J Shoulder Elbow Surg (2017) 26, 544–552

ELSEVIER

Journal of Shoulder and Elbow Surgery

www.elsevier.com/locate/ymse

REVIEW ARTICLE

Charcot shoulder and elbow: a review of the literature and update on treatment

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Charcot arthropathy of the shoulder and elbow is a rare disease process initially described in the 1700s; however, it was not until the 19th century that physicians understood its association with other disease processes such as cervical spine pathology and diabetes. A primary complaint is painful or painless joint dysfunction, meaning the orthopedic surgeon is regularly the first physician to evaluate the patient. Frequently, the condition of these patients is misdiagnosed. Although the pathogenesis of the disease is controversial, the etiology is commonly due to syringomyelia. The key to successful management is a thorough history and examination along with a workup including specific laboratory testing and imaging to rule out other disease processes. Most neuropathic shoulders and elbows have historically been managed conservatively because of poor outcomes with operative interventions. Newer data have emerged hinting that early neurosurgical intervention can stabilize this degenerative process. If clinical and radiographic stabilization occurs, recent studies have outlined surgical indications that can provide surgeons with a guide as to patients in whom successful operative outcomes can be achieved in the face of failed conservative management.

Level of evidence: Narrative Review

Keywords: Charcot; shoulder; elbow; syrinx; syringomyelia; neuropathic arthropathy

Syringomyelia is the leading cause of neuropathic (Charcot) arthropathy of the upper extremity, with the shoulder and elbow being the most commonly affected joints. Although neurologic deficits are occult and easily overlooked, the primary complaints of most patients are symptoms of neuropathic arthropathy, specifically painless joint swelling.[44] Thus, the orthopedic surgeon is often the first physician to evaluate the patient with syringomyelia and a Charcot shoulder and/or elbow. Early diagnosis and proper management of the joint and neurologic cause are critical. Referral to the neurosurgery department and conservative arthropathy management were traditionally the mainstay of treatment, as surgical treatments produced unfavorable results. Newer literature has advocated surgical management of the Charcot shoulder and elbow and has reported early successful outcomes.

History

The first description of neuropathic arthropathy was by Musgrave in 1703, when he described the swollen, inflamed joints of a patient who was left "flaccid by paralysis." In 1831, Mitchell reported on a patient with spinal cord paralysis due to tuberculosis and noted "bizarre" joint changes. During the latter part of the 19th century, Jean-Marie Charcot reviewed patients with tabes dorsalis and provided a detailed

Institutional review board or ethical committee approval was not applicable to this study.

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1058-2746/$ - see front matter
http://dx.doi.org/10.1016/j.jse.2016.10.015

description of the rapid development of joint deterioration and instability.[2] It was Sokoloff in 1892 who described the association of neuropathic joints of the upper extremity with syringomyelia. In the mid 1900s, neuropathic changes were described in association with diabetes and intra-articular corticosteroid injections.[8,24]

Etiology

Neuropathic arthropathy typically occurs due to syringomyelia but has also been associated with diabetes mellitus, tabes dorsalis, chronic alcoholism, end-stage renal disease, gigantism, intra-articular steroid injections, peripheral neuropathy, meningomyelocele, multiple sclerosis, myelodysplasia, leprosy, amyloidosis, and congenital insensitivity to pain.[2,52] Neuropathic arthropathy will develop in 25% of patients with syringomyelia, with 80% of these cases occurring in the upper extremity.[2,21] Although a few reports have indicated that the elbow is more commonly affected than the shoulder, most case reports have described the shoulder as the most commonly affected joint of the upper extremity.[4,20,25,43] Case reports of neuropathic arthropathy of the wrist and interphalangeal joints are rare.[12] Twenty percent of patients have multiple joints involved.[2] Cervical syringomyelia is the etiology of shoulder neuropathic arthropathy in 75% of cases and is the most common cause of elbow neuroarthropathy.[25]

Pathogenesis

Syringomyelia is a chronic and slowly progressive spinal cord disease with a fluid-containing cavity (syrinx) inside the spinal cord. The disease can be congenital or may result from infection, trauma, tumor, vascular abnormalities, or degeneration.[10,14] The decussating fibers of the lateral spinothalamic tract that harbor nerve fibers for pain and temperature sensation are the first structures to be damaged by a syrinx. This leads to abnormal innervation of the affected joint or joints[10,23]; as a result, a condition called *dissociative anesthesia* occurs, in which proprioception and motor function are preserved while pain and temperature senses are lost.[10,49] As the syrinx enlarges, the damage to the dorsal column and anterior horn will produce areflexia, loss of muscle strength, and atrophy. The eventual joint destruction can occur early or late in the disease process.[43]

There are 3 phases of the neuropathic joint. In the destructive phase, the joint is hyperemic and swollen and there is osteoclastic bone resorption associated with repetitive trauma. The reparative phase follows with the formation of dense fibrous tissue and coalescence of the debris. Finally, the quiescent phase is characterized by decreased vascularity and osseous sclerosis.[16]

The pathogenesis of syringomyelia and neuropathic arthropathy is not fully understood, and many theories have been proposed. Mitchell and Charcot hypothesized in the 19th century that damage to the central nervous system trophic centers disrupts bone and joint nutrition and causes osteolysis. Volkmann and Virchow theorized that after a loss in pain sense, joint destruction was caused by years of subclinical trauma. More recently, neurovascular and neurotraumatic theories for the development of neuropathic arthropathy were developed.[4]

According to the neurovascular theory, sensory loss disrupts normal neurovascular reflexes at the joints. The resulting hyperemia and activation of osteoclasts cause bone resorption.[7,52] The neurotraumatic theory involves loss of somatic muscle reflexes, which prevents protective proprioception. This leads to recurrent, unnoticed microtrauma, causing joint destruction from extremes of joint motion. Not all reported cases have a history of trauma, as neuropathic arthropathy is also seen in paraplegic, bedridden patients. Therefore, the most widely accepted current theory is that osteolysis starts because of neurovascularity processes and continues because of neurotraumatic processes.[7,22,52]

In 1997, Gough et al[19] proposed a mechanism of inflammation leading to excess osteoclastogenesis. This is supported by an elevation in blood marker levels showing osteoclast activity (tumor necrosis factor α, IL-6) in patients with acute Charcot arthropathy and an elevation in proinflammatory cytokine levels in bone samples from surgery.[6,39]

Ultimately, there is no widely accepted theory as to why syringomyelia causes neuropathic arthropathy. A large question remains as to why this disease process tends to occur unilaterally or is monoarticular. Although the syrinx does involve both halves of the cord, usually only one side of the body is affected. Multiple studies have reviewed the cervical magnetic resonance imaging (MRI) scans of the syrinx and confirmed that it is often slightly asymmetrical, with every patient having neuropathic arthropathy on the affected side.[1,12,27,37]

Diagnosis

The clinical findings in neuropathic shoulder arthropathy are widely variable. Usually, joint symptoms manifest earlier than neurologic symptoms, with patients presenting initially to orthopedic clinics.[21] Patients may present with painful or painless joints, joint instability, swelling, and dysfunction with or without a history of trauma. Most patients with a neuropathic elbow present with elbow instability and/or subluxation.[11,27,53] Physical examination reveals joint laxity, crepitus, effusion, and often, decreased muscle strength.[2] On examination, sensory and temperature changes are often revealed, most notably along the patient's back and arms in a cape-like distribution. Also noted are a loss of hot or cold hand sensation and the presence of asymmetrical reflexes.[12,23,52]

To firmly diagnose a syrinx and neuropathic arthropathy, other conditions must be ruled out. The differential includes septic arthritis, arthritis, synovial chondromatosis, soft-tissue sarcoma, tumoral calcinosis, idiopathic osteolysis, Winchester syndrome, Gorham disease, trauma, and blood clot.[4,15,25] Another condition in the differential is Milwaukee