

Neuropathic arthropathy of the shoulder associated with chronic inflammatory demyelinating polyneuropathy: A case report

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Abstract

Neuropathic arthropathy is a chronic degenerative arthropathy that results from a neurosensory deficit of the affected joint. According to neurovascular theory, the loss of sensation of the joint is responsible for its formation. Chronic inflammatory demyelination polyneuropathy (CIDP) is a rare disease with sensorimotor involvement in which demyelination and axonal damage occur in the peripheral nerves as a result of an abnormal immune response. A case is presented of a 64-year-old male patient with a diagnosis of CIDP who had Charcot arthropathy of the right shoulder.

Keywords: Charcot arthropathy, shoulder, polyneuropathy

Introduction

Neuropathic arthropathy or Charcot joint is a form of chronic degenerative arthropathy that results from neurosensory deficits of the involved joint. Although upper extremity involvement is rare in neuropathic arthropathy, the shoulder joint is usually affected [1]. Shoulder involvement (often called Charcot shoulder) has been reported in 5-6% of all neuropathic arthropathies [2]. Neuropathic arthropathy is often caused by syringomyelia, but other causes include diabetes mellitus, tabes dorsalis, leprosy, peripheral neuropathy, multiple sclerosis, chronic alcoholism, intra-articular steroid injections, gigantism, end-stage renal disease, congenital insensitivity to pain, amyloidosis, meningomyelocele, and myelodysplasia [3,4].

Chronic inflammatory demyelinating polyneuropathy (CIDP) is a rare disease in which an abnormal immune response causes demyelination and axonal damage of peripheral nerves. It characteristically shows slow progressive symmetrical sensorimotor involvement. The exact etiology of CIDP is unknown [5]. Patients with CIDP experience progressive weakness, sensory and motor dysfunction in the legs and arms, decreased deep tendon reflexes (areflexia), and fatigue [6]. The most accepted diagnostic criteria in recent clinical studies are based on clinical history, examination, electrodiagnostic criteria, and exclusion of other causes [7].

Although chronic fatigue, depression, sleep disorders, and respiratory failure related to CIDP have been reported in the literature, no development of neuropathic arthropathy has been found [8]. This report presents a case with neuropathic arthropathy in the shoulder joint due to CIDP. This case is important because it is the first report of Charcot shoulder to develop due to CIDP in the literature.

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Informed Consent

The authors stated that the written consent was obtained from the patient presented with images in the study.

Conflict of Interest

No conflict of interest was declared by the authors.

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Case presentation

A 69-year-old male patient was admitted to our clinic due to difficulty in walking, loss of strength in his arms and legs, neuropathic pain, and swelling in the right shoulder that had recurred intermittently since 1 month prior, and restricted movement. The patient was diagnosed with CIDP in 2016. He had a history of coronary artery disease and myocardial infarction. The patient's right shoulder developed bruising and swelling for the first time in January 2019 without pain or trauma. When he visited the orthopedic clinic with this complaint, X-rays indicated that he had a dislocated shoulder (Figure 1 and 2).

The patient had used a shoulder splint for a while and had experienced swelling, bruising, and redness in the right shoulder from time to time without pain, even after the splint was removed. Due to these complaints, the patient visited the orthopedic clinic in February 2021. No malignant cells were found in the histochemical staining of a fluid sample from the right shoulder-joint. Blood analysis was performed, and PET-CT images were taken for a paraneoplastic study. The results showed no pathology suggestive of neoplasm.

Magnetic resonance imaging (MRI) of the shoulder showed humeral head dislocation, areas with significant destruction, and complete rupture of the rotator cuff tendons. A preliminary diagnosis of Charcot arthropathy was considered by the radiologist. Cerebral cervical, thoracic, and lumbar MRIs were performed in February 2021 to examine the etiology of Charcot shoulder but revealed no syrinx cavity, discopathy, or spondylosis findings. The patient was referred to the rheumatology clinic in January 2022 for inflammatory arthritis and vasculitis, but no pathology was found in this regard. The results of a Brucella tube agglutination test were also negative.

In the examination of the patient, the muscle strength was 2/5 in the right shoulder group globally, 2/5 in the hip extensor and abductor, 3/5 in the bilateral hip rotators, 3/5 in the bilateral knee flexors, and 3/5 in the toe group. The strength in other muscles was 4/5. His right shoulder flexion was 80°, abduction was 75°, and internal and external rotations were limited. The patient's deep sense was decreased in all 4 extremities. The patient described bilateral hypoesthesia from the C4 dermatome area. The deep tendon reflex (DTR) was hypoactive in both upper extremities, but DTR could not be obtained in the bilateral lower extremities.

During the patient's hospitalization in our clinic, his blood glucose, biochemistry, hemogram, and acute-phase reactants were normal. The infectious diseases department was consulted for evaluation for infectious causes of the patient's Charcot shoulder, such as tabes dorsalis or Lyme disease). The patient also consulted the neurology department to undergo lumbar puncture (LP), and electromyography (EMG) was planned.

ARB, Gram staining, direct microscopy, cerebrospinal fluid (CSF) biochemistry, syphilis VDRL, and Lyme PCR analyses were performed. No pathology was detected in the results. The anti-HIV test came back negative. Infectious pathology was not considered according to the anamnesis and examination results of the patient. The results were reexamined by the infectious diseases department. EMG showed axonal type

sensorimotor polyneuropathy with prominent diffuse sensory and motor involvement in the lower extremity, and the result supported the diagnosis of CIDP. A physical therapy program including electrotherapy and strengthening exercises was provided to the patient. Information was given about protective measures for the shoulder joint. Informed consent form was obtained from the patient for presentation.

Figure 1: X-ray image of the shoulder



Figure 2: X-ray image of the shoulder



Discussion

Neuropathic arthropathy of the shoulder is rarely seen, and the etiology can include peripheral neuropathies that result from pathologies such as syringomyelia, syphilis, diabetes, chronic alcoholism, and leprosy [9]. The neuropathic joint undergoes 3 phases. In the destructive phase, the joint is hyperemic and swollen, and there is osteoclastic bone resorption associated with repetitive trauma. The reparative phase continues with the formation of dense fibrous tissue and coalescence of debris. Finally, the silent phase is characterized by decreased vascularity and bone sclerosis [10].

The pathogenesis is not fully understood, but two different theories have been proposed: neurovascular and

neurotraumatic theories. According to the neurovascular theory, loss of sensation at the joint level impairs normal neurovascular reflexes. This results in osteoclast activation, which causes hyperemia and bone resorption. According to the neurotraumatic theory, the loss of somatic muscle reflexes that maintain the range of motion of the joint causes repetitive traumas. As a result, joint destruction occurs [4,11].

The most common initial symptoms of neuropathic shoulder arthropathy are swelling, redness, decreased range of motion, and loss of function. Considering the symptoms, septic arthritis, synovial chondromatosis, idiopathic osteolysis, tumoral calcinosis, and neoplasia (soft tissue sarcoma) should also be considered in the differential diagnosis [12,13]. In the present case, shoulder joint biopsy had been performed previously in the orthopedic clinic to aid in differential diagnosis, and no neoplasm or synovial chondromatosis was detected.

Radiological findings in neuropathic arthropathy include sclerosis, joint destruction, new bone formation, bone fragmentation, subluxation, osteophytes, osseous debris, effusion, heterotopic ossification, and periarticular soft-tissue swelling in the involved joint [14]. Radiographically specific findings for Charcot shoulder include superomedial flattening of the humeral head, periarticular soft tissue calcification, and glenoid sclerosis [15]. In the present case, radiological findings were consistent with neuropathic arthropathy in the shoulder X-ray. Despite the significant joint destruction, the absence of pain and sensory deficit in neurological examination, normal follow-up of acute phase reactants and white blood cell count, advanced stage destructive arthropathy findings in radiological imaging, and sensorimotor polyneuropathy detected in EMG were compatible with neuropathic arthropathy.

Cervical syringomyelia is the cause of 75% of neuropathic arthropathies in the shoulder [13]. Cases have been reported in which Charcot shoulder developed due to diabetes, chronic alcoholism, intra-articular steroid injection, and congenital painlessness syndrome [4]. In the present case, no syrinx cavity was found in cervical MRI, and there was no history of diabetes, alcoholism, or steroid injections. The literature showed no case of neuropathic arthropathy of the shoulder developing with CIDP etiology as in our case.

Physical therapy, effusion aspiration, nonsteroidal anti-inflammatory drugs, immobilization, and a shoulder sling can be applied as conservative treatments for neuropathic arthropathy of the shoulder [9,12]. If there is no response to conservative treatments, shoulder hemiarthroplasty, total shoulder arthroplasty, glenohumeral arthrodesis, arthroscopic irrigation, and debridement can be applied as surgical treatments [12]. In our clinic, electrotherapy, exercises to protect the range of motion, and isometric strengthening exercises were used to treat the patient's shoulder.

Conclusion

Sensory nerve damage in CIDP may predispose patients to the development of neuropathic arthropathy. The reason is that sensory deficit in the related joint causes the development of neuropathic arthropathy by disrupting the neurovascular reflex according to neurovascular theory. Although neuropathic shoulder arthropathy is a rare disease, it should not be ignored as it may develop in patients with a diagnosis of CIDP. We think that careful

evaluation of patients' joints is important for early diagnosis and treatment.

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