Internal Medicine
The Japanese Society of Internal Medicine
doi: 10.2169/internalmedicine.7985-21
Intern Med Advance Publication
http://internmed.jp

【 CASE REPORT 】

Postpolio Syndrome from Non-Paralytic Poliovirus Infection

Kengo Maeda and Yoshiko Sugihara

Abstract:
A 73-year-old man presented with muscle weakness and atrophy of his right arm. Atrophy of his left brachia and left calf had occurred 13 years before without any improvement or deterioration. His sister and cousin had a history of paralytic poliomyelitis. Serum poliovirus type 2 neutralizing antibody was elevated to 128x. Electromyography revealed chronic denervation potentials not only in the muscles affected previously but also in the unaffected muscles. Acute and chronic denervation potentials were found in the newly affected muscle. Postpolio syndrome should be considered in patients with unilateral muscular atrophy even when they have no history of paralytic poliomyelitis.

Key words: postpolio syndrome, non-paralytic poliomyelitis, electromyography

(Intern Med Advance Publication)
(DOI: 10.2169/internalmedicine.7985-21)

Introduction

The term "postpolio syndrome (PPS)" is applied to paralytic poliomyelitis survivors who develop new weakness or fatigue after achieving a long-term stable condition (1). The new weakness and atrophy affect both previously affected and unaffected muscles. Typically, these are asymmetrical or segmental (2). Polioviruses damage spinal motor neurons in the anterior horn. When the damage is less severe, paralysis is not recognized. An electromyographic (EMG) study revealed that unaffected muscles of PPS patients have large motor units as a result of effective reinnervation (3). There is a possibility that PPS may occur in people with non-paralytic polio.

Case Report

Our patient was a 73-year-old Japanese man who, at 60 years old, had noticed weakness and atrophy of his left arm and calf. At that time, a blood test, including evaluations of anti-Jo-1 antibody, anti-nuclear antibody, anti-acetylcholine receptor antibody angiotensin converting enzyme, lactate/pyruvate, vitamin B1 and B12, anti-scl-70 antibody, and thyroid hormones, a cerebrospinal fluid (CSF) examination, and magnetic resonance imaging (MRI) of the whole spinal cord were all normal. At 69 years old, he came to our hospital complaining of persistent weakness of these muscles. The weakness had been unchanged without additional weakness for nine years. He had a family history of paralytic poliomyelitis in his sister and cousin in their childhood. He had never been abroad. No facial or bulbar palsy was found. His left deltoid, biceps and triceps brachii, and calf muscles were weak and atrophic (Fig. 1). No fasciculation was observed in those muscles. He did not show any sensory disturbances. Neither deep tendon reflexes nor pathological reflexes were elicited. A nerve conduction study (NCS) was normal. Serum neutralizing antibodies for poliovirus types 1, 2, and 3 were <4, 128x, and <4, respectively. He was thus diagnosed with PPS.

We simply advised him not to exercise too much. He stopped coming to our hospital until he recognized weakness of the right arm four years later. The level of serum creatine kinase was slightly elevated to 317 IU/L. An NCS including the bilateral median, ulnar, radial, tibial, peroneal, and sural nerves did not show any evidence of demyelination. No findings of pleocytosis, increased protein or IgG index values, or oligoclonal IgG band were detected in the CSF, nor were any neutralizing antibodies for poliovirus. MRI of the cervical cord revealed mild spondylosis with canal stenosis. There was no lesion in the spinal cord (Fig. 2). EMG findings were characteristic (Table).

The muscles affected previously (left deltoid and left anterior tibial) showed chronic denervation potentials, such as

Department of Neurology, National Hospital Organization Higashi-Ohmi General Medical Center, Japan
Received: May 17, 2021; Accepted: January 14, 2022; Advance Publication by J-STAGE: March 5, 2022
Correspondence to Dr. Kengo Maeda, maeda@vories.or.jp

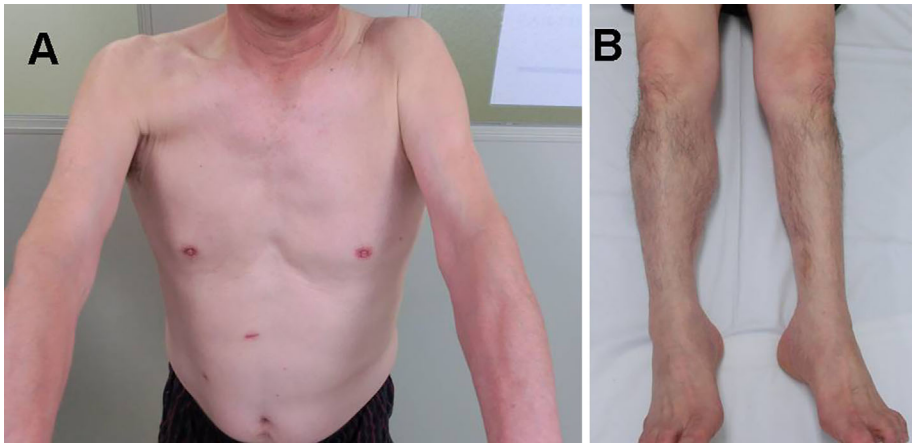

Figure 1. Photographs of the patient at the first visit. Muscular atrophy of the left arm (A) and calf (B) is prominent

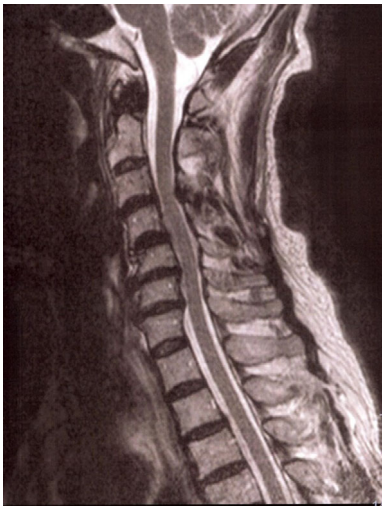

Figure 2. Sagittal view of the cervical spinal cord on T2-weighted MRI.

single oscillation of giant motor unit action potentials without acute denervation potentials. The right biceps brachii muscle, newly affected, showed both acute and chronic denervation potentials. The unaffected muscles (left extensor digitorum communis, left iliopsoas, and right iliopsoas) showed chronic denervation potentials with effective compensation. The unaffected right anterior tibial muscle showed acute and chronic denervation potentials, suggesting that weakness of this muscle might be ongoing.

Discussion

Our patient had a family history of paralytic poliomyelitis affecting his sister and cousin. He has never received the oral poliovirus vaccine (OPV), nor had he ever been to polio-endemic areas. He did not suffer from acute paralytic poliomyelitis. The presence of neutralizing antibody against type 2 poliovirus suggested that he might have been infected, possibly due to close contact with his sister. Based on the clinical features, serological evidence, and EMG findings, we diagnosed him with PPS from non-paralytic polio infection.

However, a careful differential diagnosis is necessary in cases with asymmetrical and/or segmental weakness with muscular atrophy. First, amyotrophic lateral sclerosis (ALS) was excluded based on the fact that the patient's symptoms of weakness and muscular atrophy of his left arm and calf had not progressed for more than 10 years and that some muscles with a normal strength showed chronic denervation potentials without acute denervation potentials. Furthermore, the patient had no symptoms or signs of upper motor neurons. He also did not develop bulbar palsy or respiratory failure. Second, cervical spondylotic amyotrophy known as Keegan's cervical spondylosis should be excluded. This disease affects limited muscles innervated by the locally compressed anterior roots. Widespread abnormalities on an EMG do not indicate this disorder. However, the presence of cervical canal stenosis may influence the patient's symptoms. Motor systems that were pre-injured by a polio infection and decreased in the number of motor neurons might be susceptible to the compression whose power is not strong enough to cause muscle weakness and muscular atrophy in normal condition. Third, multifocal motor neuropathy (MMN) should be excluded. MMN is an immune-mediated demyelinating neuropathy. NCSs including the bilateral median, ulnar, radial, tibial, peroneal, and sural nerves excluded demyelinating neuropathy in the present case based on the absence of decreased nerve conduction velocity or conduction block. Fourth, poliomyelitis itself, not PPS, should be considered. Our patient had never traveled abroad. In Japan, a live vaccine was used for polio vaccination (OPV) until 2012. The OPV is a cause of vaccine-induced poliomyelitis; however, our patient did not receive the OPV. His grandsons had received the OPV one- year before the onset of this patient, but our patient did not have a fever, or other infectious symptoms at that time, and the interval of one- year seems be too long for this to have been a case of vaccine-induced poliomyelitis. Fifth, Hirayama disease affects young people, and the onset age differs markedly from this patient. Al-

Table. **Results of Electromyography.**

Muscles	Resting	Weak contraction	Interference
Previously affected			
L deltoid	F (-), P (-)	giant, 8mV	poor
L anterior tibialis	F (-), P (-)	giant, 9mV	poor
Newly affected			
R biceps brachii	F (-), P (+)	giant, 9mV	poor
Unaffected			
L extensor digitorum communis	F (-), P (-)	giant, 12mV	normal
L iliopsoas	F (-), P (-)	giant, 7mV	fair
R extensor digitorum communis	F (-), P (-)	normal	normal
R iliopsoas	F (-), P (-)	giant, 12mV	poor
R anterior tibialis	F (+), P (-)	giant, 8mV	fair

L: left, R: right, F: fibrillation potential, P: positive wave

though the serum level of CK was increased slightly, muscle diseases, such as myositis and muscular dystrophy, were denied by the EMG findings. Hypothyroidism had been excluded at the previous hospital. Mononeuropathy multiplex induced by vasculitis was not suggested due to the patient's symptom (no sensory symptoms), NCS findings, and blood test results.

The present patient showed a stepwise deterioration. We therefore believe that our patient experienced a deterioration of his symptoms at least three times. When we saw him for the first time (at 69 years old), his left arm and left calf had been already affected. We do not believe that they were affected at the same time. One study followed 52 patients who had previously had poliomyelitis for 4 years. That study detected only small changes in the muscle strength and disability (4). However, some case studies have observed deterioration in different motor innervation segments in PPS (5, 6).

A total of 95% percent of polio-infected patients develop non-paralytic disease, presenting with a flu-like illness; however, in 5% of cases, paralytic polio develops (7). The diagnostic criteria for PPS are 1) prior paralytic poliomyelitis; 2) a period of partial or complete functional recovery after acute poliomyelitis, followed by an interval of stable neurological function; 3) the gradual or sudden onset of progressive and persistent new muscle weakness or abnormal fatigability; 4) symptoms persisting for at least one year; and 5) the exclusion of other neurological, medical, and orthopedic problems (8). Our patient did not meet these criteria since he did not have paralytic poliomyelitis. However, PPS-like weakness or fatigue has been observed in patients with non-paralytic polio (9-13). In a twin- study, 71% of paralytic poliomyelitis patients developed PPS, whereas only 41% of other twins of non-paralytic poliomyelitis patients developed PPS (10). Halstead et al. reported that patients with a typical clinical course, physical findings, and electrophysiological findings without paralytic poliomyelitis should be included in PPS (12). Motor neuron damage in unaffected muscles has been reportedly observed both electrophysiologically (3) and pathologically (14-16). PPS should be considered in patients presenting with segmental muscle weakness or atrophy with an inactive interval.

The authors state that they have no Conflict of Interest (COI).

References

1. Gonzalez H, Olsson T, Borg K. Management of postpolio syndrome. Lancet Neurol **9**: 634-642, 2010.
2. Maeda K, Joko M. Segmental muscular atrophy in a patient with postpolio syndrome. Intern Med **46**: 75-76, 2007.
3. Luciano CA, Sivakumar K, Spector SA, Dalakas MC. Electrophysiologic and histologic studies in clinically inaffected muscles of patients with prior paralytic poliomyelitis. Muscle Nerve **19**: 1413-1420, 1996.
4. Willen C, Hou L, Stibrant Sunnerhagen K. A very long-term longitudinal follow-up of persons with late effects of polio. Eur J Phys Rehabil Med **56**: 155-159, 2020.
5. Oki R, Uchino A, Izumi Y, et al. An autopsy case of progressive generalized muscle atrophy over 14 years due to post-polio syndrome. Rinsho Shinkeigaku (Clin Neurol) **56**: 12-16, 2016.
6. Enghelberg S, Love IY, Rapoport M. Post-polio syndrome causing late onset respiratory failure. IMAJ **22**: 395-396, 2020.
7. Kidd D, Williams AJ, Howard RS. Poliomyelitis. Postgrad Med J **72**: 641-647, 1996.
8. Halstead LS. Assessment and differential diagnosis for post-polio syndrome. Orthopedics **14**: 1209-1217, 1991.
9. Ramlow J, Alexander M, LaPorte R, et al. Epidemiology of the post-polio syndrome. Am J Epidemiol **136**: 769-786, 1992.
10. Nee L, Dambrosia J, Bern R, et al. Post-polio syndrome in twins and their siblings: evidence that post-polio syndrome can develop in patients with nonparalytic polio. Ann NY Acad Sci **753**: 378-380, 1995.
11. Bruno RL. Paralytic vs "nonparalytic" polio: distinction without difference? Am J Phys Med Rehabil **79**: 4-12, 2000.
12. Halstead L, Silver J. Nonparalytic polio and postpolio syndrome. Am J Phys Med Rehabil **79**: 13-18, 2000.
13. Falconer M, Bollenbach E. Late functional loss in nonparalytic polio. Am J Phys Med Rehabil **79**: 19-23, 2000.
14. Kosaka T, Kuroha Y, Tada M, et al. A fatal neuromuscular disease in an adult patient after poliomyelitis in early childhood: consideration of the pathology of post-polio syndrome. Neuropathology **33**: 93-101, 2013.
15. Bodian D, Howe HA. Experimental nonparalytic poliomyelitis: frequency and range of pathological involvement. Bull Johns Hopkins Hosp **76**: 1-10, 1945.

16. Bodian D, Howe HA. Non-paralytic poliomyelitis in the chimpanzee. J Exp Med **81**: 255-274, 1945.