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Case Report

Septic and Dislocated Elbow as the Initial Manifestation of a Cervicothoracic Syrinx: Case Report

Kenneth Ford, MD[1]; Angus Brooks, MD[1]; Adeet Amin, MD[1]; Zachary Hoelscher, MD[1]; Camden Tissue, MD[1]

Abstract

Case: A previously healthy 56-year-old male presented with a painless, swollen elbow and septic shock following a minor puncture wound. Imaging and subsequent arthrocentesis revealed a dislocated elbow with concurrent septic arthritis, yet the patient demonstrated minimal pain and near full range of motion. This discordant clinical picture prompted further investigation, revealing an extensive, previously undiagnosed cervicothoracic syrinx.

Conclusion: Septic arthritis with simultaneous acute-appearing joint dislocation represents an unusual initial presentation of neuropathic arthropathy secondary to syringomyelia. Clinicians should maintain high suspicion for underlying neurological pathology when encountering patients with severe joint abnormalities but disproportionately minimal pain or functional limitation.

Keywords: Syrinx, Elbow Dislocation, Septic Arthritis

Introduction

Syringomyelia is condition characterized by a fluid filled cavity, syrinx, found within the parenchyma of the spinal cord.[1] While many cases are discovered incidentally, symptomatic patients typically present with pain, hyperesthesias, myelopathic symptoms, or occipital headaches.[2]

Charcot arthropathy is a chronic, neuropathic arthropathy characterized by progressive joint destruction. Cervical syrinx has an established relationship with Charcot arthropathy of the upper extremity, with the shoulder being the most commonly affected joint, followed by the elbow.[3] Despite this well described correlation, there are relatively few cases of cervical syrinx and Charcot arthropathy of the elbow.[4,5,6] A recent systematic review identified only 50 such cases in the literature, 33 of which were isolated elbow arthropathy as in our patient.[7] More unusual still, only two previous case reports have described septic arthritis of the elbow in patients with

Corresponding Author:
Angus Brooks, MD
University of Texas Health Science Center at Houston
Department of Orthopedic Surgery, Houston, TX, USA
Email: angus.c.brooks@uth.tmc.edu

Figure 1. (A) Clinical photograph of the affected elbow showing significant soft tissue swelling and a posterolateral wound. (B) AP radiograph of the affected elbow showing aggressive periosteal reaction along the medial aspect of the humerus concerning for possible osteomyelitis. (C) Lateral radiograph of the affected elbow showing posterior dislocation with a large effusion.

neuropathic arthropathy secondary to cervical syrinx.[8,9] Notably, all previously reported cases presented with advanced Charcot changes in the affected joint.

Here we outline the presentation and subsequent follow up of a patient with previously undiagnosed cervicothoracic syrinx who presented with concurrent septic arthritis and a radiographically acute appearing elbow dislocation.

Case Description

A 56-year-old man with no past medical history presented to the emergency department with progressive swelling to the right elbow which had begun the previous month following a punctate wound to the posterolateral elbow. His physical exam was notable for significant elbow swelling though a painless arc of motion from 5 to 135 degrees. He was febrile to 39.4 degrees Celsius with an elevated heart rate of up to 130, and elevated ESR and CRP. He met sepsis criteria at the time of presentation. Further neurological examination revealed diminished proprioception in his feet and inability to heel toe walk along with decreased pain and temperature perception to his bilateral upper extremities.

Radiographs in the emergency department demonstrated a dislocated ulnohumeral joint without evidence of joint destruction (Fig. 1). The workup was continued with CT (Fig. 2) and MRI (Fig. 3) of the elbow which demonstrated a posterior elbow dislocation with a large elbow effusion concerning for septic arthritis and aggressive periosteal reaction along the medial aspect of the distal humerus suggestive of osteomyelitis. The patient subsequently underwent urgent surgical irrigation and debridement of the right elbow. He was initially started on broad-spectrum antibiotics, cefepime and vancomycin, which were later narrowed to cefazolin based on culture sensitivities of methicillin sensitive staph aureus.

The patient's unusual presentation of painless joint dislocation with infection prompted further neurological investigation. MRI cervical spine demonstrated an expansile T2/STIR signal hyperintense signal nearly completely replacing the cord parenchyma extending from the level of the dens through T3 level which was consistent with syrinx formation (Fig. 4). Postcontrast MRI was obtained and did not demonstrate an underlying neoplasm. Neurosurgical consultation was obtained, though the

Figure 2. Coronal (A), Sagittal (B), and Axial (C) CT of the humerus and elbow demonstrating a posterior elbow dislocation with periosteal reaction and cortical erosion of the posteromedial aspect of the distal humerus.

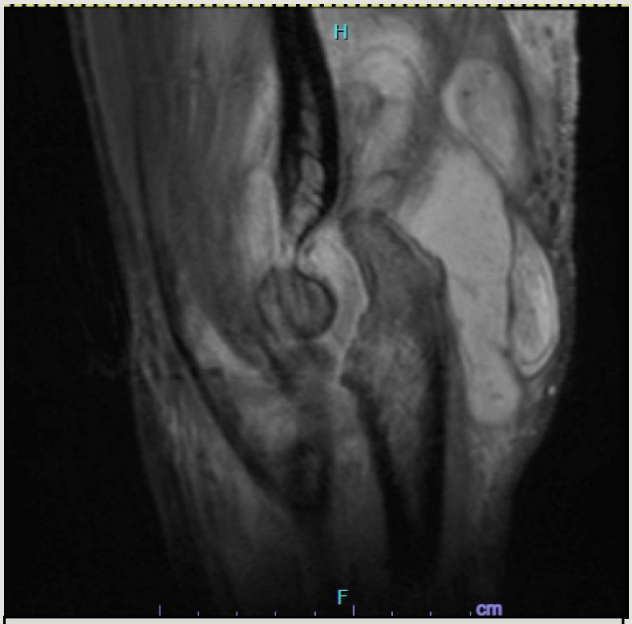

Figure 3. Sagittal proton density fat suppressed magnetic resonance imaging scan of the elbow demonstrating destructive changes of the coronoid with extensive

Figure 4. Sagittal T2-weighted (A) and T1-weighted (B) magnetic resonance imaging scan of the cervical spine showing a syrinx extending from the level of the dens through T3. (C) Axial T2-weighted magnetic resonance imaging scan of the cervical spine showing a syrinx near completely replacing cord parenchyma.elbow effusion which is continuous with subcutaneous and muscle edema.

patient ultimately declined surgical intervention, stating he had minimal perceived disability and was concerned about potential surgical risks. Over the next six days, the patient continued to have spiking fevers with elevated inflammatory markers which necessitated a third irrigation and debridement. Following this third surgery he was discharged with a peripherally inserted central catheter and six weeks of antibiotics per infectious disease.

At his initial postoperative visit, radiographs demonstrated rapid onset of neuropathic changes with progressive osteolysis of the elbow, near-complete absence of the coronoid process and portions of the olecranon, and heterotopic ossification peripherally. At 10-week follow-up, the patient's elbow remained clinically unstable though still pain-free, with near-full range of motion. Radiographs showed progressive findings of neuropathic arthropathy with persistent posterolateral dislocation (Fig. 5). The patient was counseled regarding elbow arthrodesis at 90 degrees to address the instability, but he declined, believing the position would severely limit his occupational activities. At one-year follow-up, the patient reported no pain in his elbow but noted progressive loss of dexterity in his hands, and loss of range of motion, now with a roughly 30-degree arc of motion. Despite these symptoms, he remained uninterested in surgical management of either the syrinx or the elbow, as he desires to preserve any motion possible to the elbow to maintain his employment. He continues to follow with the neurosurgical and orthopedic clinics for monitoring of the progression of his symptoms.

Discussion

Neuropathic arthropathy, also known as Charcot arthropathy, is a chronic condition in which there is progressive fragmentation of bones which ultimately results in destruction of the affected joint. The pathophysiology has been proposed to involve a combination of neurovascular and neurotraumatic mechanisms. The neurotraumatic theory states that a loss of protective sensation occurs in the joint which allows repeated trauma to go unnoticed which over time causes degradation of the joint.[10] Alternatively, the neurovascular theory states that increased blood flow to the neuropathic joints results in increased osteolysis and demineralization.[11] Charcot arthropathy can present secondary to a multitude of underlying pathologies, most commonly diabetes in the lower extremity

and syringomyelia in the upper extremity.[12] Presenting symptoms vary based on the severity of the disease and can include swelling, deformity, erythema, and warmth in the affected area.

Syringomyelia is a progressive, chronic condition caused by abnormal cerebrospinal fluid flow and is characterized by a fluid-filled cavity present in the spinal cord. It is most commonly associated with a Chiari I malformation, but can also be seen years after spinal trauma, meningitis, intramedullary or extramedullary tumors, or can be due to an idiopathic cause.[13] The most common presenting symptoms are paresthesias, hyperesthesia, and non-radicular segmental pain. Other common symptoms are gait ataxia and hand muscle weakness with loss of fine motor function. Classically, a centrally located syrinx can selectively compress the dorsal columns of the spinal cord, leading to isolated loss of pain and temperature sensation.[2,14]

Treatment of a syrinx varies based on the underlying pathology. Patients with a Chiari I malformation often are deemed to benefit from craniocervical decompression, which consists of a removal of the posterior arch of C1 and a suboccipital craniectomy with removal of arachnoid adherences.[15] If these treatments fail, or if the syrinx has an idiopathic cause, a shunt is often used to decompress the syrinx. This, however, is generally not preferred due to high complication rates and the shunt's lack of addressing the underlying pathology[16]. First-line treatment for the extremity manifestations should address the underlying syrinx when possible. Orthopedic management options for Charcot elbow are limited, with arthroplasty being contraindicated. Arthrodesis is regarded as the most reliable option for providing stability, though at the expense of motion.[17] Our patient's case illustrates the complex decision-making involved, as the patient prioritized motion preservation for employment purposes.

This case is distinctive for several reasons. First, unlike previously reported cases of septic, neuropathic elbows,[4-7] our patient presented with what radiographically appeared to be an acute dislocation without the typical findings of advanced Charcot. This suggests that the infection and the resulting inflammatory cascade may have disrupted the joint capsule and ligamentous structures, precipitating a fairly acute dislocation in an elbow already compromised by early neuropathic changes. Second, the patient had no prior diagnosis of

Figure 5. 10-week follow-up AP (A), external oblique (B), and lateral (C) elbow radiographs demonstrating erosive arthropathy with bone loss and extensive heterotopic ossification at the elbow joint.

or symptoms from his extensive cervicothoracic syrinx, despite its impressive size on MR imaging. This reinforces the often-insidious nature of syringomyelia, where patients may adapt to gradually developing neurological deficits without recognition.

Conclusion

This case highlights a previously undescribed presentation of neuropathic elbow arthropathy secondary to a previously undiagnosed cervicothoracic syrinx, manifesting initially as septic arthritis with concurrent joint dislocation. While the patient initially presented with relative sparing of the joint architecture, his follow-up has gone on to demonstrate the expected neuropathic changes with extensive destruction of the joint. Clinicians should maintain a high index of suspicion for underlying neurological pathology when encountering patients with significant joint pathology but disproportionately minimal pain or functional limitation. A thorough neurological examination is essential in such cases and may reveal previously unrecognized underlying pathology.

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Affiliations:
[1] University of Texas Health Science Center at Houston Department of Orthopedic Surgery