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RESEARCH ARTICLE **OPEN ACCESS**

POST-POLIO SYNDROME AND THE PHANTOM OF ACUTE PREVIOUS POLIOMYELITIS: A SYSTEMIC ENTITY

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ABSTRACT

Introduction: Poliomyelitis, often referred to as infantile paralysis or polio, was first described in 1840 by Jakob Von Heine, a German orthopedic physician. It is an acute and infectious disease caused by an enterovirus of worldwide distribution. With the implementation of immunization, the incidence of cases has reduced exponentially worldwide. However, a portion of individuals who have already developed polio, are now manifesting the late effects of polio, called post-polio syndrome (PPS). **Case Report:** PRF, male, system analyst. Diagnosis of acute previous polio at the 18th month of age in a hospital in Fortaleza in 1965. At the age of 18 he started to present a new clinic of muscle fatigue, weakness and inability to perform his daily activities. **Discussion:** The Post-polio syndrome (PPS) was first reported in 1875, by Raymond, when reporting the case of a 19-year-old man, previously infected with acute previous polio and at that time had presented a new clinic of muscle weakness and atrophy. Post-poliomyelitis syndrome is defined as a presentation of the delayed effects of polio with the appearance of new neuromuscular symptoms that occur at least 15 years after clinical and functional stability in the natural history of patients with a previous history of acute previous polio. It can be characterized by: new muscle weakness, atrophy and pain, fatigue, sleep disorders, joint pain, cold intolerance, recent weight gain, respiratory distress and dysphagia. Criteria that support the diagnosis are used, such as: (1) Having been affected with acute previous polio; (2) at least 15 years of clinical stability; (3) new clinic of muscle weakness and fatigue; (4) complete or partial recovery of the functions lost in the poliomyelitis period, with a subsequent decline; (5) no other clinical conditions that explain the case. However, effective monitoring by a multidisciplinary team is necessary to assess the degree of functional and motor deterioration, respiratory capacity, as well as the psychological assessment of the patient. **Conclusion:** Although acute previous poliomyelitis has been eradicated in Brazil, as well as in several countries in the world, it is estimated that today there are still about 12 million individuals with poliomyelitis after-effects.

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INTRODUCTION

Poliomyelitis, often referred to as infantile paralysis or polio, was first described in 1840 by Jakob Von Heine, a German orthopedic physician.

The word polio comes from the Greek, where "polio" means gray, "myelo" refers to the spinal cord and "ite" refers to inflammation (Chamberlin, 2005). It is an acute and infectious disease caused by an enterovirus of worldwide distribution, constituted by a simple chain of ribonucleic acid (RNA), in which the transmissibility occurs via oral-fecal, through the

contact with objects, food and / or water contaminated with feces. In addition, direct transmission from person to person can occur through nasopharyngeal secretions (Cohen, 2004).
In 1908, the etiological agent of polio, poliovirus, was recognized by the physician and biologist Karl Landsteiner. This virus belongs to the genus Enterovirus and the family Picornaviridae. Poliovirus has a high infectivity capacity, and its power to penetrate and multiply is by 100%. However, its pathogenicity is low, from 0.1 to 2.0% of affected individuals the paralytic form of the disease occurs. Lethality ranges from 2 to 10%, however, this can be higher, depending on the clinical form of the disease (Chamberlin, 2005). Although 90% of those affected by the poliovirus do not show symptoms, as the infection is restricted to the gastrointestinal tract and the nasopharyngeal region, there is a portion that can present a diversity of symptoms if the poliovirus reaches the bloodstream. Only in 0.1 to 2.0% of cases, the virus reaches the central nervous system (CNS), infecting and suppressing, especially, motor neurons, causing acute flaccid paralysis, hyporeflexia, muscle weakness and fasciculation (Orsini, 2008).

Based on the assumption that the virus affects spinal motor neurons when it affects the CNS, it is stated that poliomyelitis is a neuromuscular disease. The diseases classified in the neuromuscular scope are configured as a group of disorders that affect the motor unit, that is, the cell body of the lower motor neuron, its respective extension, the neuromuscular junction or the skeletal striated muscle. Among neuromuscular diseases, motor neuropathies stand out, representing pathologies where there are morphological or biochemical changes in the neuron. Such conditions are configured by the involvement of the lower motor neuron cell body and the main prototypes are: acute previous poliomyelitis, motor neuron disease and progressive spinal muscular atrophy (Oliveira, 2007). In acute poliomyelitis infection, poliovirus enters the CNS, causing partial or total injury to spinal motor neurons, with denervation of muscle fibers, followed by flaccid paralysis. During the patient's rehabilitation phase, through neuronal plasticity, axonal sprouting takes place, which reinnervates the denervated muscle fibers due to acute infection, recomposing the functional muscle capacity, even if partially. This rehabilitation of functional capacity is called the latency period, also called the stability plateau, and is essentially related to the number of neurons preserved. This hard neurophysiological complex lasts between six to eight weeks and constitutes “giant motor units”, considering that a motor neuron that previously coordinated 200 muscle fibers starts to coordinate from 800 to 1,000 fibers. In addition, it contributes to the rehabilitation of muscle functional capacity, the atrophy of the remaining muscle fibers and the increase in the amount of type I fibers (slow-twitch fibers) (Dalakas, 1995). The poliovirus contaminated and killed thousands of people, and only with the creation of the vaccine composed of inactive virus in 1955, by Jonas Salk, and then by Albert Bruce Sabin in 1961, there was a minimization of the number of cases of poliomyelitis in the world. In 1961, the Sabin vaccine was officially sanctioned in Brazil, replacing Salk, thanks to its low cost, ease of administration, harmlessness, as well as its greater protective effect and ability to pluralize in the gastrointestinal system, enabling the eradication of vaccine viruses in the environment (Orsini, 2010). In 1984, in Brazil, it was the last record of the poliomyelitis epidemic, the last case being documented only in 1989. From 1990, Brazil met all the requirements determined by the International Commission for the Certification of Poliomyelitis Eradication, obtaining the certificate of interruption of native poliovirus transmission in 1994 (Lira, 2009). With the implementation of immunization, the incidence of cases has reduced exponentially worldwide. However, a portion of individuals who have already developed polio, are now manifesting the late effects of poliomyelitis. Such effects present themselves clinically as a new clinic of muscle weakness and fatigue, muscle and joint pain, dysphagia, breathing and sleep disorders, among other symptoms, in which in their totality they are called Post-Polio Syndrome (PPS). Thereby, the objective of the present study is to expose a case of post-polio syndrome in a patient previously affected by acute previous poliomyelitis (Orsini, 2011).

CASE REPORT: PRF, male, system analyst. Diagnosis of acute previous poliomyelitis at 18th month of age in a hospital in Fortaleza in 1965. At the time, he was hospitalized with the flu. Weeks after hospital discharge, he already had amyotrophy in the left upper limb and in the contralateral lower limb. He underwent numerous rehabilitation treatments. He underwent shoulder joint surgery at age 15, partially recovering motor function. At the age of 18 he started to present a new clinic of muscle fatigue and weakness and inability to perform his daily activities. The disease goes on with depression, cold intolerance and myoarticular pain. In use of: Gabapentin, Duloxetine, muscle relaxants and opiates. He presents scoliosis due to inadequate movement synergies and discrepancy between the lower limbs, evidence of reduced respiratory function, dyspnea on small efforts (Fig.1). ENM: pattern of chronic pre-ganglion denervation and neurogenic impairment. The diagnosis of post-poliomyelitis syndrome was established about 10 years ago according to the Dalakas criteria.

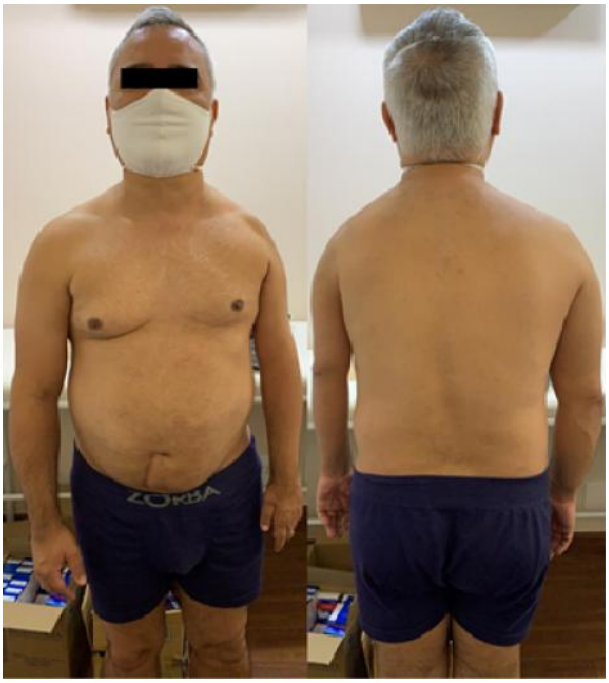

Figure 1. Scoliosis due to inadequate movement synergies and discrepancy between the lower limbs

DISCUSSION

Post-poliomyelitis syndrome (PPS) was first reported in 1875, by Raymond, when reporting the case of a 19-year-old man, previously infected with acute previous poliomyelitis and at that time had presented a new clinic of muscle weakness and atrophy (Nollet, 2004).

In addition, paresis was observed in the left upper and left lower limb. Approximately 100 years after the event, research has shown that individuals with a previous history of acute previous poliomyelitis could develop, after years of clinical and functional stability, new signs and symptoms, as well as muscle weakness and atrophy. Since then, PPS has come to be recognized as a clinical condition that can affect patients previously with poliomyelitis (Kayser-Gatchalian, 1973; Mulder, 1972). Post-poliomyelitis syndrome is defined as a presentation of the delayed effects of poliomyelitis with the appearance of new neuromuscular symptoms that occur at least 15 years after clinical and functional stability in the natural history of patients with a previous history of acute previous poliomyelitis. It can be characterized by: new muscle weakness, atrophy and pain, fatigue, sleep disorders, joint pain, cold intolerance, recent weight gain, respiratory distress and dysphagia (Orsini, 2009). Neuromuscular symptoms can occur in limbs previously damaged by injury or not. The natural history is characterized by a slow worsening of the signs and symptoms of PPS, thus being characterized as a progressive disease (Trojan, 1997; Gawne, 1995). Although the pathophysiology of the symptoms is not well understood, several hypotheses have been raised. Among such mechanisms, the most widely accepted suggests that the degeneration or dysfunction of giant motor units, presented through peripheral deterioration (neuromuscular junction and / or axon), are possibly the consequence of the abundant metabolic convocation of giant motor units (muscle overuse) (Borg, 1989; Grimby, 1989).

However, there are several hypotheses correlated with the pathophysiology of PPS, to mention: predisposition to motor neuron degeneration thanks to glial, vascular and lymphatic detriment, muscle disuse, growth hormone result, decrease in motor units with age, poliomyelitis' associated immunological components, pain, weight gain, reactivation of poliovirus or persistent infection, combined effect of overuse, among other pathologies (Dalakas, 2005). Without elaborating, the patients most likely to evolve with PPS are those who have experienced the history of the most severe acute poliomyelitis. However, several patients with a classic report of PPS have had a history of mild acute poliomyelitis with excellent clinical resolution. Some reasons for the new progressive weakness are highlighted: report of hospitalization and use of ventilatory support during the acute phase of poliomyelitis; initial age of infection (older people tend to be at higher risk for developing new neurological symptoms); excessive weakness in the acute stage of poliomyelitis; period of clinical stability, recent weight gain; muscle pain associated with physical effort; paralytic involvement in all limbs (quadriparesis); and age (Trojan, 1994).

With regard to the diagnostic criteria of PPS, it requires a clinical evaluation in order to exclude other neurological, orthopedic, psychiatric diseases, even the natural senescence process, since they would be able to develop the same signs and symptoms as PPS. In addition, criteria that support the diagnosis are used, such as: (1) Having been affected with acute previous polio; (2) at least 15 years of clinical stability; (3) new clinic of muscle weakness and fatigue; (4) complete or partial recovery of the functions lost in the poliomyelitis period, with a subsequent decline; (5) no other clinical conditions that explain the case. It is important to note that there are no biochemical and / or physiological markers for the diagnosis, making it necessary to distinguish between the symptoms of PPS and other health conditions that affect the elderly population (Dalakas, 1995). It is evident that such patients need with extreme rigor to be evaluated and continuously monitored by a team of health professionals composed of rheumatologist, pulmonologist, neurologist, orthopedist, physical educator, nutritionist, physiotherapist and psychologist, in order to measure the degree of involvement and suggest new treatment strategies. Although the patient who has had acute previous poliomyelitis is psychologically stable in accepting his limitations and lives, within his physical disabilities, with motivation and perseverance, when the symptoms of post-polio syndrome start to appear these individuals become able to effectively monitor the multidisciplinary team to assess the degree of functional and motor deterioration, respiratory capacity, as well as the psychological assessment of the patient (Lira, 2009). In addition, excessive physical exercise in order to obtain muscle mass can be one of the triggering components of PPS. However, some activities have a beneficial effect and are essential to prevent changes resulting from immobility. Previous research (Trojan, 2005) has highlighted the importance of muscle conditioning in the patient's quality of life. Symptoms of weakness, fatigue and pain are precipitated when performed with activities gradually increasing in difficulty.

The exercises performed in heated pools, on the other hand, usually assist in physical conditioning, providing mobility and reducing pain (Lira, 2009). Patients with respiratory distress need to be monitored with extreme care, in addition to needing observation regarding the risk of developing pulmonary infection. They should receive antibiotic prophylaxis, immunization for influenza and pneumococci, and preserve themselves from smoking. Mobility, in turn, can be impaired by excess weight, which can lead to the development of osteoarthritis and respiratory failure thanks to hypoventilation and obstructive sleep apnea. However, there is usually a difficulty for these patients to lose weight due to reduced mobility, and thus the nutritionist is extremely important (Orsini, 2007). As for pain management, it is necessary to highlight that this condition can be difficult to manage because it is generally generalized and not located in a joint or limb. Simple physical interventions such as heat, cold, massage and passive stretching can be of great value to these patients, as well as transcutaneous electrical nerve stimulation and acupuncture (Willén, 2007).

CONCLUSION

Although acute previous polio has been eradicated in Brazil, as well as in several countries in the world, it is estimated that today there are still about 12 million individuals with poliomyelitis after-effects. These patients overload the health system, especially those diagnosed with post-polio syndrome. Therefore, studies in different areas of health sciences are extremely important for understanding the pathophysiological mechanisms and with the aim of improving the quality of life of these patients.

Studies that address the biochemical and physiological issue can be useful for understanding the neurodegeneration that occurs in PPS, in addition to composing material for the study of the repercussion of a physical / motor after-effect on one or more organic systems and on functional capacity.

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