



## Charcot's Elbow Following Syringomyelia: Revisited

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### ABSTRACT

Neuropathic arthropathy of the elbow joint is uncommon with a reported incidence of 3-8% only. The natural history, pathogenesis and management approaches are debatable with an absence of clear guidelines. We report two cases of Charcot's elbow following syringomyelia with ulnar nerve manifestations. One patient required debulkingsynovectomy and ulnar nerve decompression while the other was managed conservatively. The article aims to enrich the limited knowledge of managing Charcot's elbow.

**Keywords:** charcot's joint; elbow; syringomyelia; ulnar nerve.

### INTRODUCTION

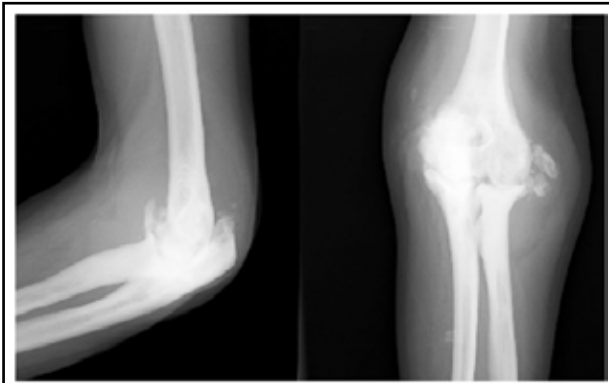
Neuropathic arthropathy or Charcot's joint is characterized by painless progressive joint destruction due to an underlying neurologic disorder.<sup>1</sup> It was first described by Charcot in 1868.<sup>2,3,4</sup> Syringomyelia is an important cause in the upper limb.<sup>5</sup> The primary objective of this paper is to cast more light on the clinical presentation and optimal surgical management of the same. We report two cases of syringomyelia of cervicodorsal spine with Charcot's arthropathy of the elbow joint with ulnar nerve neuropathy.

### CASE REPORT

A 55 year old gentleman presented with painless progressive swelling of left elbow for four years and paresthesia of the left forearm and hand for six months. There was no history of trauma or pain in the neck. Local examination revealed massive, non-tender, boggy elbow swelling with crepitations and loose bodies. Synovium was hypertrophic with thick, non-tender ulnar nerve. Painless elbow flexion 0-160° with 15° of hyperextension was present. The opposite normal elbow had 0-135° flexion. Pronation and supination were full and painless. Elbow was grossly unstable medio-laterally

within a 12° range. Neurological examination revealed reduced touch, temperature and pain sensations over C5 to T2 dermatome on left side and paraesthesia along the ulnar nerve distribution. Elbow and wrist power were grade 4/5. Grip strength was 26 kg as measured with dynamometer on the affected side as compared to 42 kg on normal side. Plain radiograph revealed grossly disorganized elbow joint with multiple loose fragments (Figure 1). Magnetic resonance imaging (MRI) of cervical spine identified a syrinx at C4-C7 level (Figure 2). Patient underwent debulkingsynovectomy of the left elbow with decompression and neurolysis of the ulnar nerve. Postoperatively, elbow was mobilized in a hinge splint for three weeks. At six months follow up, patient's sensory symptoms improved and grip strength increased from 26 kg to 35 kg.

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**Figure 1.** Anteroposterior and lateral views of the left elbow reveals severe destruction of the elbow joint and multiple loose bodies.



**Figure 2.** MRI (T2 sequence) of cervical spine showing syrinx from C4 to C7 level.

## Case 2

A 33 year old female was referred from general surgical outpatient department for suspected sarcoma around the left elbow. She complained of painless, progressive swelling of her left elbow and clumsiness of left hand for three months. Upper limb tone was normal but lower limbs revealed hypertonia. Intrinsic muscles of hand revealed weakness in ulnar and median nerve distribution. Left hand grip strength was 13 kgs as compared to 45 Kgs on normal side as measured by dynamometer. Sensations were decreased on left side from C5 to T2 dermatome. Left elbow revealed swelling on medial side. Synovial hypertrophy was felt with gross crepitation. The olecranon had migrated proximally. Flexion was  $10^{\circ}$  to  $160^{\circ}$  and painless. Pronation was  $60^{\circ}$  and supination  $40^{\circ}$  but painless. Elbow was unstable on varus stress. Radiography revealed grossly disorganized elbow joint with multiple loose bodies (Figure 3). MRI revealed syrinx from C4 to T2 level (Figure 4). Neurosurgical opinion was sought as there was lower limb hypertonia indicating upper motor neuron involvement. Syrinx decompression

and ulnar neurolysis was planned. However, patient refused surgical intervention and was discharged on elbow hinge splint and physical therapy. Eight months later, she regained another  $10^{\circ}$  each of pronation and supination, without any further worsening in upper and lower limbs neurological symptoms.



**Figure 3.** Plain radiograph of elbow reveals typical neuropathic changes in bones with subluxation of the elbow.



**Figure 4.** MRI (T2 sequence) of cervical spine showing syrinx from C4 to T2 level.

## DISCUSSION

Neuropathic arthropathy of the elbow following syringomyelia with peripheral nerve manifestations is a rare presentation.<sup>1,6</sup> Very few authors reported more than three cases.<sup>1,2,6</sup>

Charcot's joints commonly involves weight bearing joints, the common causes being tabes, diabetes and leprosy.<sup>1,2</sup> Upper limb involvement is rare, syringomyelia being the commonest cause,<sup>2,3,4,5,6,7</sup> with incidence in

elbow being 3-8% only.<sup>6,7</sup>

Pathogenesis is controversial with multiple theories including the neurotraumatic, neurovascular and neurotrophic theories.<sup>3,5,7,8</sup> Though classically described as painless, one-third of the patients do have pain.<sup>1,8</sup> Both of our patients had painless destruction of the elbow.

Unnanuntana has described the radiological appearance as "5 D's - debris, density, destruction, disorganization and dislocation".<sup>7</sup> Radiological appearances can be hypertrophic (productive) and atrophic (destructive) types.<sup>1,6,7</sup> Deirmengian points out that all reported elbow cases were hypertrophic, whereas Kwon YW opines that upper extremity cases are usually atrophic.<sup>1,6</sup> Both of our cases appeared to be hypertrophic.

There are some reports of peripheral nerve involvement. Deirmengian describes three cases of ulnar neuropathy, probably due to distortion of cubital tunnel, of which two needed decompression.<sup>1</sup> Meyn has also reported ulnar transposition for his case.<sup>3</sup> In our study, considering the positive result of debulking synovectomy and decompression, we recommend the procedure, if the patient is refractory to conservative measures.

The aim of treatment is to reduce further articular damage while retaining a functional joint,<sup>1,4</sup> by usage of efficient bracing, calipers or corset.<sup>1,4,7</sup> Yeap has suggested initial immobilization till active process subsides.<sup>9</sup> Surgery is considered when conservative treatment has failed and the integrity of surrounding soft tissues is threatened.<sup>8</sup> Surgical options include prosthetic replacement, resection arthroplasty or arthrodesis.<sup>7</sup> Arthroplasty is contraindicated when muscles are paralyzed with porotic bones.<sup>10</sup> Neuropathic elbows are difficult to arthrodesis with poor functional results at any angle of fusion due to the sclerotic poorly vascular bone.<sup>9</sup> Peripheral nerve decompression for entrapment is worthwhile.<sup>1</sup> It helps in improving the overall function of the affected limb. One patient who underwent ulnar nerve decompression showed improved sensori-motor function over a period of 6 months. Neuropathic elbow arthropathy due to syringomyelia, though uncommon, needs early identification. The role of surgery in cases of peripheral nerve entrapment is worthwhile and safely recommended.

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