The Complexity of the Diagnosis of Post-Polio Syndrome

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Abstract
The diagnosis of post-polio syndrome (PPS) might be difficult as the diagnosis PPS mostly depends on the patient’s subjective description of the symptoms. Furthermore, the PPS patients have reached an age when concomitant diseases are common. However, an adequate diagnosis is important for treatment and prognosis. The aim of this case report is to describe the complexity of the PPS diagnosis.

Case Report
A 50 year old woman from Iran who contracted polio at the age of two years with remaining moderate paresis of both legs. Since five years she suffers from fibromyalgia with tender-points and diabetes mellitus. She suffers from increasing weakness of her arms/hands and a numbness of her fingers. Electrophysiological examination and neurological evaluation confirmed a diagnosis diabetic polyneuropathy.

Discussion
The patients in the case report suffers from a clinical deterioration with pain, fatigue and muscle weakness which are main findings in the diagnosis PPS. However, the new or increased symptoms could be explained by concomitant diseases like fibromyalgia, polyneuropathy and diabetes mellitus and not by PPS and therefore she was not diagnosed with PPS. The present case report underlines the necessity to exclude concomitant disorders before the patients receive a PPS diagnosis.

Introduction
Poliomyelitis was before the vaccination against polio started in the 1950's a huge medical and social problem. The last great epidemic took place in Sweden 1953 leaving a large number of persons with polio sequelae. Nowadays many of the prior polio patients suffers from a severe disability including muscular atrophy, loss of muscular function and in severe cases paresis of the respiratory muscles [1]. Furthermore, some patients after a stable period develop new or increased symptoms such as increased muscle weakness, pain, and fatigue and thus are diagnosed with post-polio syndrome (PPS) [2,3]. For the diagnostic criteria of March of Dimes (Table 1). The cause of PPS remains unclear, but is likely due distal degeneration or loss of enlarged post-polioymyelitis motor units [2]. The prevalence of PPS varies in the literature between 20-80% depending on the studied population and the diagnostic criteria. In the study by Ramlow the prevalence was found to be 28.5% [4,5] and in a recent Italian study 41% [4]. The risk of developing PPS increases with time since acute polio infection [4]. However, ages at acute polio infection have shown to have no impact on the development of PPS. Furthermore, female patients and patients with initial paresis of the upper extremities might have a higher risk of developing PPS.

PPS is diagnosed according to the criterions of March of Dimes with the last criteria being that concomitant disorders that may explain the new health problems should be excluded [6]. However, the patients and the examiner might not be aware of concomitant disorder as the background for new or increasing symptoms [7]. PPS might be difficult to diagnose as the diagnosis depend on the patient’s own subjective description of the symptoms. Furthermore, polio patients in Europe have reached the age when concomitant disorders like hypertension, cardiovascular diseases, and diabetes mellitus are commonly present. The aim of the present case report was to illustrate the complexity of the PPS diagnosis and the necessity to exclude concomitant disorders as the background to new or increasing symptoms in patients with prior polio.

Case Presentation
The patient is a 50 year old woman who contracted polio at the age of two years. Since the onset of her polio infection she suffers from moderate paresis of both legs but was able to walk unaided. During the last five year she suffered from generalized pain. She was referred to a pain clinic and was found to have trigger points and was diagnosed with fibromyalgia. Furthermore, she was diagnosed with diabetes mellitus but received no treatment. She reported increasing fatigue and loss of both motor and sensory functions in upper extremities since two years. Her fatigue increased after exercise and she had to rest after walking 100 m. She worked part-time in white collar work. Due to her pain her sleep quality was poor. She did not suffer from a spinal cord disorder, cervical hernia, cervical spondylosis or myofascial pain. She underwent a thorough neurological examination including muscle function, reflexes, sensibility, and presence of muscular atrophy. The muscle strength in key muscles was graded 0-5. 0 means no muscle function, 5, the muscle contracts normally against full resistance. At the examination, her sensitiveness in the upper extremities was normal but she reported numbness in the fingers. Reflexes in the upper extremities were normal but absent in the lower extremities. She showed no atrophy of muscles in the upper extremities. Her bloods samples including hemoglobin, C-reactive protein and thyroid hormones showed normal results except for an elevated level of blood glucose. A standard EMG

1. A history of paralytic poliomyelitis, as confirmed by history, neurologic examination, and electromyography.
2. A period of partial or complete functional recovery, followed by a stable period of at least 15 years.
3. Gradual or sudden onset of progressive and persistent new muscle weakness or decreased endurance, with or without generalized fatigue, muscle atrophy, or muscle and joint pain. (Sudden onset may follow a period of inactivity, trauma or surgery.)
4. The symptoms should persist for at least a year.
5. Exclusion of other causes

Table 1: The criterias of March of Dimes.

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demonstrated increased amplitude and duration of motor units action potentials, increased percentage of polyphasic potentials and a decrease in the numbers of motor units in the lower extremities but not in the upper extremities. The neurophysiologic examination confirmed the diagnosis of diabetic polyneuropathy. She underwent treatment with Immunoglobulin intravenously, 30 g per day during three consecutive days without effect on her pain and fatigue.

Discussion

The diagnosis of PPS is to some extent relying on subjective description of the new or increased symptoms. The present patient suffered from loss of motor function in the upper extremities, pain as well as fatigue which are main findings of PPS. In the upper extremities she had no loss of tendon reflexes and no muscular atrophy which are two of the main findings in PPS. Overuse of muscles may lead to pain but her pain was general and not localized to polio affected muscles which is usually the case in pain after poliomyelitis. Her pain might therefor be caused by her fibromyalgia. However, numbness in the fingers and neurophysiological findings pointed to the diagnosis of diabetic poly-neuropathy. The conclusion is that she did not fulfill the diagnostic criteria’s for PPS and her fatigue might be explained by her diabetes mellitus in combination with poor sleep quality. For the differential diagnosis to fatigue (Table 2). Her sensory and motor loss could not be explained by a spinal cord disorder, cervical hernia, spondylosis or myofascial pain and her poly neuropathy might be the plausible explanation for her sensory disturbances.

The most common concomitant diagnosis are hypertension, other cardiovascular diseases, diabetes mellitus, poly-neuropathy and degenerative disorders as earlier described [8] and might need investigation and treatment in other units. Therefore patients with prior polio needs a careful examination and evaluation at a PPS unit to confirm the diagnose PPS. The examination at the PPS unit must include a complete neurological examination including reflexes and sensibility and a neurophysiologic examination to confirm the diagnosis of PPS and to exclude confounding disorders. Concomitant diagnosis may give symptoms similar in PPS like increasing paresis, pain and fatigue. However, with a correct analysis of the symptoms and complete general and neurological examination polio patients may be correctly diagnosed and thus, may undergo adequate treatment. The treatment options for PPS are physiotherapy, treatment with immunoglobulin and exclusion of concomitant diseases. Treatment with immunoglobulin has been effective in about two thirds of the patients however; we believe it is of essential to choose the right patients for treatment.

References


Table 1: The differential diagnosis to fatigue.

| 1. | Anemia |
| 2. | Endocrinopathia including diabetes mellitus and hypothyreidism |
| 3. | Sleep disturbances |
| 4. | Cardio-vascular diseases |
| 5. | Adrenal insufficiency |
| 6. | malignancies |

Table 2: The differential diagnosis to fatigue.