Case Report

Post-polio syndrome presenting as isolated neck extensor myopathy: a case report

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Background: Post-polio syndrome (PPS) is characterized by new or worsening muscular weakness, atrophy, muscle and joint pain, and muscle fatiguability decades after paralytic poliomyelitis infection.

Case Description: A 56-year-old man was diagnosed with paralytic poliomyelitis at the age of five, which left him with flaccid paralysis and weakness of the right leg. One year before seeking chiropractic care, the patient saw his primary care physician with neck pain, low back pain, and fatigue. At the time, he had been diagnosed with degenerative spondylosis and was being treated with tricyclic antidepressants, clonazepam, and tramadol. Despite taking the drugs, his spinal pain and fatigability deteriorated, and he acquired head ptosis during the following six months. As a result, he sought chiropractic care for second opinion. Due to the patient's failure to respond to oral analgesics, radiographs were performed, which revealed degenerative spondylosis, cervical flexion deformity, right pelvic drop, and right thoracolumbar scoliosis. The patient met the PPS diagnostic criteria. PPS related isolated neck extensor myopathy (INEM) was impressed. Multimodal intervention including cervical and lumbar manipulation, spinal traction, micro-vibration deep muscle massage, and core muscle training was provided. As a result of 40-month treatment, the patient reported full resolution of physical complaints. Head posture restored, cervical curvature retrieved and pelvic obliquity relatively corrected.

Conclusions: Survivors of paralytic polio are especially vulnerable to developing leg weakness and length discrepancy, pelvic obliquity, asymmetric axial loading, and trunk muscular imbalance. The current case demonstrates a rare myopathy in a patient at post-polio stage, as well as the restoration of neck function with chiropractic intervention.

Keywords: Chiropractic; head ptosis; isolated neck extensor myopathy (INEM); post-polio syndrome (PPS); case report

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Introduction

Poliomyelitis, or polio, is a contagious disease caused by poliovirus. Approximately 70% of polio infections are asymptomatic (1). Symptomatic cases can be divided into three clinical manifestations: abortive polio (non-specific minor illness), non-paralytic polio (aseptic meningitis without paralysis), and paralytic polio (flaccid paralysis of various muscle groups). Paralytic polio occurs in less than 1% of all infections (1,2). The disease, depending on the level of involvement, is classified as spinal polio (spinal cord), bulbar polio (brainstem), or bulbospinal polio (both). Spinal polio accounts for 79% of paralytic cases and is characterized by asymmetric paralysis that most often involves the legs. The paralysis is usually permanent, but some recovery may occur through compensation from the unaffected neighboring nerve cells and muscles (1).

Polio survivors may experience a constellation of symptoms post-recovery, called post-polio syndrome (PPS) (2). PPS was first coined by Halstead in 1986, who presented criteria for the diagnosis of PPS, including new or deterioration of muscular weakness and atrophy, fatigability, muscular and joint pain, and cold intolerance several years after acute paralytic polio infection (3). All these latent sequelae of polio are the result of the damage incurred by initial infection, prolonged stress on musculoskeletal structures, and increased metabolic demand of surviving motor-neurons (4,5). Head ptosis (head drop or dropped head syndrome) is a rare condition due to the weakening of neck extensors to hold the head in an upright position. Isolated neck extensor myopathy (INEM) is a specific type of head ptosis caused by non-inflammatory myopathy restricted to the cervical extensor muscles (6).

Wild poliovirus has been eradicated in the developed countries by the introduction of the Salk (in 1955) and Sabin (in 1962) inactivated poliovirus vaccines (1). As of the year 2022, endemic circulation of wild poliovirus persists only in Afghanistan, Pakistan, and poor African countries where vaccination is difficult (7). About 15% to 80% of the polio survivors will get a deterioration of symptoms, effective treatment for the neuromuscular sequelae, especially core muscle weakening, in the 20 million global patients of paralytic polio is desperately needed (8). There is a paucity of data describing or reporting on the outcomes of conservative treatment for PPS and head ptosis. The current study highlights the potential effectiveness of a chiropractic approach in addressing head ptosis and posture problems in a patient at a post-polio stage. We present the following case in accordance with the CARE reporting checklist (available at https://acr.amegroups.com/article/view/10.21037/acr-22-76/rc).

Highlight box

Key findings

- Isolated neck extensor myopathy presenting as a part of the post-polio syndrome is a likely but rarely documented occurrence.

What is known and what is new?

- Following recovery from polio, survivors may experience a constellation of symptoms caused by prolonged stress on musculoskeletal structures and increased metabolic demand on surviving motor neurons.
- This case demonstrates that truncal hypertonia can occur, but it is not always recognized.

What is the implication, and what should change now?

- A multimodal intervention focused on core muscle training, cervical and lumbar manipulation, and avoiding overuse of the weak muscles significantly improved neuromuscular symptoms, global posture, and spinal function.

Case presentation

A 56-year-old Asian businessman reported that he got paralytic poliomyelitis when he was 5 years old, which subsequently resulted in right lower limb flaccid paralysis. The patient had made a partial recovery and kept his neuromuscular functioning largely steady for the next five decades after the infection. He has been able to walk ever since with the aid of a cane and a foot orthotic. However, he started experiencing deep ache in the nuchal and low back regions, and general fatigue one year before first seek for chiropractic care. The patient denied any previous injuries. His primary care physician radiographically diagnosed him with degenerative cervical and lumbar spondylosis at the time. Tricyclic antidepressants, clonazepam, and tramadol were prescribed for pain relief on an as-needed basis. Despite taking the aforementioned medications, the neck and back pain and fatigue got worse, and the difficulty to hold the head up progressed over the course of the following six months. The spinal pain and head ptosis had severely limited his activities of daily living more than before, and forced him to seek further treatment.

The patient presented with a dropped head posture and was barely able to hold his head up for 5 minutes. The head ptosis was correctable in the supine position. He ambulated with a cane with a short step length and was accompanied

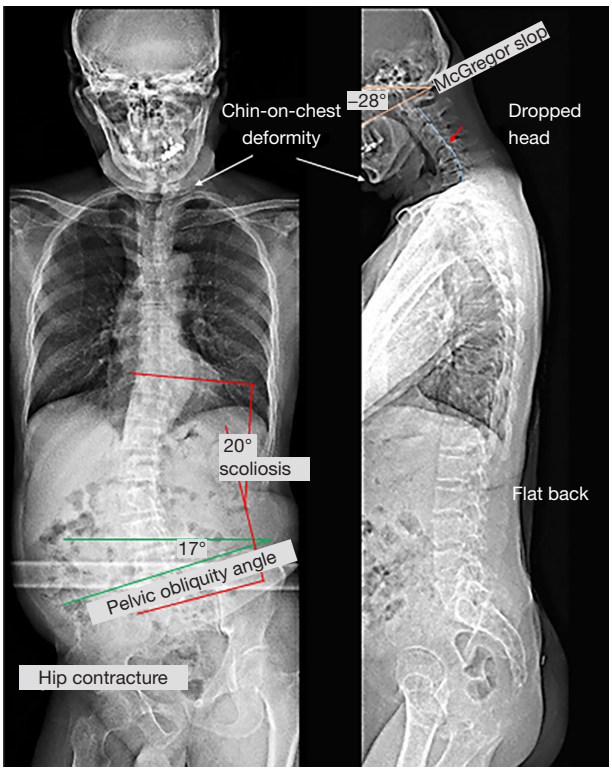

Figure 1 At initial presentation, coronal radiograph shows right pelvic drop, right hip contracture with abduction and a right thoracolumbar curvature with a 20° Cobb angle between T10 and L5. Lateral view reveals reverse cervical lordosis with anteroflexion deformity (white arrow), –28° McGregor slop, mild subluxation of C4 relative to C5 (red arrow), osteophytes in the lower cervical and lumbar spine, and interspace narrowing of the C3/C4, C5/6, C6/C7, C7/T1, and L4/5 levels. McGregor's line is a hypothetical line drawn between the hard palate and the most caudal point of the occipital curve.

by a foot drop on the right side. In addition, the patient had atrophy on the right leg. Palpation revealed hypertonicity of the bilateral upper trapezius, rhomboids, and quadratus lumborum muscles, and intersegmental motion restrictions were found from C5–7, T3–8, and L2–5. The intensity of the neck and low back pain was rated at 5–6/10 on a 1–10 numeric pain rating scale, where 0 means no pain and 10 means the worst imaginable pain. His range of motion in the cervical region was recorded at 90° of flexion, 30° of extension (normal 50°–70°), 10° of lateral flexion (normal 20°–45°), and 90° of rotation to both sides.

The manual strength grade of the right lower limb was 2/5. The deep tendon reflex test, using a reflex hammer, was conducted by a chiropractor, showing hyporeflexia of the right knee-jerk and ankle-jerk. The neurological findings of the left leg were normal. Standing full-spine EOS® radiographs showed reverse cervical lordosis with anteroflexion deformity, osteophytes in the lower cervical and lumbar spine, and interspace narrowing of the C3/C4, C5/6, C6/C7, C7/T1, and L4/5 levels, suggestive of degenerative spondylosis. A coronal view revealed right pelvic drop, right hip contracture with abduction, and a long thoracolumbar curve (T_{10}–L_5 Cobb angle of 20°) with a right convexity as compensatory changes for pelvic obliquity (*Figure 1*). The World Health Organization Quality of Life (WHOQOL-OLD) (9) scoring was used, and the results were linearly translated to a 0–100 scale, with higher scores indicating better quality of life. The patient received a score of 48. Static surface electromyography (sEMG) was conducted using MyoVision® 4000 (Precision Biometrics/MyoVision, San Carlos, CA, USA) revealed intense myoelectric activity of bilateral paraspinal muscles throughout the spine (*Figure 2*). This apparatus has been shown to be reliable among both healthy and neck-pain individuals (10). The patient met the diagnostic criteria for PPS (2). PPS related isolated neck extensor myopathy (INEM) was impressed.

The initial phase of chiropractic care was aimed at relieving pain and improving core muscle functions. During each visit, the intervention included: (I) 15-minute mechanical cervical traction to reduce the pressure on the nerves; (II) 8-minute cervical and lumbar manual therapy to improve mobility; (III) 8-minute deep muscle massage using a handheld micro-vibrator (Strig®, STRIG Inc., Korea) to relax the hypertonic muscles; and (IV) 15-minute isometric core trunk muscle training (11) to strengthen the paraspinal muscles and improve spine posture. The patient was seated in a commercially robotic chair (AllCore360° Core Training System®, AllCore360, GA, USA) for the workout. The device is designed to allow for increasing the strength of the core muscles by progressively contracting the trunk muscles while the chair slowly rotates at various angles and the patient keeps his spine in a neutral position (*Figure 3*).

Initial treatment sessions were scheduled 3 times a week for the first month, focusing on pain symptoms. Then, the frequency of treatment was reduced to 2 times a week for 2 months, focusing on relieving pain and improving core muscle functions. Afterwards, the patient was seen once a week for 10 months, focusing on correcting both pelvic and spinal postures. The frequency of treatment was further reduced to once a month for 30 months,

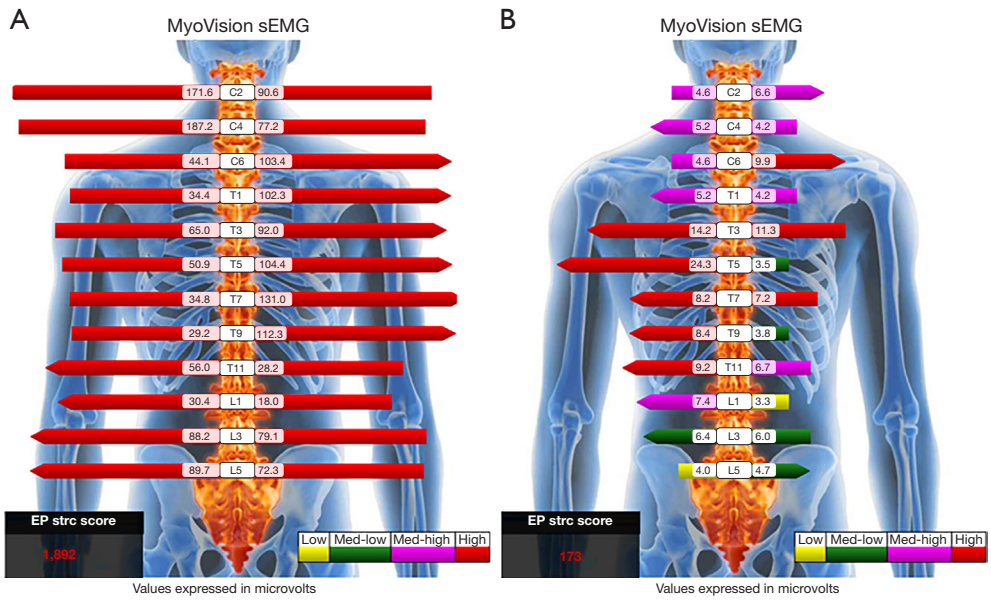

Figure 2 Comparison of pre- and post-intervention sEMG. Handheld electrodes are used to measure paraspinal muscle groups. In a schematic representation, myoelectric activity at each paraspinal level is recorded and represented by different colored bars. EP stress score is the sum of all muscle activity on the spine measured in microvolts. (A) sEMG revealed substantial levels of muscular tension at all spinal segments during the initial presentation. Intense neck activity indicated the significant effort required in the nuchal muscle to prevent the head (chin) from falling onto the chest. (B) The sEMG signal of most muscle groups normalized and was favorably linked with symptom alleviation following 40 months of treatment. The less myoelectric activity in the lumbar region is due to asymmetric paralysis of the quadratus lumborum following initial polio infection (2). sEMG, surface electromyography; EP, electrophysiological.

focusing on monitoring the symptoms, spinal adjustment, and core muscle training. A home stretching program (muscle strengthening, mobility training, and ergonomic recommendations) was also used as an adjunct to care. When training with exercise programs, the standard advice is to avoid exercising too hard and to train at submaximal but progressively increasing levels.

The patient reported that his numeric pain scale was reduced from 6 to 2 on the 1–10 scale after a few visits. His neck mobility was nearly normal and his pain and muscle weakness were in complete remission after four weeks of treatment. His WHOQOL score increased from 48% to 78% at 11 months, indicating improvement in the quality of life. Treatment was continued until either the patient's improvement plateaued or the maximal improvement was achieved. Forty months after the treatment initiation, follow-up radiographs and sEMG were taken. Comparing to pre-intervention sEMG, paraspinal myoelectric activity returned to normal after intervention at the majority of spinal levels (*Figure 2*). Repeated radiographs revealed improved head posture, relatively restored normal pelvic obliquity, and lumbar scoliosis reduced to a T_{10}–L_5 Cobb angle of 16° (*Figure 4*). No pathological or treatment-related adverse effects were observed.

All procedures performed in this study were in accordance with the ethical standards of the institutional and national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

Discussion

Acute polio is no longer a persistent threat to people in the polio-free areas of the world, but there are an estimated 20 million polio survivors with PPS globally (12,13). The etiology and pathogenesis of PPS are unknown. Survivors frequently recover only partially from their paralysis. It has been proposed that a certain amount of recovery is due to the ability of unaffected neighboring nerve cells to "sprout"

Figure 3 Exemplification of a core strengthening workout. The demonstrator is placed in the robotic device (AllCore360° Core Training System®, AllCore360°, GA, USA), which slowly rotates at varied angles of inclination. At the same time, the demonstrator contracts the core muscles isometrically to maintain a neutral, seated posture while clutching a big ring in the hands and a ball between the knees. The device and its usage are depicted in this exercise demonstration.

and reconnect to the paralyzed muscles. The gradual and latent deterioration of muscle weakness is related to years of overuse of the compensatory motor units that can no longer respond properly, unveiling the neurological impairment caused by the initial illness (13,14). The European Federation of Neurological Societies (EFNS) criteria for the diagnosis of PPS (2,3) are as follows: (I) previous episode of paralytic polio; (II) rehabilitation followed by over 15 years of steady neuromuscular function; (III) new or worsening muscle weakness; (IV) symptoms last at least a year; (V) exclusion of other possible causes of symptoms. Common symptoms include muscle weakness, fatigability, and muscle and joint pain, which are hypothesized to be caused by muscle overuse, leg length discrepancy, asymmetric biomechanics, asymmetric loading on the axial spine, and age-related changes (12). PPS symptoms and functional impairments can have an influence on quality of life and capacity to accomplish everyday duties.

Head ptosis is a rare disorder characterized by the inability to extend the neck due to the weakening of the neck extensor muscles, resulting in a chin-on-chest deformity (6,15). It can be painful, disfiguring cosmetically, impeding swallowing and impairing forward gaze. The pathophysiology of ptosis of the head remains unclear. It might be the presenting symptoms of a vast array of metabolic, dystrophic, myopathic, or neurological disorders (16). INEM is one of the probable causes of head ptosis due to isolated myopathic changes resulting from chronic injury and overloading of the cervical extensor muscles (6). There is a paucity of documented cases reporting a link between head ptosis and PPS. Our interpretation of this association is that polio left our patient with right leg weakness, a length discrepancy, and pelvic obliquity, which led to asymmetric axial loading, scoliosis, and imbalanced core muscles. Overuse and fatigue of the cervical paraspinal muscles (neck extensors) contributed to the inability of the patient to maintain an erect head position. In addition, the scoliotic curvature exerts an additional asymmetric strain on the neck muscles, resulting in INEM. Initial sEMG findings of intense myoelectric activity were consistent with the significant effort require by the cervical paraspinal muscles to prevent the head from dropping onto the chest. A decrease in lumbar myoelectric activity was noted when compared with sEMG before and 40 months after intervention (*Figure 2*). This was attributed to the patient's asymmetric quadratus lumborum paralysis following the initial polio infection.

There are currently no established regimens that can prevent the deterioration of PPS. Generally, the overall goal of the management of PPS is symptomatic, aiming to reduce pain and improve function by minimizing the excessive metabolic stress on the muscles (17). Evidence suggests that in order to treat new weakness in patients with PPS, supervised training programs are required (18). These programs focus on boosting muscle strength through non-fatiguing exercises that involve using submaximal or maximal strength with short-duration repetitions on alternate days (2,18). Such program yielded favorable results, by facilitating a full recovery from the exercises within the constraints of fatigue and pain (19) and preventing the overuse of the weak muscle (17). On follow-up sEMG (*Figure 2*) and radiographs (*Figure 4*), relaxation of paraspinal muscle spasm, improvement of spinal alignment, and restoration of normal pelvic obliquity may have contributed to enhanced spinal functional capacity of the current case. This case illustrates

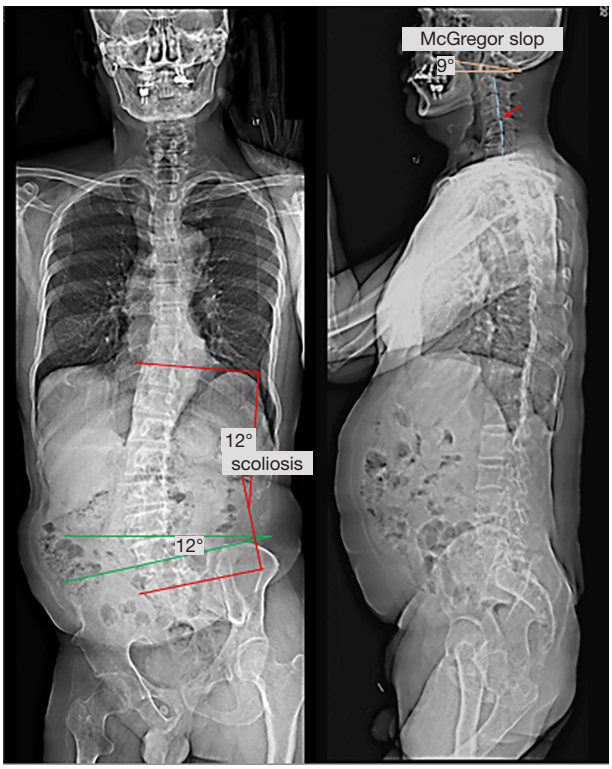

Figure 4 Comparing the result of 40-month treatment in the same patient shown in *Figure 1*. Repeat radiographs taken 40 months after the first one show patient's head ptosis has been corrected, the McGregor slop returns to 9°, and the majority of other radiographic parameters have improved. The coronal Cobb angle of the main thoracolumbar curve is 16° and pelvic obliquity angle is 12°.

that hypertonic muscle may be present, a fact that is not always acknowledged.

This study has potential limitations. First, due to the rarity of both PPS and INEM, this was a single case study and was not controlled. Second, PPS was basically diagnosed on a medical history and the exclusion of other conditions that might explain the symptoms. Electromyography and magnetic resonance imaging for a thorough investigation of soft tissue alteration in the neck had not been taken. Furthermore, the precise effectiveness of chiropractic intervention could not be clarified by the multimodal treatment strategy. The mechanisms of symptom relief and posture retrieval warrant further scrutiny. The strength of this work is that it provides information indicating that the occurrence of INEM decades after paralytic polio infection is a probable but rarely documented event.

Conclusions

The current study was the first to report INEM among patients with PPS. The suggested multimodal intervention with long-term spinal manual therapy has shown significant improvement in neuromuscular symptoms, the head and global posture, and spinal function.

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Footnote

Reporting Checklist: The authors have completed the CARE reporting checklist. Available at https://acr.amegroups.com/article/view/10.21037/acr-22-76/rc

Conflicts of Interest: Both authors have completed the ICMJE uniform disclosure form (available at https://acr.amegroups.com/article/view/10.21037/acr-22-76/coif). The authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

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