Bilateral Charcot arthropathy of shoulder secondary to syringomyelia: An unusual case report

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Abstract

Neuropathic arthropathy of the shoulder is a relatively rare disorder characterized by destruction of joint secondary to loss of sensory innervation. Bilateral Charcot arthropathy is an even rarer disorder, with very few cases reported in the English literature. We herein present a case of bilateral shoulder arthropathy secondary to syringomyelia with classical clinical and radiological findings. Radiological finding on one side was of resorptive type and resorptive mixed with productive on the other side.

Key Words

Charcot joint, shoulder, syringomyelia

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Ann Indian Acad Neurol 2012;15:202-4

Introduction

Neuropathic arthropathy, also known as Charcot joint, is a destructive arthropathy resulting from loss or diminution of proprioception, pain and temperature perception.[1-3] Patients with diabetes mellitus, syringomyelia and syphilis are particularly prone to this disease.[4] Diabetic patients tend to have involvement of foot and ankle, while knee is the common site in syphilis. Patients with syringomyelia commonly suffer with shoulder and elbow involvement.[5]

We conducted a Medline search with the keywords Charcot arthropathy shoulder, and found a total of 110 related articles. As such, Charcot arthropathy involving shoulder joint is an uncommon disorder, with less than 70 patients reported in the English literature.[6] Among them, we found only three cases where bilateral involvement was observed. Herein, we report a case that is felt to be of interest because of two reasons, one because of bilateral involvement with unusually late presentation and second because of resorptive appearance on one side and combined resorptive–productive appearance on the other side radiologically. To the best of our knowledge, this is the first case with bilateral shoulder involvement where one side showed resorptive and the other side showed mixed resorptive–hypertrophic destruction of joint.

Access this article online	
Quick Response Code:	Website: www.annalsofian.org
	DOI: 10.4103/0972-2327.99718

Case Report

A 62-year-old male presented to our outpatient department with painless swelling of both his shoulders and restricted movements of the bilateral shoulder joints. His condition began when he was approximately 35 years old and he started experiencing reduced sensation in the bilateral upper limbs. No medical treatment was taken and the patient went to some osteopath who gave him some oil for local application. Patient condition did not improve and he started experiencing reduced sensations in the bilateral lower limbs. Despite that, he did not seek medical attention and his condition gradually worsened. There was no history of significant trauma in the past. Physical examination revealed generalized swelling about the shoulder, more on the left side [Figure 1]. On the right side, there was abnormal motion, distal to where the shoulder joint would be expected . On the other hand, the left shoulder joint was found to be dislocated, with humeral head anterior to glenoid cavity, leading to significant restricted joint movement. On palpation, the shoulder joints were nontender. Bilateral shoulder movements were restricted. Active forward flexion was 40 on the right and 30 on the left side, abduction 30 on the left and 25 on the right side and internal rotation bilaterally up to the sacrum. He had 4/5 shoulder abductor strength and 4/5 shoulder flexor and extensor strength bilaterally. Range of motion for passive movements was significantly higher than on active movements, more on the right side. Passive movement was painful on terminal range of motions. His biceps

strength was 4/5, triceps strength was 5/5 while motor strength in bilateral distal extremities was 5/5. There was decreased sensation involving the entire upper extremities bilaterally and healed trophic ulcers found on the fingers and dorsal and ulnar border of the hand and forearm . The biceps, triceps and brachioradialis reflex were absent. There was muscle wasting in both upper limbs.

Patient was worked up and X-ray, complete blood counts,erythrocyte sedimentation rate,fasting blood sugar,venereal disease research laboratory test level of vitamin B12, electromyography (EMG) and nerve conduction velocity of both upper limbs, measurement of the crystal for gout and pseudogout, magnetic resonance imaging (MRI) of cervical and thoracic spine and abdominal ultrasonography were performed. Although the overlying skin temperature was normal, still joint aspiration was performed to rule out the remote possibilities of an infected joint. No organism grew on culture. Other than radiography and MRI, all the hematological investigations were within normal limits, on the basis of which syphilis and diabetes were ruled out. A purified protein derivative test was nonreactive. X-ray of right shoulder showed complete destruction of humeral head with fragmentation, which falls into the resorptive variant of Charcot shoulder [Figure 2]. X-ray of the left shoulder showed anterior dislocation of shoulder with fragmentation of head and heterotopic new bone formation [Figure 3]. No biopsy was performed. MRI (magnetic resonance imaging)of cervical and dorsal spine showed large syrinx from the 3rd cervical to the 3rd dorsal spine [Figure 4]. At that time, various modalities of treatment were considered. Limb elevation, shoulder abduction brace and nonsteroidal anti-inflammatory drugs were advised. His swelling gradually subsided after 10 days of conservative treatment. As instability and not pain was the chief complaint, a custom-made shoulder abduction brace was advised on both the sides and underwent intensive shoulder rehabilitation programme. After that, the patient returned monthly and remained asymptomatic for the next 2 years. At the time of writing, the patient was doing well on this conservative mode of therapy and performing his daily activities and was able to perform his self-care.

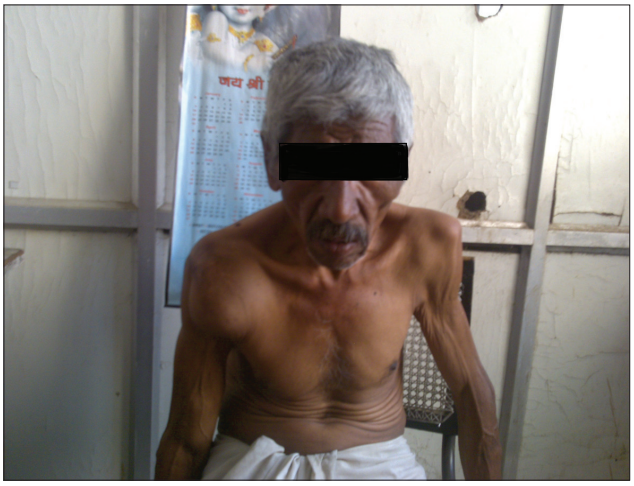

Figure 1: Clinical photo of the patient showing involvement of bilateral shoulder joint

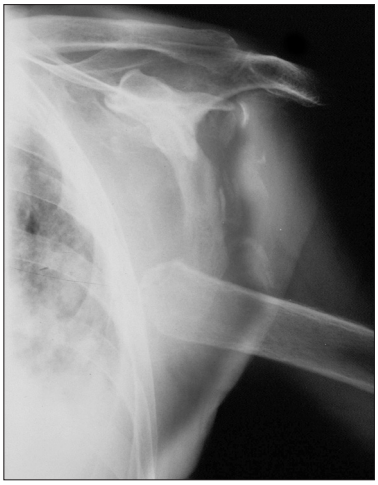

Figure 2: X-ray of the right shoulder showing resorptive changes with total destruction of the proximal humerus. Scapula and clavicle seem to be intact

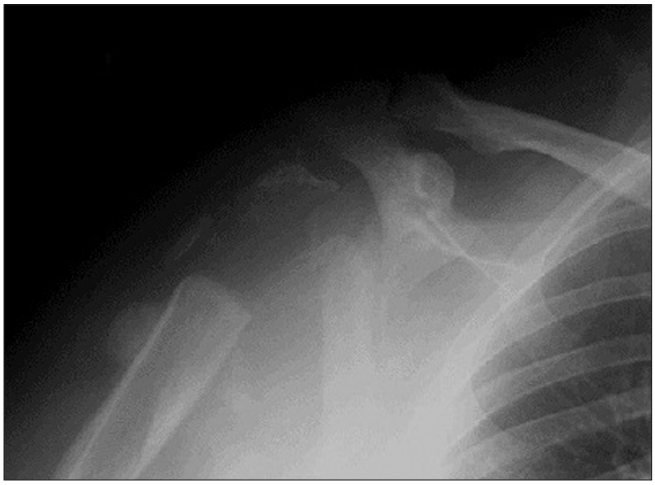

Figure 3: X-ray of the left shoulder showing hypertrophic dislocated shoulder joint

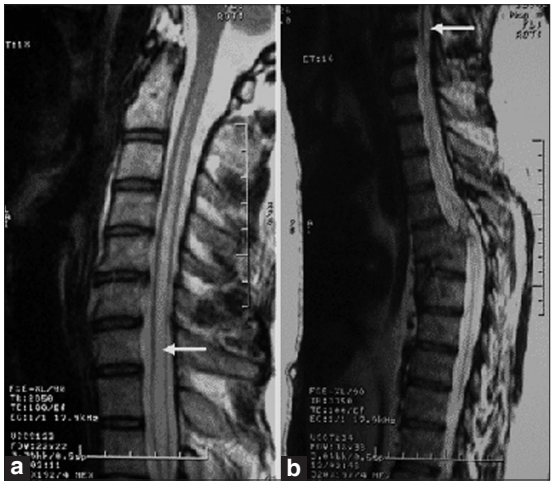

Figure 4: Magnetic resonance imaging showing syrinx in the cervical and dorsal regions

Discussion

Charcot arthropathy is located in the shoulder in only 5% cases. Meyer *et al*. stated that arthropathies develop in 25% of the cases of syringomyelia, and that 80% of syringomyelic arthropathies involve the upper limb. (Exact numbers were not reported in that study.)[7] There are two theories describing the pathogenesis of neuropathic osteoarthropathy. These are the neurotraumatic and the neurovascular theories. The neurotraumatic theory, first described by Johnson in 1967, involves repetitive trauma sustained by an insensate joint. The neurovascular theory, proposed by Allman and colleagues, describes active bone resorption by osteoclasts secondary to sympathetic dysfunction and a neurally mediated persistent hyperemia. If fractures and other forms of trauma are involved, this theory suggests that they occur secondarily.[4,7]

Neuropathic arthropathy of the shoulder often presents in a striking fashion, producing extensive and rapid destruction of the proximal aspect of the humerus and the glenoid. The differential diagnosis often includes primary and metastatic malignant tumor, tuberculous and microbial infection, and Gorham disease (vanishing bone disease). Syringomyelia is a disorder involving a fluid-containing cavity (syrinx) within the spinal cord. These cavities commonly occur in the lower cervical and upper thoracic segments, and the distension may propagate proximally. Its causes include congenital, traumatic, infectious, degenerative, vascular or tumor related.[2,3,8,9] MRI is considered the gold standard for visualization of a syrinx.

Syringomyelia is a potential cause of neuropathic osteoarthropathy of the shoulder, or "Charcot shoulder." The work-up of a patient with shoulder dysfunction should include a thorough history and physical examination and radiographic plain films. Other pertinent investigations such as CBC(complete blood counts), ESR(erythrocyte sedimentation rate), FBS(fasting blood sugar), VDRL(venereal disease research laboratory test), level of vitamin B12, EMG(electromyography) and nerve conduction velocity of both upper limbs, measurement of the crystal for gout and pseudogout and abdominal ultrasonography should be performed. Once a neuropathic joint has been diagnosed, its etiology should be pursued with aseptic joint aspiration to look for infection or tumor, and an MRI to evaluate for syringomyelia if the etiology remains in doubt. Syringomyelia may present as instability of the shoulder, and instability or frank dislocation is not unusual in the neuropathic shoulder.

In the present case, the unusual delay in presentation urged us to perform an extensive literature search and we found this interesting fact that our case has presented for the first time after the longest delay ever reported. No where in the literature has it been previously described that a patient who started experiencing neurological symptom of syringomyelia during his 20s first reported to the physician after 60 years of age.

Previous studies reported failure when arthrodesis was performed to treat neuropathic arthropathy of the shoulder, and concluded that this condition is a contraindication to arthrodesis.[3,10] Those authors also stated that synovectomy is not helpful and that a neuropathic shoulder should be treated nonoperatively, with an emphasis on the maintenance of function. The therapy for neuropathic arthropathy is conservative. Preventing trauma to the joint with proper splinting is key to treatment. Aspiration of large effusions and splinting will prevent further ligamentous laxity.[1,2]

We concur with these conclusions and agree that the maintenance of function, rather than immobilization, is the keystone of treatment of neuropathic arthropathy. We managed our patient conservatively, as suggested by the majority of the literature, and the patient was satisfied with the treatment. To our knowledge, this is the first report of bilateral Charcot arthropathy of the shoulders from the Indian subcontinent with an unusually delayed presentation. It is hoped that this case report will increase awareness of physicians about this destructive joint disease.

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How to cite this article: Panagariya A, Sharma AK. Bilateral Charcot arthropathy of shoulder secondary to syringomyelia: An unusual case report. Ann Indian Acad Neurol 2012;15:202-4.

Received: 19-09-11, **Revised:** 13-11-11, **Accepted:** 28-12-11

Source of Support: Nil, **Conflict of Interest:** Nil