Original research article

Post-polio syndrome. Cases report and review of literature

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Abstract

It is estimated that around 15 million people survived polio infection worldwide since early twentieth century. In 1950 effective vaccination was used for first time. Since that time number of affected people decreased. The last epidemic of Haine–Medine disease in Poland was in 1950s. Another rare cases of infections were observed till 1970s. About at least 15 years after polio virus infection, slowly progressive muscle limbs paresis with muscle atrophy, joints pain, paresthesia were observed in polio survivors. That constellation of symptoms was called post-polio syndrome (PPS). PPS frequency among people after paralytic and nonparalytic polio infectious is ranged from 30% to 80%. Fatigue that leads to physical and mental activity deterioration is another important symptom that is observed in 90% of patients with PPS. Etiology of disease remains elusive. Probably it is an effect of spine frontal horns motoneurons damage during acute virus polio infection that leads to overloading and degeneration of remaining ones. The most important risk factors of PPS are female sex and respiratory symptoms during acute polio infection. Electromyography is an important part of PPS diagnostic process. Electrophysiological abnormalities are seen in clinically affected and unaffected muscles. The most frequent are fasciculations and fibrillations during rest activity, extension of motor unit area, time duration and amplitude. In this study we described three cases of people who developed PPS years after Haine–Medine disease and correlation between their EMG results and clinical status. We also analyzed electromyography results both after one month since first PPS signs occurred as well as after few years. Presentation of dynamic changes in EMG was the most important aim of that study.

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Sixty-two years old male with hypertension who underwent polio virus infection at age of six years developed at time of disease weakness and atrophy of right lower limb muscles. After infection he was able to walk unassisted and his nervous system progressed quite normally. After 39 years from recovery new neurological symptoms occurred. He complained at intensification of muscles atrophy with accompanying pain and numbness (Photo 1). Symptoms were more intensive during winter and after longer physical activity. Patient claimed that neurological deterioration occurred when he started to work as a postman and partially decrease after buying a car and use it to deliver letters. He also complained at problems with sleeping and concentration, general weakness. Patient has gone to neurologist after four years since that new neurological status deterioration. Blood tests and magnetic resonance (MR) of brain were normal. Electroneurography (ENG) and electromyography (EMG) were done. ENG was normal. EMG study in clinically affected muscles of right leg revealed features of neurogenic dysfunction, significant extension of amplitude, time duration and motor unit area. Signs of chronic denervation were also observed in left leg muscles. Maximal voluntary activity was diminished and single fasciculations and fibrillations were observed during rest activity in muscles (Table 1).

### 2. Case 2

Sixty-years old male with hypertension and digestive heart failure was admitted to neurology department because of progressive weakness and atrophy of right limbs muscles. Symptoms started month earlier. At age of two years he suffered from polio infection. As a result of this, he had right limbs paresis and muscle atrophy (Photos 2–4). During all this years neurological condition was stable. He was able to work with the use of right hand and to walk on crutches. After admission to hospital his neurological state was bad. He had right limbs paresis was severe, he was able to perform only small movements with the hand, could not walk and use a wheelchair. He was depressed and had problems with sleeping. Computer Tomography (CT) of brain did not reveal any abnormalities. ENG study was normal. In EMG study there

### Table 1 - EMG examination of first post-polio patient.

<table>
<thead>
<tr>
<th>Clinically affected muscles</th>
<th>Mean amplitude</th>
<th>Mean time of duration</th>
<th>Percentage of polyphasic potentials</th>
<th>Rest activity</th>
</tr>
</thead>
<tbody>
<tr>
<td>Right tibialis anterior muscle</td>
<td>2924</td>
<td>16.9</td>
<td>32.0</td>
<td>Fasciculations, fibrillations</td>
</tr>
<tr>
<td>Right quadriceps femoris muscle</td>
<td>3162</td>
<td>15.9</td>
<td>29.3</td>
<td>Fasciculations, fibrillations</td>
</tr>
<tr>
<td>Clinically unaffected muscles</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Right biceps</td>
<td>444</td>
<td>10.2</td>
<td>0</td>
<td>–</td>
</tr>
<tr>
<td>Left interosseus dorsalis muscle</td>
<td>752</td>
<td>9.9</td>
<td>0</td>
<td>–</td>
</tr>
<tr>
<td>Left tibialis anterior muscle</td>
<td>1215</td>
<td>12.4</td>
<td>6.7</td>
<td>Fasciculations</td>
</tr>
<tr>
<td>Left quadriceps femoris muscle</td>
<td>1298</td>
<td>12.5</td>
<td>6.7</td>
<td>Fasciculations</td>
</tr>
</tbody>
</table>
were signs of active denervation in right limb muscle, right and left femoral and right calf muscles. There was no voluntary activity in muscles of right arm and signs of chronic denervation in other muscles of left limb were observed (Table 2).

3. Case 3

Fifty-eight years old female with hypertension only who survived from polio infection at age of 6 years, complained at progressive general weakness, problems with sleeping and concentration, joints pain, mood deterioration. Symptoms started five years earlier after respiratory tract infection. She was treated with amitriptyline 25 mg once a day with mild effect. After polio she had paresis of left leg and walk on crutches. Now she claims that also right limbs seem to be disabled from about four years. In ENG study median and ulnar nerves were damaged. In EMG study there were features of chronic denervation in muscle of both arms and right leg while there was no voluntary activity in left leg muscles. Signs of acute denervation were not observed despite of the fact that her neurological state was deteriorating (Table 3).

Post polio syndrome (PPS) is a disease that occurs 30-40 years after acute paralytic and nonparalytic polio virus infection. The most characteristic symptoms are slowly progressive muscle weakness and atrophy accompanied by joints and muscle pain [1]. That symptoms can be observed in clinically affected and nonaffected muscles during primary polio infection. Post polio progressive muscular atrophy (PPMA) is a term used when there is a weakness of only limbs muscles. Usually muscle weakness progression is slow and last for many years. In case of second patient that process was fast and led to severe disability after one month. In PPS also bulbar and respiratory muscles can be affected but in presented study those symptoms were not observed in any of three cases. PPS symptoms are often preluded by infectious, injuries, operations or intensive physical activity like in first and second presented patients. In case of second patient with the most severe disease course, cause of that deterioration

<table>
<thead>
<tr>
<th>Clinically affected muscles</th>
<th>Mean amplitude</th>
<th>Mean time of duration</th>
<th>Percentage of poliphasic potentials</th>
<th>Rest activity</th>
</tr>
</thead>
<tbody>
<tr>
<td>Right I interosseus dorsalis muscle</td>
<td>1806</td>
<td>10.9</td>
<td>9.1</td>
<td>Fibrillations</td>
</tr>
<tr>
<td>Right quadriceps femoris</td>
<td>2172</td>
<td>11.5</td>
<td>13.3</td>
<td>Fasciculations, fibrillations</td>
</tr>
<tr>
<td>Right tibialis anterior</td>
<td>3976</td>
<td>11.9</td>
<td>28.6</td>
<td>Fibrillations, positive sharp waves</td>
</tr>
<tr>
<td>Clinically unaffected muscle</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Left I interosseus dorsalis muscle</td>
<td>1636</td>
<td>10.0</td>
<td>0</td>
<td>–</td>
</tr>
<tr>
<td>Left biceps</td>
<td>1251</td>
<td>10.4</td>
<td>0</td>
<td>–</td>
</tr>
<tr>
<td>Left vastus lateralis muscle</td>
<td>1959</td>
<td>10.1</td>
<td>13.3</td>
<td>Fibrillations</td>
</tr>
<tr>
<td>Left tibialis anterior muscle</td>
<td>12.2</td>
<td>958</td>
<td>5</td>
<td>–</td>
</tr>
</tbody>
</table>
remains unknown. About 90% of PPS patients also complain at general fatigue, problems with concentration and mood disturbances [2,3]. Those symptoms have severe influence on patients’ quality of life, social and occupational activity and they were most evident in third patient. Intensity of general weakness and fatigue can be unstable during the day. In PPS patients reticular activating system (RAS) is damaged and this is probably cause of cognitive performance deterioration and chronic fatigue syndrome [4]. This theory was confirmed in histopathological examination. That studies revealed abnormalities in brainstem of PPS patients [5]. Joints and muscle pain is observed in 70% of PPS patients. Muscle atrophy and intolerance of low temperatures are more rare and are observed in 50% PPS patients [6]. Apnea and dysphagia are observed in case of 30% patients with PPS [7]. The most important risk factors for PPS are female sex and severe course of initial infection [8]. Cause of PPS remains elusive. Probably it is an effect of motoneurons damage during acute virus polio infection that leads to overloading and degeneration of remaining ones in spine frontal horns [8,9]. Diagnostic criteria for PPS include prior paralytic poliomyelitis, period of functional recovery after acute poliomyelitis, symptoms lasting at least 15 years, gradual onset of new muscle weakness in muscles affected and unaffected during initial infection with coexisting general weakness, joints pain, problems with sleeping [1]. In differential diagnosis it is important to exclude orthopedics and neurological diseases similar to PPS like amyotrophic lateral sclerosis, cervical spondylosis, tumors of the cervical and thoracic cord and other causes of chronic fatigue syndrome, myasthenia gravis, myopathies and cardiac, hematologic, endocrine, cancer, chronic systemic infections [10,11]. In PPS diagnostic process important role plays electromyography. Although electrodiagnostic studies cannot differentiate patients with post-polio syndrome from asymptomatic post-polio ones, these studies are important to exclude other neuromuscular diseases like amyotrophic lateral sclerosis, radiculopathy, polyneuropathy, myasthenia gravis, and myopathies. It is also important to confirm past poliomyelitis episode involving frontal spine horns [12,13]. All that abnormalities are observed in PPS patients. Electroneurography is usually normal in PPS patients. In presented study only third patient had damage of median and ulnar nerves connected with comorbid carpal tunnel syndrome and ulnar neuropathy of the elbow. Hachisuka studied 43 polio survivors and 20 healthy controls with motor nerve conduction studies of the median and tibial nerves. They found a significant increase in abnormally stereotyped F-waves and a reduction of F-wave persistence in both nerves in the polio patients compared to the control group. The authors conclude that F-wave reveal electrophysiological features of anterior horn cells in polio survivors and reflect their motor unit loss [14]. Electromyography can reveal both chronic and acute denervation in post polio patients. Due to increased number of muscle fibers innervated by one neuron, motor unit action potentials have abnormally large amplitudes, time of duration and are polyphasic [13]. The most characteristic finding in EMG study in PPS is presence of giant potentials that were present in all three cases. That units are greater than 8 mV and occurs as a result of chronic denervation that lasts for many years and reinervation by sprouts from the same motor unit. Due to high amplitude and large area of motor units size index is also elevated in PPS patients [15]. In Chang study 31 patients with PPS were diagnosed with the use of EMG. In 41 (78.8%) of the 52 muscles motor unit time of duration was increased. In 43 (82.7%) of the 52 muscles increased motor units amplitudes were observed. Polyphasia of more than 20% was found in 46.1% of the affected muscle. Spontaneous activities such as fibrillation and positive sharp waves were observed in about 30% of the tested muscles in this study. Fasciculations occurred in case of 26.9% PPS patients [16]. In presented study two patients had fibrillations and positive sharp waves in muscles connected with acute denervation while third one had only signs of chronic denervation despite of the fact that her neurological state was deteriorating. It is also important to notice that in case of second patient EMG study was done at the beginning of new neurologic symptoms while in case of first and third patient symptoms of PPS last for years before EMG study was done. It is possible that acute denervation was present at time when new neurological deterioration occurred. In case of second patient EMG result was similar to those seen in first one. All patients had electrophysiological abnormalities in clinically affected and nonaffected muscles during polio infection. In another study Thompson revealed that in group of 15 post polio patients all had abnormalities in EMG study but none of them had acute denervation [17]. On the other hand many polio survivors with no neurological symptoms can have fibrillation and positive sharp waves in muscle for years after recovery. The reason for this remains elusive. Proposed mechanisms include ineffective renovation of motor units due to metabolic abnormalities connected with the polio virus infection [18]. That data was confirmed in

<table>
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<th>Mean time of duration</th>
<th>Percentage of polyphasic potentials</th>
<th>Rest activity</th>
</tr>
</thead>
<tbody>
<tr>
<td>Left quadriceps femoris muscle</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>Left tibialis anterior muscle</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>Clinically unaffected muscle</td>
<td>Left biceps</td>
<td>584</td>
<td>12.5</td>
<td>7.8</td>
</tr>
<tr>
<td>Left biceps</td>
<td>668</td>
<td>12.4</td>
<td>5.2</td>
<td>–</td>
</tr>
<tr>
<td>Left interosseus dorsalis</td>
<td>1138</td>
<td>10.9</td>
<td>10</td>
<td>–</td>
</tr>
<tr>
<td>Right quadriceps femoris muscle</td>
<td>3812</td>
<td>16.7</td>
<td>5</td>
<td>–</td>
</tr>
<tr>
<td>Right tibialis anterior</td>
<td>1412</td>
<td>11.3</td>
<td>10</td>
<td>–</td>
</tr>
</tbody>
</table>

Table 3 - EMG examination of third post-polio patient.
histopathological studies in which signs of chronic and acute denervation were observed [19].

The main complaint in PPS patients is permanent or recurring muscle weakness that can be connected with neuromuscular junction deficits [20–22]. Reduction of compound motor action potentials on supramaximal repetitive stimulation was observed in PPS patients in many studies [23]. SFEMG (single fiber electromyography) has become the most sensitive method to diagnose abnormal neuromuscular transmission and is most frequently used in diagnosis of myasthenia. Even subclinical transmission disturbance can be detected with the use of this method [24]. In SFEMG increased jitter, fiber density and blocking were observed in PPS patients [13]. SFEMG can provide information about motor unit reorganization with fiber density. Increased jitter is connected with mild abnormalities and is often asymptomatic while a moderate and severe abnormality is recognized from impulse blocking [24,26]. Cashman claims that there is a relationship between the degree of neuromuscular junction deficits and the degree of motor unit enlargement in PPS patients [13,12]. That SFEMG findings confirm the theory that PPS is a result of nerve terminals degradation that have the influence on neuromuscular junctions condition [16].

Studies with the use of macro EMG in PPS were also described [25,26]. In early stages of PPS macro-EMG amplitudes are increased even 40 times the upper normal limit because of reinnervation process [27]. With time reduction of macro-MUP amplitudes can be observed and confirm the peripheral disintegration of the motor unit [25,27].

There is no cure for PPS. The main goal of treatment is improvement the quality of life. The most important is rehabilitation and resting affected limb with the orthopedic equipment [28,29]. Occupational therapy and psychotherapy are useful. There were also attempts of treatment with pyridostigmine and amantadine but their effectiveness in that disease was not confirmed [30–32]. Also intravenous immunoglobulins are actually tested in PPS [8]. In case of patients with respiratory symptoms assisted ventilation may be necessary [3].

There is no electrophysiological test specific for PPS but EMG is very important part of diagnosis. It helps to exclude other neurological conditions and confirm past poliomyelitis episode. Further electrophysiological studies especially macro EMG and SFEMG may provide more information about the pathophysiology and help to find the cure for PPS.

Conflict of interest

None declared.

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None declared.

Ethics

The work described in this article has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans; Uniform Requirements for manuscripts submitted to Biomedical journals.

REFERENCES