Neuropathic Arthropathy of the Shoulder - Case report of two cases with Review of the Literature

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Two cases of neuropathic arthropathy of the shouldder secondary to syringomyelia are presented. Radiographs revealed resorption of the humoral head in both patients. Magnetic resonance images revealed a syrinx of the central cord in both patients.

Key: Neuropathic arthropathy, Shoulder, Syringomyelia.

Introduction

Charcot in 1868, first described a chronic progressive arthopathy associated with tabes dorsalis. Subsequently neuropathic joint disease (Charcot's joint) has been described in patients with diabetes mellitus, leprosy, amyloidosis, acromegaly, yaws, spina bifida, meningomyelocoele, syringomyelia, myelopathic pernicious anaemia, peripheral neuropathy secondary to alcoholism and avitaminosis, peripheral nerve injury, spinal cord injury, multiple sclerosis, congenital insensitivity to pain and intra articular injection of steroids.[1,2,3,4,5] Neuropathic arthropathy of the shoulder is a rare disorder that has been described in fewer than sixty patients in the world literature.[1]

The present report describes the cases of two patients who had neuropathic arthropathy of the shoulder and reviews the literature on this topic, with emphasis on clinical and radiological manisfestations.

Case Reports

Case 1

A 50 year-old man who had no history of medical problems was evaluated for pain and swelling in his right shoulder for the past six years. There was no history of trauma, alcohol abuse and steroid therapy. The right shoulder was enlarged, crepitant and unstable but not tender, fluctuant or warm to palpation. Neurological examination revealed sensory hypoaesthesia in right upper limb but no motor deficit. The initial radiograph done six years back showed mild degenerative changes with predominant involvement of acromio clavicular joint (Fig. 1-A). Routine laboratory tests revealed normal findings. Repeat radiograph on referral to our institution, revealed significant destruction of the humeral head and acromion with purposeless new bone formation in the adjoining soft tissues (Fig. 1-B). A diagnosis of neuropathic arthropathy was considered and magnetic resonance images revealed a large syrinx of the caudad portion of hte cervical cord.

The patient was advised to continue active and passive exercises to maintian a maximum range of motion of the right shoulder.

Figure 1-A: Radiograph at initial presentation, showing mild degenerative changes involving the acromioclavicular joint.

Figure 1-B: Radiograph, made six years after the initial presentation, showing destruction of humeral head and acromion with periarticular soft tissue calcification.

Case 2

A 50 years old female was seen because of mild pain and swelling in the left shoulder for two months. Physical examination revealed cystic, fluctuant, non tender swelling around the left shoulder. The active movements of the left shoulder were grossly restricted in all directions but on passive movements, the affected shoulder was very unstable, hypermobile and crepitant. The neurological examination revealed sensory loss involving the fifth, sixth and seventh cervical dermatomes. The radiograph showed resorption of nearly fifty percent of the humeral head (Fig. 2). Findings from routine laboratory studies were normal. The diagnosis of neuropathic arthropathy was made and magnetic resonance imaging demonstrated a cervical syrinx. The patient was put on rehabilitation programme of exercise.

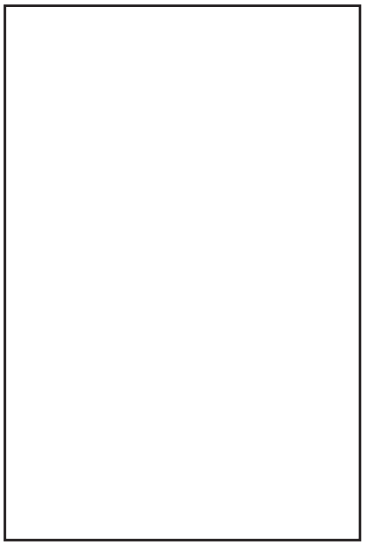

Figure 2: Radiograph showing resorption of nearly 50 percent of the humeral head.

Discussion

Neuropathic arthopathy is located in the shoulder in only 5 percent of the cases[1,6]. The differential diagnosis often includes primary and metastatic malignant tumor, tuberculosis and microbial infection, traumatic arthritis, chemical arthritis and Gorham disease[1,3].

The association of neuropathic arthropathy and syringomyelia was first noted by Sokoloff in 1892[5]. Neuropathic arthropathies develop in 25% of patients with syringomyelia, and 80% of arthropathies secondary to syringomyelia involve the upper extremities, with the shoulder and elbow being the joints most commonly affected[1,2,3]. Neuropathic arthropathy, as in our patients, may be the first clinical manifestation of syringomyelia[3]. Syringomyelia can be due to congenital, infective, traumatic, tumor, vascular or degenerative causes. The exact pathogenesis of neuropathic changes is unclear but it is believed to be due to destruction of aferent proprioceptive fibres and subsequent unrecognized repeated trauma to the joint[5].

Our review of the literature reveals that hte most frequent presenting symptom of neuropathic arthropathy is swelling, followed by pain and loss of motion[1,2]. Similar findings were observed in both our cases. Magnetic resonance imaging of the cervical spine demonstrated the presence of a syrinx in both patients in the present study. MRI has become the procedure of choice in the diagnosis of syringomyelia[1,2].

We are aware of the cases of two patients with known neuropathic arthropathy of the shoulder who were managed successfully with either arthrodesis or total shoulder arthroplasty[6]. However, there is a general consensus of opinion that arthrodesis or arthroplasty has got high failure rates in neuropathic shoulder and it should be treated non-operatively, with an emphasis on the maintenance of function[1,2]. We concur with these conclusions and agree that the maintenance of function, rather than immobilization, is the keystone of treatment.

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