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CASE REPORT

POST-POLIO SYNDROME, SPINAL CORD INJURY AND STATIN MYOPATHY: DOUBLE TROUBLE OR INCORRECT DIAGNOSIS? TWO CASE REPORTS

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Objective: **To discuss the importance of a thorough clinical examination and evaluation of symptoms in upper and lower motor neurone lesions.**
Design: **Case report.**
Methods: **Post-polio outpatient clinic, Danderyds University Hospital, Stockholm, Sweden. We describe here two patients with a past history of poliomyelitis, who were experiencing increasing muscular weakness. Clinical evaluation led to diagnoses of spinal cord injury and statin myopathy, respectively.**
Conclusion: **In order to make a correct diagnosis it is essential to distinguish between lower and upper motor neurone lesions. In the case of a lower motor neurone disorder a neurophysiological examination is necessary for a correct diagnosis, and is a prerequisite for adequate treatment and rehabilitation.**

Key words: post-polio syndrome; spinal cord injury; statin myopathy; paresis; upper motor neurone disorder; lower motor neurone disorder.

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INTRODUCTION

Although, the poliovirus is almost eradicated worldwide, the number of survivors of poliomyelitis remains high. During recent decades there has been increasing interest in post-polio syndrome (PPS), i.e. increasing or new symptoms developing after a long stable period. The most common symptoms of PPS are weakness and muscle atrophy, fatigue and/or muscle and joint pain (1, 2). PPS is defined by different sets of criteria (3, 4). An important criterion being that other neurological conditions should be ruled out.

Muscular weakness is the main symptom in most neurological disorders, such as spinal cord injury (SCI), and in particular in neuromuscular disorders. PPS is a late effect of poliomyelitis that affects the anterior horn cells, resulting in a lower motor neurone lesion with a flaccid paresis, weak tendon reflexes and a negative Babinski's sign (1–4). Other neuromuscular disorders also result in a lower motor neurone lesion due to their effect on different parts of the motor unit. SCI, however, is the result of an injury of the central nervous system resulting in an upper motor neurone lesion with increased muscle tone, exaggerated tendon reflexes, and a positive Babinski's sign (5). However, a SCI at a low thoracic or lumbar level can be a combination of an upper and lower motor neurone disorder, and a flaccid paresis may be seen. For the classification of SCI we used the American Spinal Injury Association Impairment Scale (AIS) A–E and the neurological level (6). AIS A refers to complete SCI, AIS B–D refers to incomplete SCI, and AIS E refers to normal sensory and motor functions.

In order to make a correct diagnosis it is essential to distinguish between upper and lower motor neurone disorders. There are differences, as mentioned above, in the clinical findings in the different conditions. Furthermore, there are different treatment options and rehabilitation strategies for the two conditions. We describe here two patients with a history of polio, who had developed increasing muscular weakness diagnosed as PPS, in whom clinical neurological examination and neurophysiological investigation revealed a complete spinal cord injury and a statin myopathy, respectively. These cases emphasize the need for thorough neurological examination, adequate interpretation of the results, and neurophysiology in order to make a correct diagnosis.

CASE 1

An 80-year-old woman was referred to our outpatient post-polio clinic by her cardiologist.

She had had poliomyelitis in early childhood. Her left side was affected and she recovered fully. She had had a healthy and active life and was able to walk normally.

During recent years she had developed ventricular arrhythmia and had an internal defibrillator.

In 2008 she had developed bladder problems and difficulty walking. She was not aware of where she put her feet. In August 2009 she was hospitalized due to septicaemia after a urinary tract infection. After discharge from hospital she was not able to walk. Her urologist gave her an indwelling catheter and she experienced cramps in her legs.

She lived in a nursing home and used a wheelchair, but had no pain. At the examination at the post-polio outpatient clinic her general condition was good and she had normal cognitive function.

Neurological examination revealed normal tendon reflexes in the upper body and exaggerated tendon reflexes in the lower

extremities. Babinski's sign was bilaterally positive. A neurological level was found at Th11. Her injury was complete and she had no sacral sensory or motor function. According to the AIS scale she was classified as AIS A with a motor index of 50/100. The muscle tone in her legs was slightly increased according to the Ashworth scale. She had no muscular atrophy.

The symptoms and signs were considered to be in accordance with a complete SCI (6). As a result of the SCI she had a neurogenic bladder dysfunction and her urinary tract infection was caused by incomplete emptying of the bladder. The cramps in her legs were diagnosed as spasticity (5).

She was referred for computed tomography (CT) of her lumbar and thoracic spine. It was not possible to perform magnetic response tornography due to her internal defibrillator. The CT scan showed degenerative changes resulting in a spinal stenosis. Due to her concomitant medical problems surgery was not possible.

CASE 2

A 65-year-old woman presented with a history of clinical diagnosis of poliomyelitis at the age of 2 years, which resulted in a weak left leg and difficulty walking. However, she recovered fully and was able to work until retirement at the age of 65 years. Due to a high level of cholesterol she was started on statin medication. After 2 years she experienced muscle cramps in the jaw and lower extremities, as well as increasing muscle weakness and atrophy of the left leg, and she developed difficulty walking. Clinical examination revealed paresis and atrophy of muscles in the left leg. Her tendon reflexes were weak and Babinski's sign was negative. She had normal sensibility. Neurophysiological examination showed slight signs of myopathy in the gluteus medius muscles on the left side. There were no signs of polio. A medical investigation ruled out other obvious reasons for the myopathy and she was diagnosed with a statin myopathy. Statin medication was stopped and the patient received physiotherapy, and recovered slowly, within a year.

DISCUSSION

These cases both emphasize the importance of performing a thorough neurological examination and of interpreting the results correctly when diagnosing PPS. PPS results in a lower motor neurone lesion with asymmetrical flaccid paralysis and normal sensory function. However, in case 1 there was an upper motor neurone lesion, i.e. the neurological findings were not compatible with PPS, and a SCI was diagnosed. There are different sets of criteria for PPS (3). One main criterion, as given by March of Dimes (4), is the exclusion of other neurological, medical, and orthopaedic causes of the symptoms. In this case this criterion for PPS is not fulfilled and, thus, a diagnosis of PPS cannot be made.

Case 2 shows that even if a lower motor neurone lesion is found it has to be investigated further in order to exclude concomitant disorders and to make a correct diagnosis. Patients who have had polio previously are often elderly and the symptoms of PPS are often vague. This highlights the importance of performing a general examination in order to exclude, for example, cardiovascular disorders.

A correct diagnosis is a prerequisite for adequate therapy. In both of these cases curative therapy was probably available; surgery in case 1 and stopping medication in case 2. However, in case 1 an operation was not performed due to other medical reasons. A correct diagnosis is also of importance for symptomatic therapy and for rehabilitation. For example, muscle cramps are often reported by patients with PPS. Both patients reported muscle cramps. They were, however, due to spasticity in case 1 and are treated in another way than cramps in PPS. The cramps in case 2 disappeared after stopping the statin medication.

Although the goal for rehabilitation is the same in PPS, SCI and myopathy, i.e. to optimize function according to the patient's disability and present symptoms in order to increase activity and participation, the bases for rehabilitation differ. Bladder and bowel problems, as well as sexual dysfunction and spasticity, have to be considered when dealing with patients with SCI.

The diagnosis of acute poliomyelitis in case 2 may be challenged. The patient had a clinical diagnosis of poliomyelitis, but the neurophysiological examination more than 60 years later did not reveal any traces of poliomyelitis. In a sample of patients with PPS, Sandberg et al. (7) found that 5% of patients with an alleged history of polio had normal findings on electromyography. Thus, the diagnosis of poliomyelitis is uncertain, but due to the neurophysiological findings one might conclude that there is no strong evidence of PPS. One might, however, speculate that the statin myopathy in the left leg developed due to a locus minoris resistance resulting from earlier polio.

In summary, these two cases highlight the need for a thorough neurological examination and correct interpretation of the results, as well as the importance of a neurophysiological examination in order to make a correct PPS diagnosis. It is also essential to be aware that a patient can have two conditions at the same time.

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