Frequency and clinical manifestations of post-poliomyelitis syndrome in a Brazilian tertiary care center

Frequência e manifestações clínicas da síndrome pós-poliomielite em um centro terciário brasileiro

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ABSTRACT

Objective: To determine the frequency and clinical manifestations of patients with post-poliomyelitis syndrome (PPS) in a Brazilian division of neuromuscular disorders. **Methods:** A total of 167 patients with prior history of paralytic poliomyelitis was investigated for PPS, based on international diagnostic criteria. Other variables analyzed were: gender, race, age at poliomyelitis infection, age at PPS onset, and PPS symptoms. **Results:** One hundred and twenty-nine patients presented PPS, corresponding to 77.2% of the studied population. 62.8% were women and 37.2% were men. Mean age of patients with PPS at onset of PPS symptoms was 39.9±9.69 years. Their main clinical manifestations were: new weakness in the previously affected limbs (69%) and in the apparently not affected limbs (31%); joint pain (79.8%); fatigue (77.5%); muscle pain (76%); and cold intolerance (69.8%). **Conclusions:** Most patients of our sample presented PPS. In Brazil, PPS frequency and clinical features are quite similar to those of other countries.

Key words: post-polio syndrome, poliomyelitis, prevalence.

RESUMO

Objetivo: Determinar a frequência e as manifestações clínicas de pacientes com síndrome pós-poliomielite (SPP) em um setor terciário de doenças neuromusculares brasileiro. **Métodos:** Um total de 167 pacientes com história prévia de poliomielite paralítica foi estudado para diagnóstico de SPP, de acordo com critérios diagnósticos internacionais. Além da SPP, as variáveis analisadas foram: gênero, raça, idade à época da poliomielite aguda e idade no início dos sintomas da SPP. **Resultados:** Cento e vinte e nove pacientes apresentaram SPP, correspondendo a 77,2% da população estudada. Mulheres constituíram 62,8% dos pacientes e os homens, 37,2%. A média de idade dos pacientes com SPP à época do início dos sintomas foi de 39,9±9,69 anos. Suas principais manifestações clínicas foram: manifestações novas de fraqueza em membros previamente afetados (69%) e em membros aparentemente não afetados (31%); dores articulares (79,8%); fadiga (77,5%); dor muscular (76%) e intolerância ao frio (69,8%). **Conclusões:** A maioria dos pacientes da presente casuística apresentou SPP. No Brasil, a frequência e as características clínicas da SPP são similares às observadas em outros países.

Palavras-Chave: síndrome pós-poliomielite, poliomielite, prevalência.

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Paralytic poliomyelitis is an old disease that occurred sporadically from 1600 to 1300 BC[1]. However, epidemic poliomyelitis is a modern disease related to improvement of sanitary conditions of the Western world[2]. The first epidemiological data on poliomyelitis in Brazil were known in 1910[3]. In 1994, the World Health Organization (WHO) declared Brazil as polio-free[4].

Some patients have shown a new set of symptoms decades after recovery from acute poliomyelitis, which has been called post-poliomyelitis syndrome (PPS). This is a neurological disorder characterized by a cluster of symptoms, such as new weakness, abnormal muscle fatigability, and muscle and joint pain. Less commonly, muscle atrophy, breathing and swallowing difficulties, and cold intolerance may be present[5]. For a long time, the PPS was not considered a disease, and since October 2008 the WHO, in its International Classification of Diseases (ICD), recognized it as a distinct and incurable disease (ICD=G14).

In Brazil, few epidemiological studies on the frequency and on the symptoms of PPS were performed. In this scenario, our objective was to determine the frequency and the clinical manifestations of a large series of patients with PPS in a Brazilian division of neuromuscular disorders.

METHODS

This study included 167 consecutive patients with prior history of poliomyelitis, who were admitted between March 2003 and June 2004 in the post-poliomyelitis outpatient sector of the Division of Neuromuscular Diseases from the Department of Neurology and Neurosurgery of the Federal University of São Paulo (Unifesp). All patients had paralytic poliomyelitis, were born and lived in Brazil. All of them were over 18 years of age.

PPS diagnosis was performed according to Halstead's criteria[6]. Furthermore, in our sample, we included only patients who had functional and neurological stability for at least 15 years, and the new symptoms should have persisted for more than one year[7].

Exclusion factors included: patients with paralysis who had no clear history of acute infection by the poliovirus; electroneuromyography without typical features of prior infection by the poliovirus; and patients with other motor neuron diseases, such as amyotrophic lateral sclerosis or progressive spinal muscular atrophy.

Electroneuromyography was performed in all patients, and identified typical features of prior infection by the poliovirus.

The evaluation protocol consisted of a questionnaire containing the following variables: age, gender, race, age at acute infection by poliovirus, age at onset of PPS symptoms, PPS symptoms (new weakness, muscle pain, joint pain, fatigue, cold intolerance, cramps, new atrophy, fasciculation, headache, dyspnea, and dysphagia). A neurological examination was performed by neurologists experienced in neuromuscular diseases. Fatigue was considered in those who met the score in the Fatigue Severity Scale (FSS).

RESULTS

From the 167 patients included in the study, 129 (77.2%) were classified as bearing PPS, of which 81 (62.8%) were women and 48 (37.2%) were men. Mean age of post-polio patients at the time of data collection was 47.3±10.07 years. Their average age at acute poliomyelitis infection was 1.11±2.57 years, and at PPS onset was 39.9±9.69 years. Their average time between polio and onset of post-polio symptoms was 37.0±10.75 years. Ninety percent of patients with PPS had not been previously vaccinated (Table 1).

The main clinical manifestations of PPS patients were: new weakness (n=129, 100%); joint pain (n=103, 79.8%); fatigue (n=100, 77.5%); muscle pain (n=98, 76.0%); cold intolerance (n=90, 69.8%); cramps (n=86, 66.7%); fasciculations (n=68, 52.7%); new atrophy (n=63, 48.8%); headache (n=62, 48.1%); dyspnea (n=53, 41.1%); and dysphagia (n=27, 20.9%), as seen in Table 2.

New muscle weakness occurred in the limb with motor sequelae in 69% of patients, while 31% of them developed weakness in unaffected limbs.

Table 1. Epidemiological characteristics of post-polio syndrome.

Demographics	n (%)
Total of patients	129 (77.2)
Sex	
Males	48 (37.2)
Females	81 (62.8)
Preceding vaccine	11 (9.1)
Mean age acute polio[a]	1.11 (±2.57)*
Mean age at PPS onset	39.9 (±9.69)*
Mean age at evaluation[a]	47.3 (±10.07)*

[a]expressed in years; n: number of patients; PPS: post-poliomyelitis syndrome; *mean ± standard deviation

Table 2. Clinical complaints of patients with post-polio syndrome.

Complaint	n	%
New weakness	129	100
Joint pain	103	79.8
Fatigue	100	77.5
Muscle pain	98	76.0
Cold intolerance	90	69.8
Cramps	86	66.7
New atrophy	63	48.8
Headache	62	48.1
Respiratory complaints	53	41.1
Dysphagia	27	20.9

n: number of patients.

DISCUSSION

In Brazil, there are few studies on epidemiological features of PPS. It was first reported by Oliveira and Maynard, who saw PPS symptoms in 68% of patients with prior history of poliomyelitis[8]. Moreover, Conde et al. evaluated medical records of 132 Brazilian patients with PPS and described predictor factors of its severity[9].

PPS prevalence depends on the clinical diagnostic criteria applied and on population studied, ranging from 20 to 80%[7,10]. In our sample, we found a PPS frequency of 77.2%, which is consistent with studies conducted in other countries. A weakness of our study might be the fact that most patients lived in the state of São Paulo, and the data may not properly represent the entire Brazilian population.

While many patients in our study had recent complaints of decline in muscle strength, the detailed clinical interview revealed that it occurred after a prolonged period of clinical stability, ranging from 15 to 69 years (average time between the polio and onset of post-polio symptoms was 37±10.75 years). In literature, Halstead and Rossi[11] found an average time of functional stability of 25 years and an average time between the polio and the onset of PPS of 33 years. Dalakas et al.[12] found a plateau of stability ranging from 15 to 54 years (clinical stability interval=28.8 years).

Regarding PPS symptoms, the most frequent in our sample were new weakness (100%) followed by joint pain (79.8%), fatigue (77.5%), and muscle pain (76%), which are also the most common symptoms reported in different studies[7,9,11-14].

In conclusion, our series of post-polio patients report frequency and clinical manifestations similar to other series of non-Brazilian populations and contribute to better understanding regional characteristics of the illness.

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