

# Charcot's Arthropathy of Elbow Associated with Type I Arnold-Chiari Malformation – A Case Report

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### Abstract

Jean Martin Charcot first described neuropathic arthropathy in 1868 in a tebes dorsalis patient. The joints most commonly affected are the weight bearing joints. It is classically described as painless lesion. We report a case of 50-year-old man presented with clumsiness of gait. He had undergone surgery for cervical syrinx-ACM type-I with craniocervical decompression, C1-C2 laminectomy and enlargement of foramen magnum with excision of fibrous band. During evaluation of the patient, his elbow was found swelling with mild pain on movement for which the patient was not concerned. Thorough evaluation confirmed the case to be an uncommon case of Charcot's arthropathy involving elbow associated with cervical syrinx without involving shoulder.

**Key words:** Syringomyelia, neuropathic arthropathy, Arnold-Chiari malformation (ACM).

### Introduction:

Charcot's joint or neurogenic arthropathy is a chronic progressive degenerative arthropathy associated with an underlying neurologic disorder. Charcot's arthropathy of elbow is relatively rare entity with a few case reported in the literature. Patients with Charcot's arthropathy present with a swollen erythematous joint usually in the setting of a sensory neuropathy. Common causes of Charcot's arthropathy include tebes dorsalis, syringomyelia, diabetes mellitus, alcoholism, amyloidosis, peripheral neuropathy including congenital sensory neuropathy<sup>1</sup>. Elbow joint is an uncommon site of neuropathic arthropathy.

Jean Martin Charcot first described neuropathic arthropathy in 1868 in a tebes dorsalis patient. After that

there has been much discussion about the cause and pathology of the condition. Sensory affection with mechanical disorders could produce the features of Charcot's joint. The affected joint lacks ability to respond *via* reflexes to abnormal stress. Consequently the subchondral bone of the involved joint disintegrates leading to joint collapse and considerable joint deformity. The disease is mostly of the hypertrophic type and is manifested as soft tissue swelling, subluxation, para-articular debris, osseous fragmentation and disorganisation<sup>2</sup>. The joints most commonly affected are the weight bearing joints. However hip and knee are affected in tebes dorsalis, ankles and feet in diabetes mellitus and shoulder in syringomyelia<sup>1</sup>. Although it is classically described as painless lesion in neuropathic joint one-third of the patients can have pain<sup>3</sup>. There may be history of injury recently or in distant past. The diagnostic features of neuropathic arthropathy includes "5Ds" i.e. debris, density (sclerosis), destruction, disorganisation and dislocation<sup>4</sup>.

The purpose of this report is to present an uncommon condition with affection of a non-weight bearing joint by Charcot's arthropathy which was also mild painful. A thorough evaluation is essential to diagnose such a condition which mimics and contrast neuropathic arthropathy.

### Case Report:

A 50-year-old man came to our department with the

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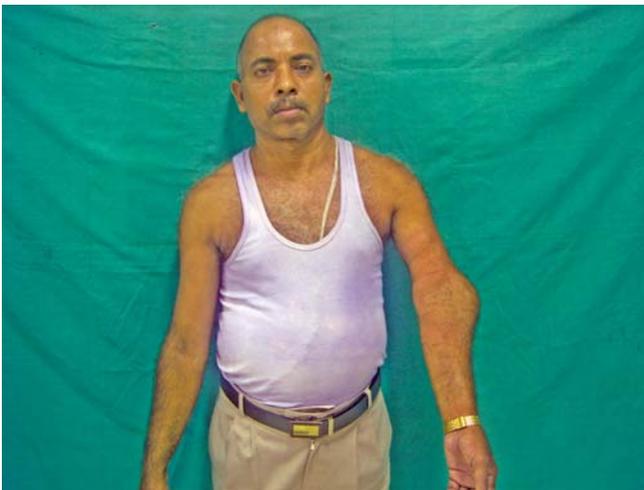
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complaints of progressive swelling and deformity of left elbow since last two and half months associated with pain on movement of elbow (Fig 1). He has difficulty in walking with clumsiness of the gait. His medical record shows he had pain neck radiating to left upper limb for 15years and low back pain for 6years. Pain was confined to back for one year subsequently radiated to left lower limb. Last 3years he had increased neck pain and back pain and found difficulty in walking and clumsiness of the gait. He was advised MRI of whole spine. MRI finding shows–syringomyelia with AC malformation type-I (Figs 2&3). Then he had undergone surgery for cervical syrinx-ACM type-I with craniocervical decompression C1-C2 laminectomy and enlargement of foramen magnum with excision of fibrous band. After surgery pain was relieved and gait improved. Since one year, increased intensity of pain and gait further deteriorated. He had occasional pain in his left shoulder also. Gradual swelling of elbow was marked with



**Fig 1-** Swelling and Deformed Left Elbow



**Fig 2-** MRI Showing Syringomyelia with AC Malformation

restricted joint range of motion. The patient was non-diabetic and normotensive and no history of sexually transmitted disease.

On examination, the left elbow was markedly swollen and fluctuant. Normal skin over the joint without any features of inflammation. On sensory evaluation, superficial sensations diminished in whole left upper limb. He was able to perceive pain sensation to some extent but unable to differentiate between sharp and dull pain. Pressure and vibration and cortical sensations were intact. Motor evaluation features included- tone normal for both upper limbs and in lower limb tone was MAS Gr 1. Voluntary motorfunction (VMC) was poor for elbow and poor to fair for shoulder and wrist. Deep tendon reflexes were not elicitable for biceps and triceps tendons, hypoactive on left upper limb and that of normal on right upper limb. Due to gross swelling and dislocation of elbow the apparent joint range motion of elbow was around 50°. Forearm was in supinated position. Passive pronation possible up to 45°. There was marked instability in varus and valgus test at elbow. Shoulder movements were restricted and painful at terminal range. Grip strength was poor to fair.

His x-ray of left elbow showed marked destruction of articular surfaces of bone ends, diminution of joint space, dislocation of joint, condensation of subchondral bone with fragmentation and intra-articular calcification with new heterotrophic calcification in surrounding tissue (Fig 4).

Laboratory studies and other investigations-Hb, DC, TLC, platelets, ESR, FBS, BUN, creatinine, Na, K, bicarbonates, chlorides values were within normal limit. Inflammatory markers like CRP, ESR were within normal range. VDRL test was negative.



**Fig 3-** MRI Showing Syringomyelia with AC Malformation

Management of neuropathic arthropathy is usually conservative, aiming towards reducing further articular damage by prevention of repetitive trauma. A number of surgical procedures have been described in literature, but none of them are satisfactory. On physiatrist point of view, goal setting was done considering functional outcome and vocation of the patient. The goal of management was directed towards improvement of gait pattern, hand function and to reduce the rate of deformity at lowest level. The patient had undergone a course of therapy and considerably improved his gait pattern. He was trained about simple modification of his activities of daily living. Protection of the joint that is susceptible to trauma and the same time useful joint movement for ADL achieved with use of special designed splint (Fig 5). A static splint with soft lining (Fig 6) was used as night splint to prevent progression of the deformity. Another splint i.e. elbow cage with free joint and pull

over strap (Fig 7) was fitted for day time use. With the use of that splint patient was able to drive motor cycle and rejoined back to his job.

### Discussion:

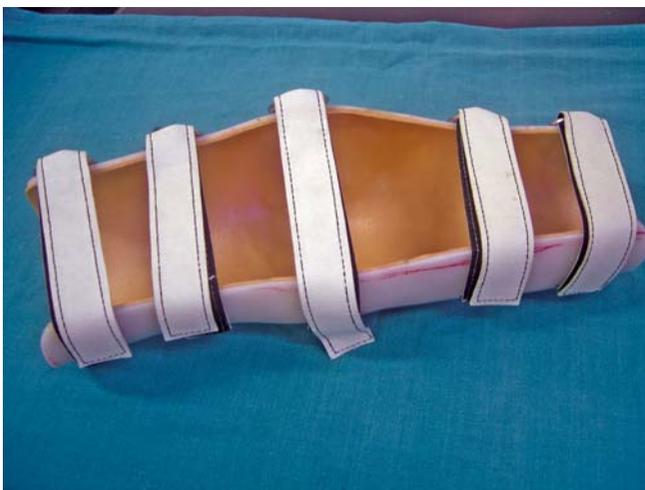
Neuropathic arthropathy of elbow is a rare condition. On literature review there are approximately 3-8% elbow neuropathic arthropathies out of all the cases of neuropathic arthropathy<sup>3,5</sup>. Most of the documents report only one or two cases except one study by Carl showing 5 cases of Charcot's arthropathy of elbow<sup>6</sup>. A variety of diseases had been reported in literature associated with neuropathic arthropathy of elbow out of which syringomyelia being the commonest one<sup>7</sup>. The current concept on aetiopathology includes the prominent role of misuse or abuse of insensitive joints. Loss of pain and proprioceptive sensations lead to relaxation of soft tissue structures around the joints which cause injury,



**Fig 4-** Straight X-ray Left Elbow Showing Features



**Fig 5-** Splint



**Fig 6-** Static Splint with Soft Lining



**Fig 7-** Splint for Day Time Use

malalignment and abnormal loading of the joints even with normal physiological loads of daily activities. Cumulative injury leads to progressive degeneration and disorganisation of articulation. Repeated trauma results in fibrillation and fragmentation of joint cartilage resulting in formation of loose bodies. Joint capsule often stretched beyond tolerance both by haemarthrosis and the stress on the joint.

The neurovascular theory describes active bone resorption by osteoclasts secondary to sympathetic dysfunction<sup>8</sup> and a neutrally mediated persistent hyperemia<sup>9,10</sup>. Radiologic findings can be classified as hypertrophic (productive) or atrophic (destructive). Neuropathic arthropathy of ankle, knee and elbow most often exhibits hypertrophic changes whereas the foot, hip and shoulder more commonly revealed atrophic changes<sup>10</sup>. MRI is the most effective modality for visualising a syrinx. Syringomyelia develops in 75%-85% of patients with a type I Arnold Chiari malformation<sup>11</sup>.

Management of neuropathic arthropathy varies from simple conservative methods to extreme surgeries. Several studies have demonstrated a beneficial effect of bisphosphonates in the treatment of neuropathic joints. Bisphosphonates reduce disease activity and bone turn over as a result of inhibition or apoptosis of activated osteoclasts<sup>13,14</sup>. Other treatment options include prosthetic replacements, resection arthroplasty or arthrodesis. Total joint replacement in neuropathic joint has bad outcomes because of associated adverse factors which include, lack of protective pain sensation and reflexes, presence of osteoporotic bone and laxicity of the surrounding ligamentous and muscular tissues. Non-operative treatment with the use of braces is probably the best solution for long-term management of these cases.

### Conclusions:

Neuropathic arthropathy of elbow is a rare condition. If neuropathic arthropathy is found in elbow as diagnosed from radiographic pictures, the underlying cause should be found out. The principle of management is to treat

the underlying disease and reduce the severity of the deformity and at the same time retaining the functional abilities. Conservative treatment with therapy and brace has good outcome especially for non-weight bearing joints affected with neuropathic arthropathy.

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