Syringomyelic Neuropathic Arthropathy of the Elbow

Neil Stewart, Kevin Karpik

ABSTRACT

Neuropathic Arthropathy or Charcot joint is a progressive, destructive arthritis that is associated with an underlying neurological disorder. We present a case of a 30 year-old male who, three years prior, had ruptured his right distal biceps tendon with subsequent development of a deep infection. At representation, the patient's clinical picture was consistent with the re-emergence of a deep elbow infection. Laboratory testing found no evidence of infection. Magnetic Resonance Imaging (MRI) of the patient's spine revealed a syringomyelia and a NA was diagnosed. The purpose of this report is to raise awareness of a unique presentation of a rare clinical condition in the early stages of the disease.

Neuropathic Arthropathy (NA) is a rare disorder which typically affects the joints of the lower limb. Involvement of the upper limb is uncommon. Among patients with NA, the elbow joint is thought to be affected in only 3% of cases. NA of the elbow as a presenting symptom of syringomyelia is extremely rare. We describe a unique case of NA of the right elbow preceded by a septic arthritis of the same joint. The purpose of this case report is to raise awareness of an uncommon pathology in its early stages where outcomes may be influenced by early diagnosis and management.

Case Report

A 30 year-old, right-hand dominant, non-diabetic male presented to the emergency department with a three week history of insidious onset pain, reduced range of motion, crepitus and circumferential swelling of his right elbow.

Three years prior he had ruptured his right distal biceps tendon at its musculotendinous junction while lifting heavy furniture. His tendon had been surgically repaired. He re-presented seven weeks later with swelling and erythema over the anterior aspect of his elbow, pain on elbow movement and subjective fevers. A deep infection of his biceps tendon had developed. This infection was managed with three washout and debridement procedures. Intra-operative tissue samples isolated Staphylococcus Aureus. He received six weeks of intravenous antibiotics via a peripherally inserted central catheter. Clinical follow-up at two, three and six months had noted normal inflammatory markers, radiographs and resolution of symptoms.

On representation, his right elbow was circumferentially 12cm larger than his non-affected side. Range of motion was limited to -20° extension and 120° of flexion. The limb was neuro-vascularly intact. C-Reactive Protein was 38mg/L (Reference Range [RR] 0–5), Eosinophil Sedimentation Rate was 41mm in one hour (RR 1–10) and Neutrophil count was 7.3 xE9/L (RR 1.9–7.5). Radiographs revealed a large joint effusion, calcification within the joint capsule, loose bodies, increased density within the distal humerus, proximal radius and ulnar, joint deformity and destruction (Figure 1 and Figure 2).

The patient was subsequently taken to theatre for washout and debridement of the right elbow for a suspected septic arthritis. No purulent material was seen, instead large amounts of straw-coloured synovial fluid, full thickness cartilage loss of the radial head and trochlear, complete loss of the capitulum and multiple loose bodies were identified.

Intra-operative aspirates found no crystals or organisms. Biopsies of the synovium and distal humerus failed to identify any organisms with Gram stain or extended cultures. Polymerase Chain Reaction did not identify Mycobacterium Tuberculosis DNA; and Quantiferon-TB gold testing was negative. DNA Sequence Analysis did not amplify any bacterial DNA.
Figure 1: Anteroposterior plain radiograph of the right elbow. Joint effusion, calcification within the joint capsule, loose bodies and increased density within the distal humerus, proximal radius and ulnar are evident.

Figure 2: Lateral plain radiograph of the right elbow. Joint effusion, calcification within the joint capsule, loose bodies and increased density within the distal humerus, proximal radius and ulnar are evident.
A possible diagnosis of a NA was raised. The patient underwent MRI investigation of his brain and spinal cord which identified a Chiari I Malformation and Syrinx extending from C1 to T7 (Figure 3). A NA had been confirmed.

**Discussion**

NA is a progressive, degenerative arthritis associated with an underlying neurological disorder. Charcot first reported this form of destructive arthropathy in 1868, which accompanied neurosyphilitic infection.

Table 1: Central and peripheral causes of Neuropathic Arthropathy.

<table>
<thead>
<tr>
<th>Central</th>
<th>Peripheral</th>
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<tbody>
<tr>
<td>Tabes dorsalis(^1,10,15)</td>
<td>Diabetes Mellitus(^1,10,15)</td>
</tr>
<tr>
<td>Leprosy(^7,10,11)</td>
<td>Alcoholism(^1,10,11)</td>
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<tr>
<td>Syringomyelia(^1,3,10)</td>
<td>Amyloid neuropathy(^1,10,11)</td>
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<tr>
<td>Multiple Sclerosis(^1,10)</td>
<td>Infection(^7)</td>
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<tr>
<td>Charcot Marie-Tooth(^1,10,12)</td>
<td>Familial sensory neuropathies(^1)</td>
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<tr>
<td>Neurofibromatosis(^11)</td>
<td>Pernicious anaemia(^1,10)</td>
</tr>
<tr>
<td>Herpetic encephalitis(^11)</td>
<td>Intra-articular or systemic corticosteroid use(^1,10,11)</td>
</tr>
<tr>
<td>Systemic Scleroderma(^15)</td>
<td>Congenital insensitivity to pain(^12)</td>
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<td></td>
<td>Poliomyelitis(^7)</td>
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and demyelination of the dorsal columns of the spinal cord (Tabes Dorsalis). Since first reported, multiple other causes of NA have been described (Table 1). These causes can be divided into either central or peripheral neurological insults, dependant on the distribution of nervous system involvement.1,7,9,10 In contemporary times, NA is most commonly associated with Diabetes Mellitus and its peripheral neuropathic sequelae.1,9,11,12 Weight-bearing joints such as the foot, ankle, knee and hip are typical.1,13 Involvement of the upper limb is uncommon1,9,12 but when this does occur, typically involves the shoulder joint.3,12 NA of the elbow joint is exceedingly rare.1,6,7,9,12

The pathogenesis of NA remains controversial, with several proposed theories.1,9,14 The Neurotraumatic theory suggests that repetitive trauma sustained by an insensate joint results in joint destruction.1,3,7,15 The Neurovascular theory describes active bone resorption by osteoclasts secondary to sympathetic dysfunction and a neutrally medicated persistent hyperaemia.1,7 A further theory proposes that joint changes result from damage to central nervous system ‘trophic centres’ which control bone and joint nutrition.1

The classical clinical presentation of NA involves joint swelling,1,12 erythema,1 reduced range of motion,7 crepitus,10 joint instability8 and the absence of pain.1,2,12 However, presentations involving various degrees of joint pain have now been reported.7,9,16

The radiological and pathological features of NA have been summarised into the ‘6-Ds’: dense bones (subchondral sclerosis), destruction of articular cartilage, disorganisation (joint deformity), debris (loose bodies), distention (fluid) and dislocation.7

In this case report, syringomyelia was identified as the underlying cause of the patients elbow NA. Syringomyelia is a progressive degenerative disorder of the spinal cord characterised by longitudinal cavitation (syrinx) containing a fluid which involves the cervical or cervicothoracic regions.10 Patients with a syrinx typically present with a dissociated segmental anaesthesia over the neck, shoulder and arm in a cape or hemicape pattern.16 The perturbation of sensory symptoms are the result of the interruption of the decussating fibres of the lateral spinthalamic tract that mediates pain and thermal sense while sparing fibres that mediate deep touch sensation.16 As the syrinx expands it may damage the anterior horn cells, causing a lower motor neuron lesion at the same level or damage to the descending cortical motor tracts, leading to an upper motor neuron lesion below the level of the syrinx.7

The object of treatment is to manage the underlying disease and reduce the rate of joint deformity.7,10,17 Thus, it is important to diagnose and treat NA as early as possible.17 Several management strategies have been proposed. Typically, physical therapy is recommended to ensure range of motion and function are maintained.9,10 Functional bracing may also be utilised, particularly if the joint destruction has resulted in instability.9,10 Nonsteroidal anti-inflammatory drugs can be used for the control of synovial inflammation.10 Aspiration of large effusions may prevent ligamentous laxity.16 Generally, surgical management is not recommended and has been associated with high rates of complications and unpredictable outcomes.5,9,16,18

Total elbow replacement is contraindicated due to the lack of protective pain sensation and reflexes, the presence of osteopenic bone and weakness of the surrounding ligamentous and muscular tissues.9,10,16 This causes abnormally high stress on implanted components, with associated loosening and periprosthetic fractures resulting in high failure rates.16 One report of successful arthrodesis is reported in the literature.5
Competing interests:
Nil.

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