A Unusual Case of Charcot's Arthropathy of Elbow Caused by Syringomyelia and Chiari Malformation Complicated with Scoliosis

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Abstract

Neuropathic joints, which is also known as Charcot's Arthropathy, is a destructive joint pathology resulting from loss or decrease in proprioception, pain and temperature sensations. Patients suffering from diseases associated with neuropathy like diabetes mellitus, syringomyelia and syphilis are said to be prone to the disease. Patients with syringomyelia commonly suffer with arthropathy involving shoulder. We report a case of 35 year old male patient with syringomyelia and charcot's arthropathy involving left elbow.

Keywords: *Charcot's Arthropathy; Syringomyelia; Elbow.*

Introduction

Charcot's arthropathy which was first reported by Jean-Martin Charcot in 1868[1], is a destructive joint pathology resulting from loss or decrease in proprioception, pain and temperature sensations[2-4]. The etiological factor of Charcot arthropathy includes Tabes dorsalis, syringomyelia, traumatic spinal cord injury, leprosies, congenital pain insensitivity, diabetes mellitus, chronic alcoholic intoxication, and so on. Although Charcot arthropathy associated with above mentioned factor has been mentioned in the literature, but a very less number of cases describing Charcot arthropathy of elbow caused by syringomyelia and Chiari malformation associated with scoliosis have been reported.

Case Presentation: A 35 years old male patient reported to our hospital complaining of weakness in right hand since the last 12 years and swelling over his left elbow since 3 years. Weakness was associated tingling and numbness in left upper limb. Swelling over the elbow joint was painless, diffuse and was gradually progressive. Patient underwent incision and drainage 2 years back carried out by a local practitioner and after the drainage, swelling did not subside and a gradual increase in size was noted. Patient gave a history off and on fever with loss of appetite and weight loss of around 15 kg in 3 years. Patient had no history trauma, comorbidities or any surgical history in the past.

On clinical examination, Diffuse swelling of size 5 x 6 cm approximately, non-adherent to bone or underlying structures, smooth over the lateral surface and irregular over the medial surface was present around the elbow joint and valgus deformity was present. Wasting of left forearm and hand muscles was seen. Elbow range of motion was 0-130 degrees and pain-free with full supination and pronation with medial and lateral laxity at the elbow joint. Bony crepitus was noted while performing elbow range of movements. On clinical examination of cervical spine, there was weakness in finger grip of left hand with sensory hypoesthesia over the dorsal aspect of left hand. Motor power of bilateral shoulder, elbow and wrist joints were normal. Examination lumbar spine and other limbs was normal.

Haematological examination in the form of complete blood count with Erythrocyte sedimentation rate, fasting and post meal blood sugar levels, C reactive protein levels, veneral disease research laboratory tests, uric acid, liver function test and kidney function test were carried out and found to be within normal limits except ESR level was slightly raised.

Joint aspiration was also done to rule out infection but culture did not find any organism growth.

Radiographs of Antero-posterior and lateral view of left elbow jointshowed degenerative changes in left elbow with decrease in joint space, flattening of articular margins of olecranon with marginal osteophytes at the olecranon process with periarticular sclerosis was seen. Loose bodies are seen near the radial head. Radiographs of Anteroposterior and lateral views of cervical spineshowed scoliosis at the cervico-dorsal spine with primary curve having convexity towards left side and secondary curve having convexity towards right side.

MRI of cervical spine showed a syrinx from the level of upper border of C1 vertebra till lower border of C2 vertebra. Sagittal section of T2 weighted image was suggestive of Chiari malformation with tonsillar herniation.

Patient was sent to neurosurgery department and patient was treated with posterior fossa decompression. From Orthopaedic point of view, swelling was aspirated and patient was immobilized with above elbow slab for 15 days and non-steroidal anti-inflammatory drugs were started. Swelling gradually subsided after 15 days of conservative treatment and above elbow slab was removed. Rather than pain, instability was the chief complaint. Hence, Patient was given customised Dynamic Hinged Elbow Braceand Elbow was mobilised using the brace. At the end of 1 year, patient remained asymptomatic and able to do his day to day activities.

Discussion

Charcot's Arthropathy of elbow is usually rare condition with few case report and case series being reported in literature.[6-17] Only 3% to 8% neuropathic joints, reports involvement of elbow joint. In the literature, two theories were postulated by the researchers that describe the pathogenesis of this condition, they are the neurotraumatic and neurovascular theory. As described Johnson in 1967, the neurotraumatic theory suggest the cause to be repetitive trauma to the affected joint which has lost sensations. The neurovascular theory described by Allman et al suggests active bone resorption due osteoclasts after sympathetic dysfunction and hyperaemia leading to joint destruction.[4]

On the basis of Underlying aetiology, joint involvement pattern can be identified like patients with Diabetes Mellitus are prone to develop this pathology in the ankle and foot while in Patients with syphilis, hip and knee joint involvement is common. In cases with syringomyelia, the upper limbs joints are seen to get commonly involved. Thus, in our case report underlying etiology and pattern of joint involvement resembles the literature findings.

Syringomyelia is characterized by cavitation of the spinal cord which can translates into a progressive clinical syndrome and can thereby lead to loss of neurologic functions. 16 one of the underlying aetiology of syringomyelia can be Chari malformation which was first described by H. Chiari in 1891. It was described as herniation of inferior lobe of cerebellum with elongated tonsils into the spinal canal. Thorough neurosurgical examination and investigation for consideration of surgical management should be obtained, as was carried out in our case.[18,19]

The treatment of choice for charcot's arthropathy has not been very clearly described in literature. The recent study derived guidelines suggests analgesia, elbow immobilization till swelling subsides followed by physiotherapy, and management of patient expectations.

Surgical Arthrodesis with external or internal fixation has resulted in variable surgical outcomes to some researchers.[15,16,17]. Kwon et al[15] reported a case series of 6 neuropathic elbows, out of which 3 cases were managed surgically developed post-operative complications which required additional surgery. A septic neuropathic elbow was managed with surgical debridement and external fixation resulted in instability in further follow-ups.[7] Elbow arthrodesis which was a treatment of choice after tubercular elbow is rarely performed today[20]. Arthrodesis is suggested only when there is no other reasonably reconstructive option available.[20]

As there was lack of valid guidelines for neuropathic elbow in the recent literature and patient wanted to have some function at elbow for daily activities, a trial of conservative management was considered in our case. Immobilisation of elbow with above elbow slab for 15 days was advised and as swelling was reduced, elbow was mobilised with dynamic hinge elbow brace for 3 months. At the end of 1 year follow-up, patient was asymptomatic and patient was able to perform daily activities.

The case of neuropathic elbow joint secondary to syringomyelia with chiari malformation with scoliosis has rarely been reported in the recent literature.

We conclude that the maintenance of function of a joint, rather than immobilization, is the 'motto' of treatment of neuropathic joint. We managedour patient conservatively, as suggested by the majority of theliterature, and the patient was satisfied with the treatment.

After a thorough literature search, we can say that, this is the first report of neuropathic joint of elbow, secondary to syringomyelia with Chiari Malformation with scoliosis on the Indian subcontinent.

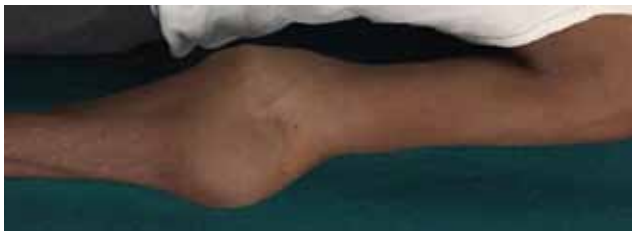

Figure 1: Showing clinical photographs depicting gross swelling over the elbow joint with wasting of muscles of forearm and hand.

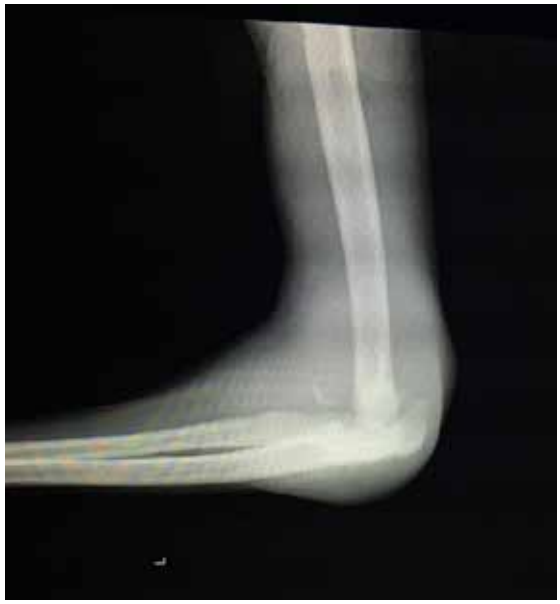

Figure 2: Antero-posterior radiographs of the Left Elbow Joint showing decrease in joint space, flattening of articular margins of olecranon with marginal osteophytes at the olecranon process.

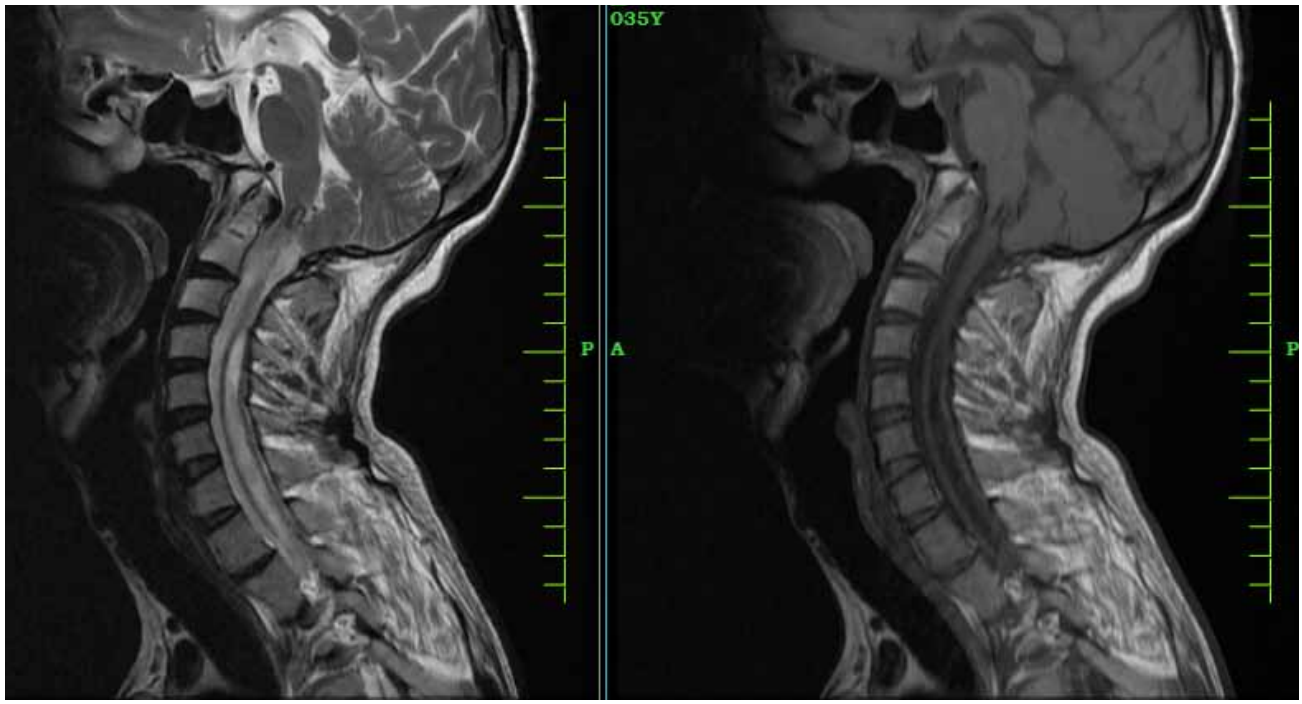

Figure 3 & 4 showing T2 and T1 weighted images of MRI showing a syrinx from the level of C1 vertebra to C2 vertebra. Sagittal section of T2 weighted image was suggestive of Chiari malformation with tonsillar herniation in the spinal canal.

Figure 5 showing clinical photograph of patient's elbow mobilised with hinged dynamic elbow splint.

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Conflict of Interest: Nil.

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