



# Neuropathic Arthropathy of the Glenohumeral Joint as the presenting symptom of a Cervical Syrinx: A Case Report

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## Introduction

Neuropathic arthropathy, also known as Charcot's joint disease, is an extreme form of non-inflammatory osteoarthritis caused by disturbed sensory innervation and is typically asymmetric. Classically, neuropathic arthropathy is found in older male patients with an unstable, painless, and swollen joint.<sup>1</sup> Radiographic manifestations of neuropathic arthropathy may include advanced destructive changes in the joint, scattered "chunks" of bone embedded in fibrous tissue, joint distension by fluid, and heterotopic ossification.<sup>2</sup> Diabetes is the most common overall cause and typically affects the foot and ankle joints.<sup>3</sup> In the upper extremity, the most common cause of neuropathic arthropathy is syringomyelia, accounting for 80% of cases.<sup>4</sup> Syringomyelia leads to myelopathy due to compression or ischemia of the spinal cord. Myelopathy is usually characterized by weakness and clumsiness, more commonly affecting the upper extremity. The patient can experience decreased manual dexterity, gait disturbances, sensory changes and spasticity. Physical exam findings in myelopathy may include hyperreflexia, radicular signs, Hoffmann sign, inverted radial reflex, myelopathy hand, finger escape sign, clonus or Babinski sign.<sup>5</sup> Myelomalacia, a radiographic hallmark of myelopathy, appears as an area of bright signal in the spinal cord on T2-weighted magnetic resonance imaging. In this case report, we present a patient who initially experienced symptoms of carpal tunnel syndrome but over several months developed, with increasing severity, many of the classic symptoms of neuropathic arthropathy associated with syringomyelia, demonstrating the importance of maintaining a broad differential diagnosis including cervical pathology.

## Case Information

A 52-year-old man with hypertension and diabetes initially presented with severe left carpal tunnel syndrome in November 2013. After an open left carpal tunnel release in January 2014, the patient reported continued sensory changes in his fingertips and loss of manual dexterity in left hand. In May 2014,

the patient reported worsening left shoulder discomfort that started while shoveling snow. Left arm weakness, acromioclavicular joint tenderness and limited range of motion were noted. Radiographs demonstrated advanced AC joint arthrosis as well as a chronic-appearing deformity of the humeral head (Figure 1). An MRI revealed a large glenohumeral joint effusion, posterior humeral head dislocation, humeral head deformity with bone marrow edema, and chronic rotator cuff tear (Figure 2). Despite immobilization, the patient's discomfort worsened and he developed significant swelling about the shoulder. Multiple aspirations yielded bloody fluid. Analysis of this fluid revealed no signs of infection, no malignant cells, and trace amounts of extracellular monosodium urate crystals. Given the severe deformity of the humeral head, the extent of soft tissue damage to the shoulder and the relative lack of pain, concern for a neuropathic joint with cervical spine pathology as an etiology was raised. He also then reported repetitive fingertip burns to both hands when he was cooking, consistent with loss of pain and temperature sensation. An MRI of the cervical spine was obtained. The MRI (Figure 3), revealed a significant syrinx from the level of C2 to T6 with myelomalacia. From these findings, the patient was diagnosed with left shoulder neuropathic arthropathy secondary to syringomyelia. He was referred to a local neurosurgeon for treatment and subsequently underwent shunt placement into the syrinx. He is now in therapy with some resolution of his myelopathic gait but with continued diminished sensation in the hands.

## Prior Reports & Relevant Literature

Neuropathic arthropathy secondary to syringomyelia is a rare condition.<sup>6</sup> Fewer than 70 cases of Charcot's shoulder have been reported in the literature. Syrinx is most highly associated with Charcot's elbow, but it is important to include the involvement of the shoulder in a differential diagnosis. In many of the cases reported in the literature, there was a significant time lapse between the initial presentation and diagnosis of Charcot's shoulder.<sup>7,8</sup> The Charcot joint can resemble other diseases such as

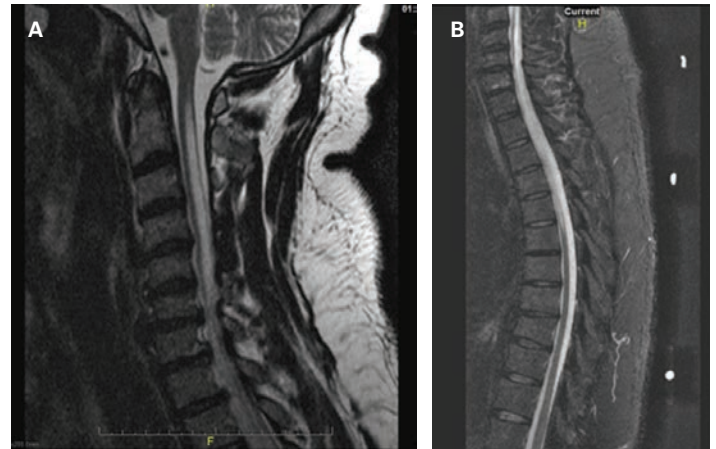
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**Figure 1.** AP Radiograph of the Left Shoulder.

inflammatory arthritis,<sup>9</sup> cancer,<sup>10</sup> or carpal tunnel syndrome,<sup>11</sup> resulting in a misdiagnosis. Several of the reported cases have unique presentations,<sup>12-14</sup> but typical symptoms include pain, swelling, and loss in range of motion. Radiographic studies usually show osteolysis, fluid collection, and degeneration resembling septic arthritis. There is no established consensus on the treatment of neuropathic arthropathy secondary to syringomyelia. Treatment strategies have ranged from neurosurgical treatment of the syringomyelia and orthopaedic

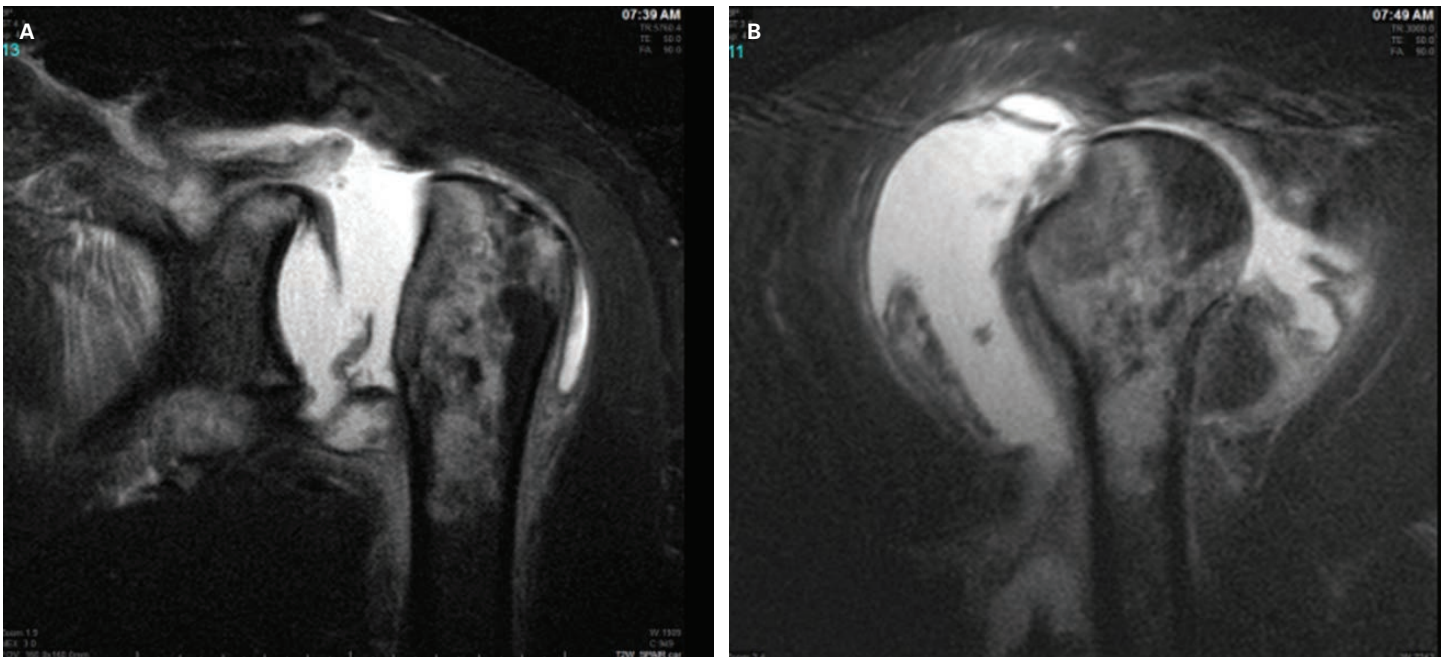


**Figure 3.** Sagittal cervical (A) and Sagittal Thoracic (B) T2-weighted magnetic resonance imaging of the spine.

interventions such as arthrodesis or resurfacing procedures to physical rehabilitation and drug treatment, with varying degrees of success.<sup>15-19</sup>

## Discussion

Establishing the diagnosis of neuropathic arthropathy and syringomyelia is often a prolonged process. This disease generally has a slow progression; the case presented above represents a relatively rapid symptom evolution over a 3-month period. There is concern that some of the neurologic changes that patients experience may become irreversible if not treated expeditiously, thus worsening the patient's prognosis. The differential diagnosis for neuropathic arthropathy includes chronic septic arthritis, sarcoma,



**Figure 2.** Coronal (A) and Sagittal (B) T2-weighted magnetic resonance imaging of the left shoulder.

idiopathic osteolysis, nephropathy, synovial chondromatosis, and Winchester syndrome in addition to cervical syrinx. Conversely, cervical pathology should be considered in the differential diagnosis of the aforementioned conditions. The definitive diagnosis of syringomyelia, made based upon the cervical MRI, raises the concern that the initial presenting symptoms were carpal tunnel syndrome-like, but in actuality due to syringomyelia.

Since diabetes leads to peripheral nerve damage, it is possible the patient's diabetes contributed to his disease process. Although the exact pathophysiology of neuropathic arthropathy is unknown, there are two predominant theories.<sup>20,21</sup> The neurotraumatic theory posits that an insensitive joint will be more prone to sustaining repetitive trauma, causing joint destruction. In the neurovascular theory, it is hypothesized that sympathetic dysfunction and sensory loss cause hyperemia and active bone resorption by osteoclasts.

Despite the wide variation in treatment of neuropathic arthropathy secondary to syringomyelia, optimal management should be focused on treating the underlying neurological cause before treating the secondary effects of syringomyelia. Aggressive orthopaedic intervention (e.g. arthrodesis and resurfacing operations) without first treating the underlying neurologic pathology has resulted in regression to the previous disease state.<sup>19</sup>

## Conclusions

The orthopaedic surgeon will be the clinician most likely to encounter this rare disease, despite its neurological origin. A detailed history, a thorough and thoughtful physical exam including strength and sensory testing, neurological and pathological reflex provocation, as well as specialized maneuvers is paramount in diagnosing neuropathic arthropathy and identifying the underlying etiology. This will prevent unnecessary surgery and expedite definitive treatment. To avoid misdiagnosis of neuropathic arthropathy, the orthopaedic surgeon should be aware of the possibility of a neurological pathologic mechanism.

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