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A painful skeleton disarticulated by acute anterior poliomyelitis

Marco Orsini, Luciana Armada, Jacqueline Fernandes do Nascimento, Adalgiza Mafra Moreno, Antônio Marcos da Silva Catharino

ABSTRACT

Introduction: Acute anterior poliomyelitis (AAP) is an endemic human disease caused by a worldwide spreading enterovirus. The late consequences of AAP include a new picture of muscle weakness together with abnormal muscle fatigue, amyotrophy, and myoarticular pain, configuring a condition known as post-polio syndrome (PPS). This new condition comprises a specific set of health problems due to the polio virus, resulting in decreased functional capacity and/or the onset of new disabilities.

Case Report: A 53-year-old female, human resources analyst, reported that she had been shaken by AAP at the age of 2. Currently she was presented with gait on the knees, osteo-myoarticular pain, inadequate synergies of movements. Imaging exams showed great disproportion in the pelvic region with marked amyotrophy and liposubstitution of the muscle, more evident in the left leg. Although there was some preservation of the plantar flexor muscles, interstitial edema was evident. In the thighs there was amyotrophy with liposubstitution of the muscle, more evident on the left. In the left knee joint there was a lesion of the cruciate ligaments with extensive deep chondral erosions in the load area of the femorotibial compartment, with exposure of the subchondral bone, without edema. Osteopenia also affected it.

Conclusion: Through the data obtained in this study, it can be seen that there are many reasons why patients affected by post-polio syndrome develops bone deformities and joint interferences that cause the individual to suffer losses and aggravations in health and quality of life. Nevertheless, there is a lack of theoretical framework available in the literature, justifying the importance of further studies on this topic.

Keywords: Fatigue, Myoarticular deformities, Pain, Post-polio syndrome

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INTRODUCTION

Acute anterior poliomyelitis (AAP) is an endemic human disease caused by an enterovirus with worldwide dissemination, which is expressed in a biphasic form. It initially presents with fever, headache, and gastrointestinal manifestations, followed by an accelerated paralysis of asymmetric character, resulting from the involvement of

the motor neurons of the anterior tip of the spinal cord [1, 2].

Several patients who developed the condition during the epidemics of the 1940s and 1950s went on to present the late consequences of AAP, which include a new picture of muscle weakness plus abnormal muscle fatigue, amyotrophy and myoarticular pain, forming a condition known as post-polio syndrome (PPS) [3]. Such consequences are correlated to the damage caused by the virus in the acute phase and to the relative muscle overuse during the clinical stability phase, motivating the degeneration of the axonal sprouts of the giant motor units that reproduced during the recovery from the AAP [4].

The incidence and prevalence of PPS are obscure in the world, as well as in Brazil. According to the World Health Organization (WHO), about 12 million people worldwide are thought to have some degree of physical limitation caused by polio [5].

This new condition comprises a specific set of health problems due to the polio virus, resulting in decreased functional capacity and/or the onset of new disabilities [6].

Based on this premise, this article aims to demonstrate, through a case report, the myoarticular deformities arising from new motor neuronal deterioration years after the acute form of the disease, which led to functional disabilities.

CASE REPORT

A 53-year-old female, human resources analyst, reported that she had been shaken by acute anterior poliomyelitis at the age of 2. At that time she was already walking. When she was affected by the viral infection, she stopped walking, presenting fever and malaise (according to her mother's report). She was immunized with Sabin, but the disease broke out after immunization. Currently she presented with gait on his knees, osteo-myoarticular pain, inadequate synergies of movements. A tendon transposition was performed in the inferior limb; this did not improve the quality of the deambulation patterns. Regarding the imaging exams, she presented great disproportion in the pelvic region (Figure 1), accentuated amyotrophy and liposubstitution of the muscle venters, more evident in the left leg. Although there was some preservation of the plantar flexor muscles, interstitial edema was evident. In the thighs there was amyotrophy with liposubstitution of the muscle venters, more evident on the left (Figure 2). In the left knee joint there was cruciate ligament injury with extensive deep chondral erosions in the loading area of the femorotibial compartment, with exposure of subchondral bone, without edema (Figure 3). Osteopenia also affected it, requiring calcium and vitamin D replacement. Regarding the visual analog scale, pain degree signaled 5. In sum, myofascial and neuropathic pain were associated with the myoarticular picture, using Gabapentin 300 mg – 3× a day. Physical therapy program began 2× a week, being oriented not to overload the already weakened muscles. Presence of pain and abnormal muscle fatigue were signs that the proposed activities were above the maximum limit. There was an overlapping between pain, discouragement, and depression.

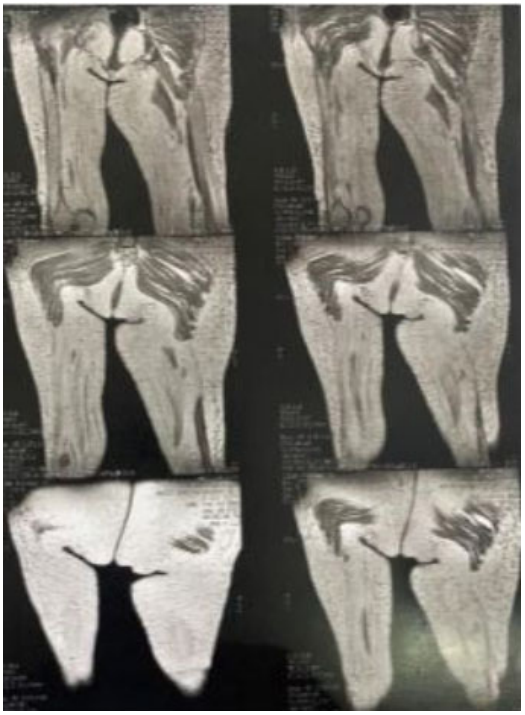

Figure 1: Great disproportion in the pelvic region.

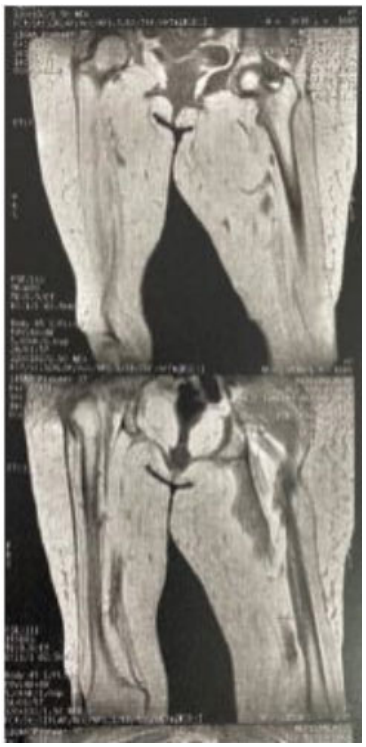

Figure 2: Amyotrophy with liposubstitution of the muscle venters, more evident on the left.

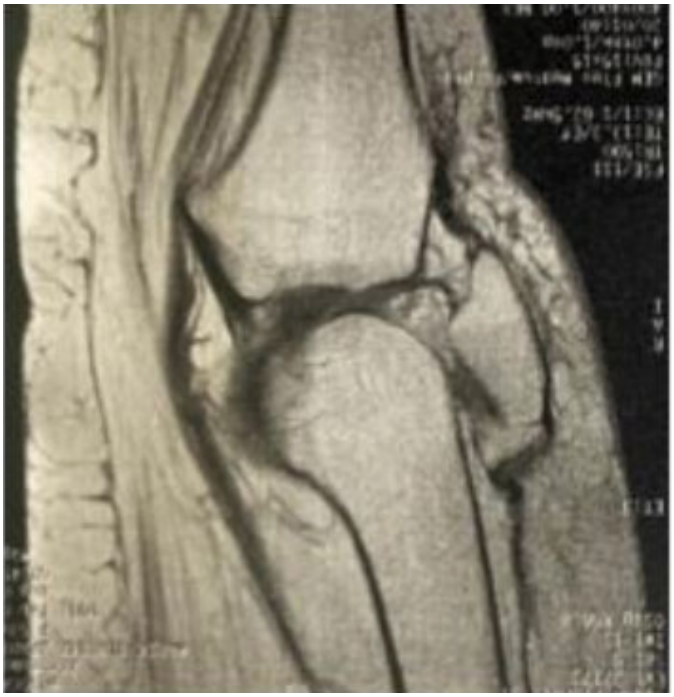

Figure 3: Cruciate ligament injury with extensive deep chondral erosions in the loading area of the femorotibial compartment, with exposure of the subchondral bone, without edema.

DISCUSSION

Post-polio syndrome is a slowly progressive disease, often insidious in onset, that can lead to disabilities, determining functional limitations in basic and instrumental activities of daily living. It is characterized by the appearance of new neuromuscular symptoms, following a minimum of 15 years of clinical stability [7]. The average interval between the AAP and the first presentations of PPS is approximately 35 years, with minimum variations 8 years and maximum71 years. The chronicity of symptoms in PPS, such as pain and myoarticular disfigurement, can lead to significant physical and emotional disability for the patient, leading to negative impacts on quality of life [8].

After acute infection, the poliovirus invades the central nervous system, damaging the cells in the anterior horn of the spinal cord with denervation of the muscle groups coincident with the motor neurons (NM) implicated, which becomes denervated, causing paralysis, amyotrophy, osteo-myoarticular pain, and inadequate synergies of movements [9].

The reestablishment of muscular strength and the reinforcement of functional capacity are achieved by means of some physiological compensatory processes, namely: modification of the muscle fiber type, muscle fiber hypertrophy, terminal sprouting, active denervation/reinnervation process and the plasticity [10]. However, as the years go by, the compensatory mechanisms fail and the striated skeletal musculature begins to deteriorate, a fact that results in bone disarticulation and numerous functional disabilities related to the synergy of movements [11].

Myoarticular pain is the first or second symptom most commonly reported by patients [12]. Vasiliadis et al., in a multicenter study of 126 patients with PPS, observed an intense relationship between female gender and the presence of myoarticular pain, which corroborates the present study. Women with PPS manifested, when compared to men, a higher risk for myoarticular and neuropathic pain [13].

Willén et al., aiming to analyze the clinical and functional changes in a specific group of patients with PPS, developed a longitudinal study with a duration of four years [14]. About 106 individuals were included in the study, and submitted to two evaluations, with the same time interval [15]. Assessments of muscle strength (Medical Research Council), gait speed and quality of life (Nottingham Health Profile) were performed. The results expressed for an attenuation of muscle strength in certain myotomes and rapid decrease in gait speed. No significant changes were confirmed in the quality of life indicator. The authors inferred that the few changes exhibited by the patients did not provide a solid establishment as to which subjects are considered to be at risk for functional deterioration [16].

Abnormal muscle fatigue is a reality in almost all patients with PPS and is one of the symptoms that most often leads to disability [17]. It is often conceptualized as an exhaustion that worsens with minimal physical activity, and usually shows a gradual course throughout the day. It is presumed to be caused by the process of distal degeneration of motor units, producing neuromuscular junction failure [18].

Pain is also mentioned as a common symptom in about 65% of the patients with PPS, and this is of muscle or joint origin, which provides restrictions on daily activities in varying degrees [19]. Koh et al., in a study that disserted as a proposal to qualify the functional deterioration and link it to the presence of pain, concluded that the "overtraining" determined to the muscles of the upper limb, especially in patients who needed assistance for mobility, was closely related to the presence of pain in this region. Moreover, muscle weakness in PPS establishes itself gradually and intervenes directly in the attenuation of functionality [20].

In view of the theoretical framework alluded to, such individuals must preserve energy, since high-intensity exercise can instigate the degeneration of neurons in the anterior tip of the spinal cord [11]. Therefore, it is suggested to attenuate mechanical stress, provide support to exhausted muscles, and stabilize abnormal movements of joints in order to minimize compensations by curbing further losses in relation to muscle strength, muscle mass, and enhance the execution of routine activities [21, 22].

There are several hypotheses for the cause of PPS, but the most accepted is that it is not caused by new poliovirus activity, but by the overuse of motor neurons over the years. The virus can damage up to 95% of the

motor neurons in the anterior horn of the spinal cord, killing at least 50% of them. With the death of these neurons, the muscles in their area of action are without innervation, causing paralysis and atrophy. Though damaged, the remaining neurons compensate for the damage by sending out branches to activate these "orphaned" muscles. With this, neuromuscular function is partially or totally recovered, depending on the number of neurons involved in the "adoption." A single neuron can launch leads to connect 5–10 times more neurons than it originally did. Thus, a neuron innervates a much larger number of neuromuscular fibers than it normally would, restoring motor function. However, overloaded after many years of functional stability, it begins to degenerate, resulting in a new symptomatological picture [23].

Patients add the damage caused by PPS, in addition to the inadequate movement synergies caused by muscle compensation mechanisms. We can conclude that the myoarticular damage, when it emerges, is not only from the PPS, but also from the excessive use of the joints [24].

The incidence and prevalence of PPS are unknown in the world and in Brazil. WHO estimates that there are 12,000 people worldwide with some degree of physical limitation caused by polio [25].

CONCLUSION

Through the data obtained in this study, it can be seen that there are many reasons why patients with post-polio syndrome develop bone deformities and joint interferences that cause the individual to suffer losses and aggravations in health and quality of life. Nevertheless, there is a lack of theoretical framework available in the literature, justifying the importance of further studies on this topic.

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Author Contributions

Marco Orsini – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Luciana Armada – Conception of the work, Acquisition of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Jacqueline Fernandes do Nascimento – Design of the work, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Adalgiza Mafra Moreno – Conception of the work, Analysis of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Antônio Marcos da Silva Catharino – Conception of the work, Design of the work, Analysis of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Guarantor of Submission

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Written informed consent was obtained from the patient for publication of this article.

Conflict of Interest

Authors declare no conflict of interest.

Data Availability

All relevant data are within the paper and its Supporting Information files.