A CASE OF MISSED UNILATERAL NEUROPATHIC ARTHROPATHY OF THE SHOULDER SECONDARY TO CERVICO-DORSAL SYRINX: A RARE PRESENTATION

S. M. Arvind Kumar¹, Chittaranjan K², Raj Kumar³, Krishna Kumar⁴

HOW TO CITE THIS ARTICLE:

ABSTRACT: A neuropathic arthropathy is a form of destructive arthropathy resulting secondary to loss of joint sensation due to various different etiological factors. It usually manifests without any pain but gross destruction of the joint. Usually seen in the weight bearing joints of the lower limb. It is highly rare for this entity to occur in the upper limbs with only very minimal cases reported. The diagnosis of the same is also difficult with many cases being misdiagnosed and treated for unrelated causes and land up with poor results. Here in we report a case of neuropathic arthropathy of the shoulder secondary to a cervico-dorsal syrinx misdiagnosed to be cuff arthropathy.

KEYWORDS: Neuropathic arthropathy, shoulder arthritis, Charcots shoulder, syringomyelia.

INTRODUCTION: Neuropathic arthropathy also known as Charcot’s arthropathy. It is a form of destructive arthropathy, secondary to loss of proprioception, pain and temperature sense in the involved joint and extremity. Neuropathic arthropathy is commonly seen to affect the lower extremities (ankles and knee), secondary to disorders such as diabetes. Neuropathic arthropathy of the shoulder is an uncommon disorder with only very few cases reported. The usual cause in cases of upper extremity Charcot’s arthropathy is syringomyelia, Hansen's disease, diabetes, alcoholism, cervical spondylosis.

Syringomyelia is a chronic, progressive process characterized anatomically by the development of a dissecting cavity in the grey matter that contains cerebrospinal fluid. Its main location is in the cervical spine and mostly affects young adults. There is a moderate correlation between the characteristics of the syrinx (length, disposition and morphology) and its neurological expression. Here we report a case of unilateral neuropathic arthropathy of the shoulder secondary to a cervico-dorsal syrinx which caused a diagnostic dilemma and a failed treatment.

CASE REPORT: This 40 yr. old male, cab driver by profession came with complaints of right shoulder swelling associated with restriction of movements for past 1 year. He attributes the symptoms following fall on out stretched hand 1 year ago. He had restricted mobility and progressive swelling of the shoulder with minimal pain. On examination he had an effusion of right shoulder, no tenderness was noted. Active range of movement'sabduction-30 degrees, flexion-40 degrees mostly scapula-thoracic movements, passive range of movements showed exaggerated movements. Clinically the shoulder joint was dislocated anteriorly and the sub acromial space was empty except for an effusion.

Neurologically motor power (shoulder excluded) was 4/5; paraesthesia from c3-t2 spinal segments and joint position sense was variable and not conclusive. MRI shoulder showed Antero-inferior subluxation of humeral head with complete tear of rotator cuff and retraction of tendons,
CASE REPORT

Antero-inferior capsule-labral tears. Lab investigations were all within normal limits. Past history patient had paraesthesia of right upper limb 5 years back. He was diagnosed to have a cervico-dorsal syrinx. Foramen magnum decompression with c1 arch removal was done. His neurological status remained the same post surgery till date. Our differential diagnosis with the correlation of history, clinical examination and MRI based evidence led us to:

1. Chronic Post traumatic unreduced dislocation with rotator cuff arthropathy.
2. Neuropathic joint.
3. Post septic sequelae.

Our diagnosis was more in favor of a chronic unreduced dislocation with a rotator cuff arthropathy (as the patient had attributed the symptoms after the fall). Patient was planned for a surgical reduction and stabilization of the shoulder. Patient was taken under GA, through a deltopectoral approach. Intra operatively there was minimal capsular remnant with no labrum visualized; the whole humeral head was eroded more than 50% with a large defect. Minimal degenerated rotator cuff remnants were seen retracted.

The articular surfaces were denuded, reduced and stabilized with k wires. The surrounding capsular and rotator cuff remnants were sutured around by osteosuturing techniques, so as to stabilize the shoulder. Post operatively patient was put under immobilization. The k wires had loosened by 5th week and were removed. Immobilization was continued. During the follow-up gradual increase in the effusion was noted and eventually his symptoms recurred as his pre-operative status.

DISCUSSION: Neuropathic arthropathy is a type of destructive arthropathy secondary to loss of proprioceptive sense in the joint. It is commonly seen in the lower extremities. It is very rare with only over 5% cases presenting as upper extremity Charcot’s. Meyer et al. stated that neuropathic arthropathy develops in 25% of the cases of syringomyelia, and among that 80% cases of syringomyelic arthropathy involve the upper limb commonly. Johnson ET al¹ & Jones ET al² suggested no surgical intervention is indicated successfully for a neuropathic arthropathy, the treatment strategy has always been conservative.

The variable options tried but without success include arthrodesis, synovectomy and alendronate therapy during the resorptive phase. Mainstay of conservative treatment is aspiration of effusion and splinting as required (Ata Can Atalar ET al,³ Ashok Panagariya ET al⁴). It is largely misinterpreted due to the unusual ways of presentation. As studies suggest it is often misdiagnosed for other disorders of the shoulder (cuff arthropathy/unreduced dislocation). And surgical procedures are carried out for the same, which invariably results in a failure of the procedure itself. The only form of treatment supported was a conservative approach with bracing/immobilization and a graduated schema of range of movements exercises (Dravaric ET al,⁵ Crowthers ET al⁶).

Here in our case misdiagnosis was due to the history of trauma, no further progressive neurological deficits from the time of decompressive laminectomy, delayed presentation of the patient, findings of MRI projecting like a cuff arthropathy. The rarity of the neuropathic shoulder led us to think in terms of a more common diagnosis of post-traumatic cuff arthropathy. The intra-op findings, failure of the procedure and recurrence of symptoms proved our mis-diagnosis.
According to Nick hatzis ET al\textsuperscript{7} often a neuropathic joint is misdiagnosed commonly especially that occurring at non weight bearing joints. Often the diagnosis is made on a retrospect thinking following failed surgeries and exclusion of causes.

REFERENCES:
CASE REPORT

Fig 1: Pre-operative clinical picture of the right shoulder.

Fig 2: Radiograph of the right shoulder-AP showing subluxated humeral head with gross arthrosis of the joint.

Fig 3 & 4: Gross effusion in the joint with fractures in the humeral head.

Fig 5 & 6: MRI of the patient showing the cavity of the cervico-dorsal syrinx.
Fig 7 & 8- Bare glenoid cavity with no capsulo-labral remanants.

AUTHORS:
1. S. M. Arvind Kumar
2. Chittaranjan K.
3. Raj Kumar
4. Krishna Kumar

PARTICULARS OF CONTRIBUTORS:
1. Associate Professor, Department of Orthopaedics, PSGIMS&R, Coimbatore.
2. Associate Professor, Department of Orthopaedics, PSGIMS&R, Coimbatore.
3. HOD, Department of Neurosurgery, PSGIMS&R, Coimbatore.
4. Junior Resident, Department of Orthopaedics, PSGIMS&R, Coimbatore.

NAME ADDRESS EMAIL ID OF THE CORRESPONDING AUTHOR:
Dr. S. M. Arvind Kumar,
Associate Professor,
Department of Orthopedics,
PSG IMS&R, Peelamedu – 04,
Coimbatore.
E-mail: arvindkumar.sm@gmail.com

Date of Submission: 06/03/2014.
Date of Peer Review: 07/03/2014.
Date of Acceptance: 18/03/2014.
Date of Publishing: 07/04/2014.