal aging, and viral reactivation¹¹. The most plausible hypothesis would be excessive metabolism demand on surviving, giant-sized motor units, which in turn would be brought on by equally excessive use of muscles over the years, thus resulting in a reduction in axonal "sprouting" in muscle fibers^{12,13}. The giant-sized motor units would develop during the AAP phase², with the goal of maximum reinnervation of previously denervated muscle fibers in order to maintain adequate function⁷. Although this process is clinically efficient, neuronal overload ensues⁴.

The process of distal wasting of motor units produces gaps in the neuromuscular junction, this probably being the main cause of fatigue, while muscle fibre denervation causes the weakness "per se" encountered in PPS. Certain risk factors for the development of PPS have been identified and include the degree of severity of AAP, advanced age of onset of AAP, number of permanent deficits present after recovery, female gender, recent weight gain, and a greater amount of physical activity during stability¹⁴. Patients who develop PPS have a history of severe, extensive paralysis at the time of the original bout of poliomyelitis¹⁴. The clinical manifestations most commonly found are muscular weakness, fatigue and pain^{3,10}. Fatigue is the most incapacitating symptom, occurring in almost 90% of patients^{9,15}. It is defined as a profound exhaustion that worsens with minimal physical activity. Patients with fatigue usually feel best during morning hours, growing worse as the day progresses². Fatigue has been characterized as a decrease in the tolerability as well as resistance to exercise, which improves upon rest^{2,9}. The new-onset weakness, which may be accompanied by atrophy, mostly affects muscles previously affected by AAP, sometimes even extending to muscle groups unaffected by the original disease^{9,11}. It is usually progressive and asymmetrical, proximal or distal^{2,11}. Other signs that signal muscle involvement include: fasciculations, cramps, and high serum creatine phosphate levels⁷.

Pain is a common complaint, and is usually the one that dominates the clinical picture and can be of either muscular or articular origin². Pain is also associated with excessive activity¹⁵, more commonly in the lower limbs and back in walking patients and in the upper limbs in wheelchair-bound or crutch wielding patients¹⁶. Patients referring pain usually limit their day-to-day activities, which can lead to disuse atrophy¹¹. Other clinical symptoms less commonly found include respiratory insufficiency or breathing difficulty in general, sleep disturbances, intolerance to cold, dysarthria, dysphagia and joint deformities^{5,9}.

The principal diagnostic criteria are: proven past history of poliomyelitis, a period of complete or partial clinical recovery followed by a period of clinical stability of at least 15 years, gradual or rarely sudden onset muscular weakness or abnormal fatigue; and finally, the exclusion of other neurologic and orthopedic causes¹⁷. Electroneuromyographic studies are useful for identifying previous neuronal loss due to AAP^{2,5}, and also to exclude other neurologic diseases, though it does not distinguish PPS from other,

asymptomatic patients that present with subclinical (AAP) polio¹³.

The goal of treatment is to provide the patient with the principles and means of self-sufficiency and to effect change in lifestyle, so as to reduce excessive metabolic demands on muscle⁷, and should include methods for energy conservation, regular periods of rest, weight loss, use of orthoses, and exercise in moderate amounts^{2,7}. The key to treatment is to discover an equilibrium between activity and rest, preventing excessive muscle use and the subsequent deterioration that comes with it. The physical limitations caused by the recent appearance of new motor symptoms added to the restrictions caused previously by AAP lead to a drastic reduction in day to day activity⁷, as well as to psychological repercussions caused by an unexpected and acute disability². Many patients, like the one included in our study, received motivation to practice exercise and to make use of extreme compensatory methods during years in order to maintain their daily functions. These compensation methods consisted of utilizing muscles to their maximum capacity, a greater caloric consumption for the realization of baseline activities, and the use of ligaments for stability which used to lead to hypermobility. However, these compensations were the cause of microtraumas and microcontusions of joints and ligaments, leading to overuse and ultimate exhaustion of neuromuscular motor units in an already constrained system⁷.

Patients who lose half (50%) of their motor neurons are still able to maintain clinically normal muscular activity/function¹⁸, therefore subclinical involvement of certain muscle groups may be present. Many patients may concentrate excess weight on a limb up until then thought to be unaffected by disease, thus new onset weakness can involve apparently unaffected limbs⁷.

The aging process, with gradual loss of neurons mainly after age 60, may be a contributing factor to PPS¹⁹. The superposition of this process in an already constrained motor system of limited motor units will result in a significant decrease of force¹³. However, this process does not fully explain the clinical aspects found in patients under 50 years of age²⁰. The signs and symptoms of PPS, though slowly progressive, can lead to changes in functional capacity and thus modify lifestyles. Despite the fact that there is no specific treatment available, rehabilitation programs may bring positive results for patients. Possible theories should be considered as probable triggers of PPS, so

that benefits may bring upon by such activities may be potentialized and that the deleterious effects of excessive use can be avoided²⁰.

Despite the fact that there is no specific medical treatment for PPS, clinical trials using human intravenous immunoglobulin and pyridostigmine have been conducted²¹⁻²³.

Kaponides et al. obtained impressive results while evaluating possible changes in motor power, physical performance and overall quality of life in patients diagnosed with PPS. Of a total of 14 patients studied, all related improvement in quality of life, however without noticeable improvement in motor power nor physical performance. The results suggest that intravenous immunoglobulin may produce substantial clinical effects resulting in improvement in overall life quality. Such an effect may be the result of a reduction in the inflammatory process of the central nervous system. Further controlled studies as well as random ones are needed, however, since the placebo effect may have influenced the outcome.²¹

Gonzales et al. conducted a randomized, double-blind clinical study in patients treated with intravenous immunoglobulin for PPS. A number of 142 patients were selected from four teaching hospitals in Stockholm, Sweden. The patients were divided into two groups. The experimental group, consisting of 73 patients, received intravenous immunoglobulin during 3 consecutive days, repeated after three months. The control group, consisting of 69 patients, received a placebo. A questionnaire with the objective of evaluating life quality was added to an evaluation of motor power. The results indicated that patients who received intravenous immunoglobulin had an increase in muscle strength when compared to the control group. Life quality did not vary between the two groups. The authors believe that the immunoglobulin may be an option to supportive treatment for patients with PPS.²²

Trojan et al. conducted a double-blind, randomized and controlled study with the object of evaluating the effects of pyridostigmine on life quality, motor power, fatigue, and blood levels of IGF-1 in 126 patients with SPP during a six-month period. The re-

sults did not show any difference between the two groups²³.

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