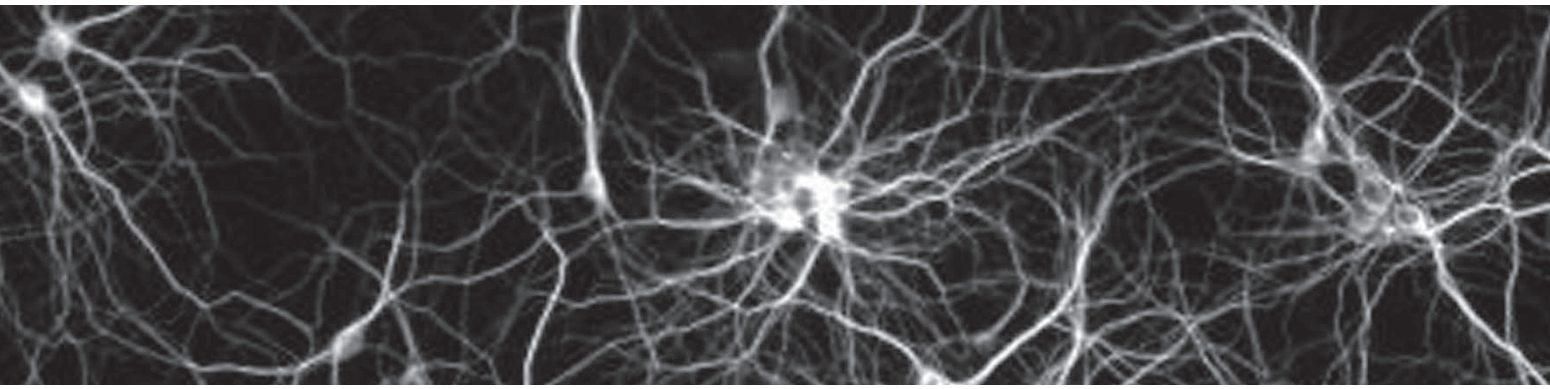

Parsonage-Turner syndrome

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ABSTRACT – *Objective:* Here we wanted to demonstrate the importance of having a broad differential diagnosis to avoid misdiagnosis in patients suffering from Parsonage-Turner syndrome (PTS). *Case description:* PTS is a rare idiopathic neuritis of the brachial plexus, characterized by sudden onset of pain in the shoulder and arm, followed by progressive weakness and muscle atrophy of the affected area. We present a case of a Caucasian 8-year-old boy from Croatia with acute onset of shoulder pain after viral infection recovery. The boy was treated with methylprednisolone and several courses of intravenous immune globulin. Over time, the patient developed necrosis of the right acetabulum, and corrective osteotomy was performed. At the age of 19, a neurological exam showed severe atrophy of the right shoulder muscles. *Results:* The diagnostic delay in our case was three years, showing the lack of specific tests when diagnosing PTS. Even though the patient received immunoglobulin and corticosteroid treatment, atrophy of the right shoulder muscles occurred, and his motor skills in the right arm were reduced. *Conclusion:* Since clinical presentation can reflect other pathological conditions, accurate diagnosis is difficult and oftentimes delayed. Our case illustrates the importance of including broad differential diagnosis in patients with shoulder pain for prompt initiation of an accurate therapy.

Keywords: Brachial plexus, neuralgic amyotrophy, Parsonage-Turner syndrome

INTRODUCTION

Parsonage-Turner syndrome (PTS), also known as neuralgic amyotrophy or brachial plexitis, is a rare neurological disorder first described by Dreschfeld in 1887. Multiple reports followed to describe the condition with Parsonage and Turner (1948) being the most important ones (1). The syndrome can vary greatly in presentation and nerve involvement. It is usually characterized by the sudden onset of acute severe pain in the upper arm and shoulder girdle followed by progressive weakness leading to flaccid paralysis and muscle atrophy of the affected area. It is usually unilateral, affecting the

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dominant side, although it can appear on both sides of the body. Even though the etiology of the idiopathic form of the disease is not yet fully understood, it is believed that most cases are due to an autoimmune response to an infection (i.e., smallpox, influenza, coxsackievirus, cytomegalovirus (CMV), human immunodeficiency virus, SARS-CoV-2 virus) (2, 3). Vaccination, surgery, pregnancy, certain medical procedures, or strenuous exercise can also lead to the development of PTS (4-7). The other, hereditary form is an autosomal dominant inherited neuralgic amyotrophy usually caused by mutations in the SEPT9 gene on chromosome 17 (8).

The estimated incidence of PTS is 1.64 per 100,000 people per year (4), mainly affecting the male population aged 20 to 60 years. Since clinical presentation can reflect other pathological conditions, accurate diagnosis is difficult and oftentimes delayed (9). The diagnostic workflow includes nerve conduction studies (NCS), electromyography (EMG), magnetic resonance imaging (MRI), and/or X-ray. Treatment is symptomatic and includes analgesics, corticosteroids, and physiotherapy. The estimated recovery rate is 75% to 90% within 2 and 3 years from symptom onset, respectively (4, 10).

CASE REPORT

An 8-year-old boy was admitted to the hospital for a tingling sensation in his right forearm and three weeks of constant pain in the right upper arm. The pain occurred after an episode of pharyngitis treated with amoxicillin and clavulanic acid. Neurological examination revealed impaired convergence of the left eye, brisk tendon reflexes of the right extremities, and a positive Oppenheim and Chaddock signs on the right site. All other aspects of the neurologic exam were normal. Laboratory workup results (sedimentation rate, complete blood count, renal and liver function tests, coagulation tests, antinuclear antibodies, rheumatoid factor, serum protein electrophoresis, copper levels, and antiganglioside antibodies) were within normal range. Cerebrospinal fluid (CSF) analysis was also within normal values. *Borrelia burgdorferi*, CMV, *Toxoplasma gondii,* and *Bartonella* serology tests were negative, while the Epstein-Barr virus test (EBV) showed positive IgG and negative IgM titers. Other microbiological findings were negative. Molecular genetic analysis for hereditary neuropathies (CMT1A/HNPP) was negative and no duplication or deletion in the PMP22 gene was found. The HLA-B27 test was also negative. EMG indicated a moderate loss of motor neurons in the foot and reinnervation potentials in the right deltoid muscle. NCS showed reduced compound muscle action potentials (CMAPs) and prolonged distal latency bilaterally in the right axillary and peroneal nerve with normal motor velocity. X-ray imaging of the sacroiliac joints showed bilateral sclerosis. Skeletal scintigraphy was normal. MRI of the brain and cervical spine showed no abnormalities. Con-

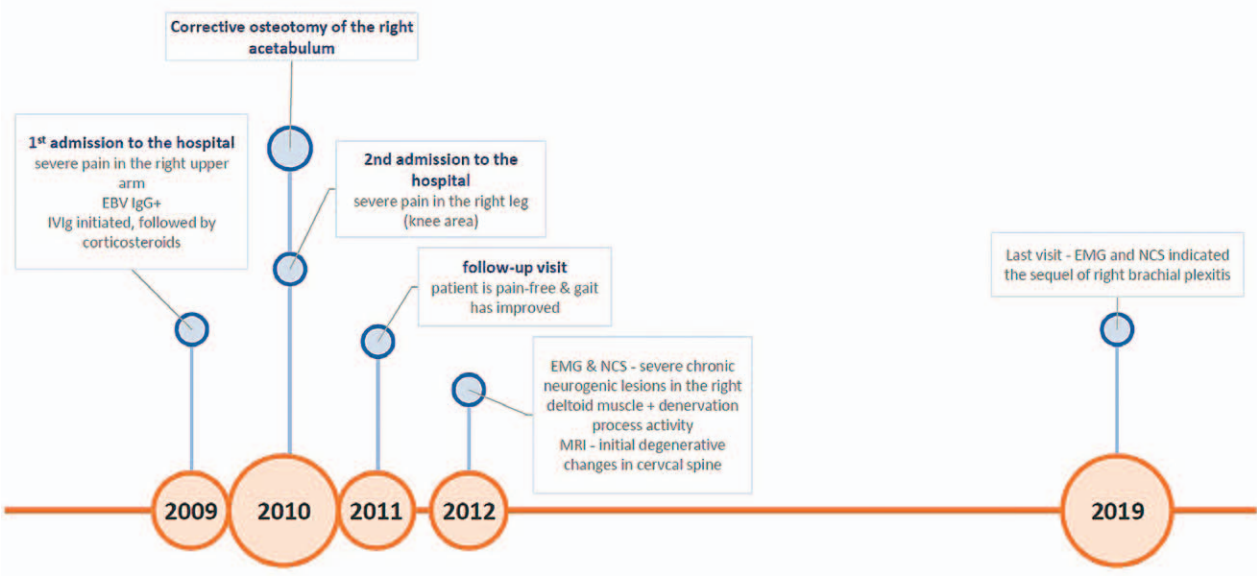

Fig. 1. *Timeline chart showing key points of the Parsonage-Turner Syndrome Case Report (EBV – Epstein-Barr virus; EMG – electromyography; IVIg – intravenous immune globulin; MRI – magnetic resonance imaging; NCS – nerve conduction study). Bubble Chart Timeline Template © 2017 by Vertex42.com*

trast computed tomography (CT) of the base of the neck, chest, abdomen, and pelvis indicated enlarged lymph nodes in the axillary and inguinal regions, with no other pathological changes. An infectious disease specialist was consulted and found no signs of acute infectious disease; the workup indicated only a past EBV infection. The patient was initially admitted under the diagnosis of an atypical form of Guillain-Barré syndrome, due to the pain and tingling sensation in his right arm. Therapy with intravenous immune globulin (IVIg) was initiated at a dose of 0.4 g/kg for 5 days and was continued with methylprednisolone orally at a dose of 64 mg daily with dose tapering. A year later the patient was readmitted to the hospital with a neurological exam showing severe atrophy of the right shoulder muscles and limited abduction and anteflexion in the right shoulder joint. The patient was given a second course of IVIg (0.4 g/kg) and was discharged with steroid therapy.

Three months later, he was readmitted to the hospital with complaints of severe pain in the right leg (knee area). The third course of IVIg was administered (0.4 g/kg), followed by the fourth course a month later. The patient was taking methylprednisolone orally until the age of 10 when it was stopped due to persistent pain in the right hip. X-ray diagnostics revealed aseptic necrosis of the right hip, and corrective osteotomy of the right acetabulum has been performed. At the age of 12, the patient's EMG and NCS findings showed a severe chronic neurogenic lesion in the right deltoid muscle, with signs of denervation, distal conductive block, slowed conduction velocity in the right axillary nerve, and proximal conductive block in the right peroneal nerve with prolonged distal latency on the right peroneal nerve. MRI of the cervical spine showed initial degenerative changes of C2-C3 and C3-C4 intervertebral discs without signs of disc herniation and with no signal changes in the spinal cord. MRI of the right brachial plexus showed hyperintensity and irregular thickening of lateral and posterior branches, a finding that may correspond to chronic inflammatory changes of nerve tissue. Also, muscle atrophy of the right shoulder girdle was noted. The patient continued with physical therapy and visited a pediatric neurologist on a regular 6-month basis. Figure 1 presents a timeline chart of key points in the boy's treatment.

During the last visit at age 19, EMG and NCS indicated the sequel of right brachial plexitis. A neurological exam showed severe atrophy of the right shoulder muscles (Fig. 2, 3), diminished deep tendon reflexes in the right upper extremity, and very limited abduction (to about 40 degrees) of the right arm in the shoulder.

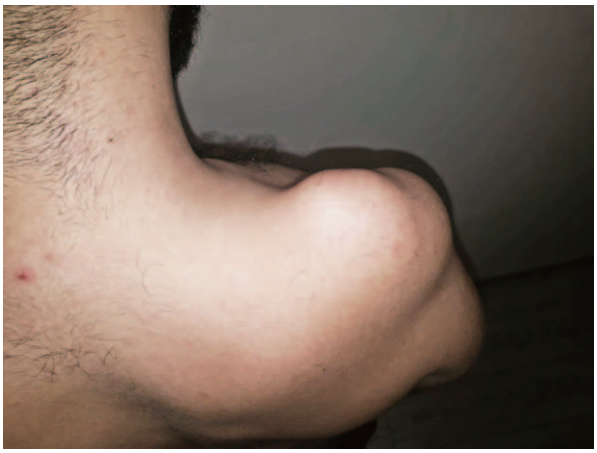

Fig. 2. *Deltoid atrophy*

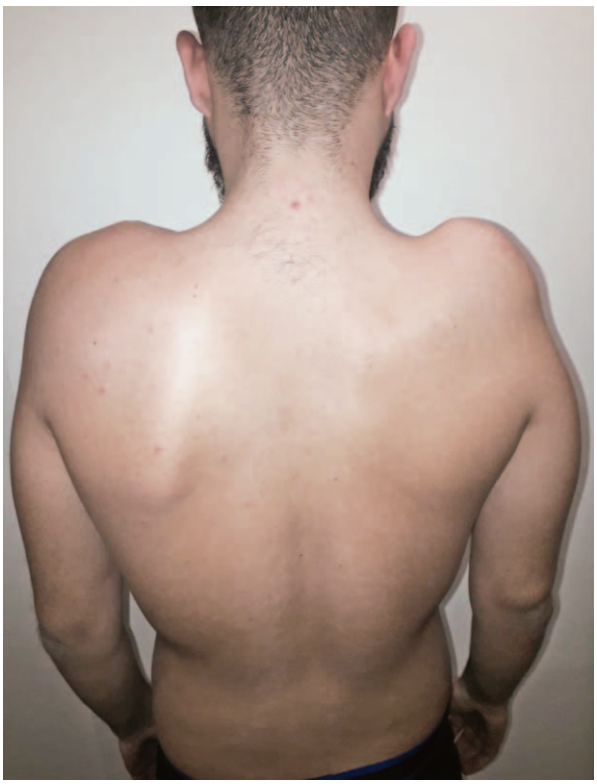

Fig. 3. *Supraspinatus and infraspinatus atrophy*

DISCUSSION

PTS is predominantly a clinical diagnosis with the classic presentation of abrupt onset, intense, unilateral shoulder girdle pain, often nocturnal at onset, with subsequent and progressive muscle weakness and varying levels of sensory abnormalities. Although the pathophysiology is not completely under-

stood, it is likely immune-mediated as there is a high reported incidence (up to 50%) of preceding infections (11). A pathognomonic characteristic of PTS is that pain, motor weakness, and sensory symptoms usually do not affect the same nerve distributions and that the passive range of motion is preserved. These characteristics are especially important when differentiating PTS from cervical radiculopathy and glenohumeral bursitis, which are the most common incorrect initial diagnoses (12). Although there are no specific tests to diagnose PTS, EMG, NCS and MRI scan (13, 14) can help in making the correct diagnosis. The diagnostic delay in our case was three years, showing that there is no specific test when diagnosing PTS. EMG and NCS are often decisive in making a diagnosis. In the case of shoulder pain, a broad differential diagnosis is needed to avoid misdiagnosis and treatment (15, 16).

In the case shown, pain in the right shoulder occurred after treatment of pharyngitis, indicating a preceding infection most likely of viral etiology given the positive EBV IgG titer with the finding of enlarged submandibular lymph nodes and hypertrophic tonsils. EMG indicated a moderate loss of motor neurons in the foot and a severe chronic neurogenic lesion with reinnervation potentials in the right deltoid muscle. NCS showed a lower amplitude of CMAP and prolonged distal latency in the right axillary nerve and peroneal nerve bilaterally with normal motor velocity. The patient was treated with four courses of IVIg and then continued with corticosteroids orally, for approximately one year. The steroids were discontinued due to avascular necrosis of the right hip, which was most likely a steroid therapy side effect (17).

After 12 years from the onset of the disease, complete loss of right shoulder muscles, and reduced motor skills of the right arm, the patient carries out daily life activities without significant difficulties.

CONCLUSION

Our case illustrates the importance of including a broad differential diagnostics workflow in patients with shoulder pain. PTS is an underrecognized cause of acute onset unilateral shoulder pain and paresis, often preceded by an inciting event. PTS is a medical condition that currently has no known cure. However, the symptoms of PTS can be alleviated with the use of oral steroids and physical therapy. Oral steroids have been found to be more effective when administered during the early stages of the condition, which highlights the importance of early diagnosis. If these measures fail to bring relief, surgery may be considered for patients with PTS who do not recover. A high index of suspicion should be maintained as prompt identification and management may decrease the high patient morbidity associated with this diagnosis.

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