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CASE REPORT

Neuropathic arthropathy of the shoulder and elbow associated with syringomyelia : A report of 3 cases

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We report three cases of neuropathic arthropathy secondary to syringomyelia. In two cases both the shoulder and elbow were involved. In the third case only involvement of the elbow was observed.

Keywords : Charcot joint ; elbow ; shoulder ; syringomyelia.

INTRODUCTION

Neuropathic arthropathy is a progressive, degenerative arthritis affecting one or more peripheral or vertebral articulations associated with an underlying central or peripheral neurological disorder. One of the most common causes besides diabetes mellitus and syphilis is syringomyelia, a chronic progressive degenerative or developmental disorder of the spinal cord. The joints most frequently involved are the shoulder and the elbow. In syphilis and diabetes the lower limbs are more frequently affected (*8, 10*).

Here, we report on three male patients with neuropathic changes in the upper limb due to syringomyelia. Interestingly, in two patients both the shoulder and ipsi- or contralateral elbow were affected ; in one patient only the elbow was affected. Reports of a concomitant neuroarthropathy of the ipsilateral shoulder and elbow, caused by syringomyelia are extremely rare (*16*). All our patients had clinical and radiological signs of syringomyelia, associated with a limited range of motion, swelling and pain in the affected joints. Initially, all deformed joints were treated conservatively and this resulted in an acceptable function with bearable pain. Two patients developed septic arthritis of the neuropathic joint, and surgical debridement was performed to eradicate the infection.

CASE REPORTS

Case 1

A 68-year-old right-handed male was admitted to our clinic in 2003 with a limited range of motion and swelling of his right shoulder and elbow. In 1975, he was seen by the neurologists because of pain in the right upper extremity, a progressive loss of sensation and a gradual loss of strength in his

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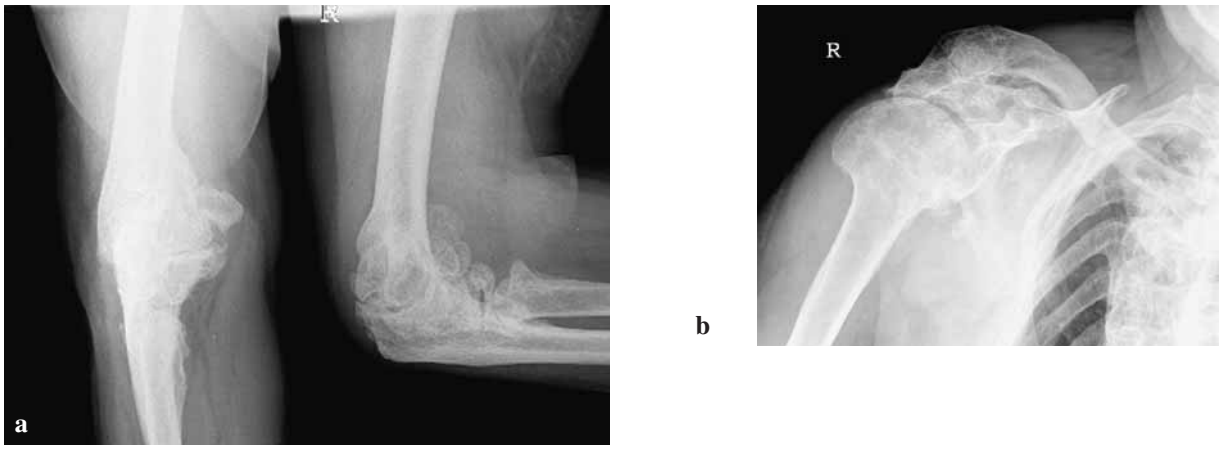

Fig. 1. — a) Neuropathic arthropathy of the right elbow with destructive joint changes, deep chondromatosis, fragmentation, subchondral sclerosis, loose intra-articular bone fragments and para-articular debris ; b) Radiograph of the ipsilateral neuropathic shoulder.

right arm. The loss of sensation caused severe painless burns on his right arm, while working in a bakery. Neurological findings included a loss of superficial touch and pain sensation in his right arm, combined with a right-sided thermo-analgesia extending from dermatomes C2-C6. Myelography confirmed the diagnosis of syringomyelia. Treatment consisted of radiation therapy.

On examination, shoulder abduction was 60°, forward elevation 45°, external rotation 0° and internal rotation to the level of the sacrum. At the elbow, extension was 20° flexion 85°. Pronation and supination were limited. The elbow was unstable but painless. Wasting of the intrinsic hand muscles was present. Radiographs revealed a destructive form of osteoarthritis with severe synovial chondromatosis (fig 1a, b). Physiotherapy was initiated to improve the range of motion.

Case 2

A 46-year-old baker was referred with pain and swelling of his left shoulder and elbow.

On examination, we found a swollen left elbow and shoulder. Shoulder abduction and flexion was 80°. Elbow flexion was 135° and extension was -20°. The elbow was unstable. Atrophy and diminished strength of the intrinsic hand musculature was noted together with bilateral loss of pain and thermal sensation in the C6-C8 dermatomes. Deep tendon reflexes were absent. Radiographs revealed fragmentation of the shoulder joint (fig 2a, b). Radiographs of the elbow showed destruction of the articular surfaces with fragmentation and subluxation of the ulno-humeral joint. Magnetic resonance imaging revealed a spinal syrinx from C2 to C7. The patient refused neurosurgical treatment. Physical therapy and a protective orthosis were prescribed. In 1990, he was admitted with a septic arthritis of his left shoulder following pneumonia. The shoulder joint was debrided and after one month of intravenous antibiotics, normalisation of the infectious parameters was observed.

Case 3

A 34-year-old truckdriver complained of pain, stiffness and swelling of his left elbow since two years. He also mentioned pain and limited mobility in his right shoulder. This was associated with a cramping sensation in the fourth and fifth digit of the left hand and sensory impairment in the left upper extremity. We noted 100° of flexion and full extension of the elbow. Abduction of the shoulder was 65°, forward elevation 85° and rotation was normal. Neurological examination revealed the presence of a segmental dissociative thermo-analgesia on the left side extending from the upper cervical region to the D2 dermatome. Deep tendon reflexes were absent in both the left and right upper

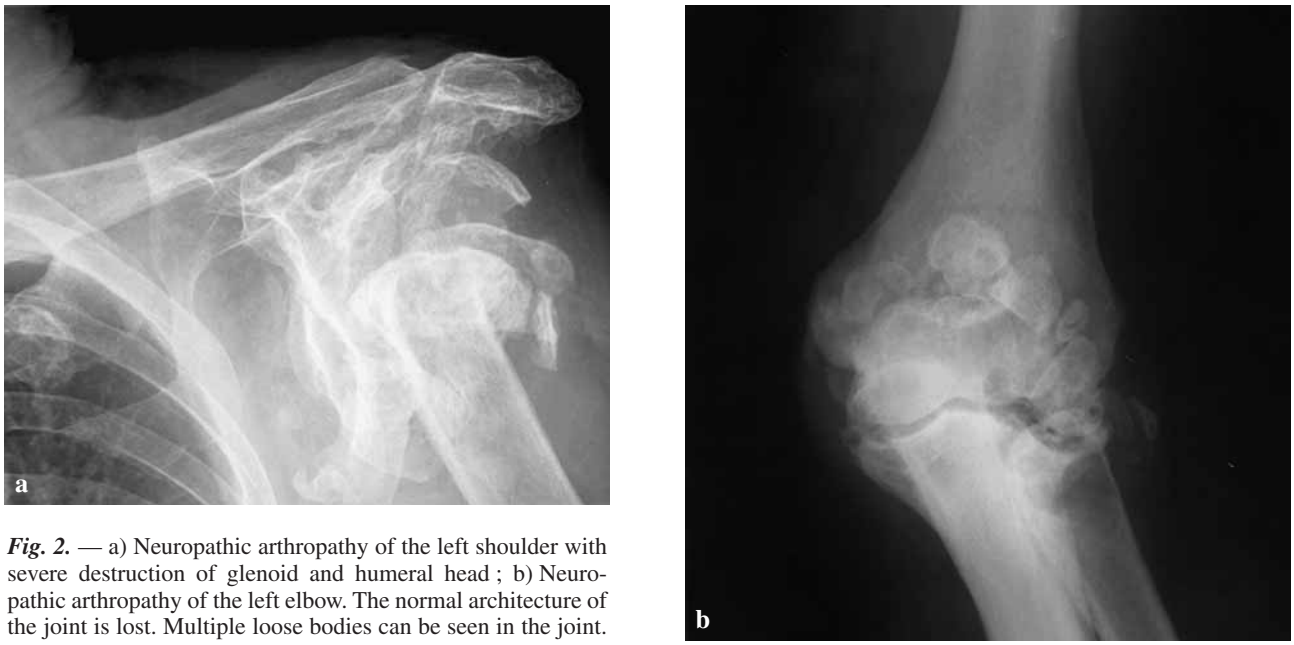

Fig. 2. — a) Neuropathic arthropathy of the left shoulder with severe destruction of glenoid and humeral head ; b) Neuropathic arthropathy of the left elbow. The normal architecture of the joint is lost. Multiple loose bodies can be seen in the joint.

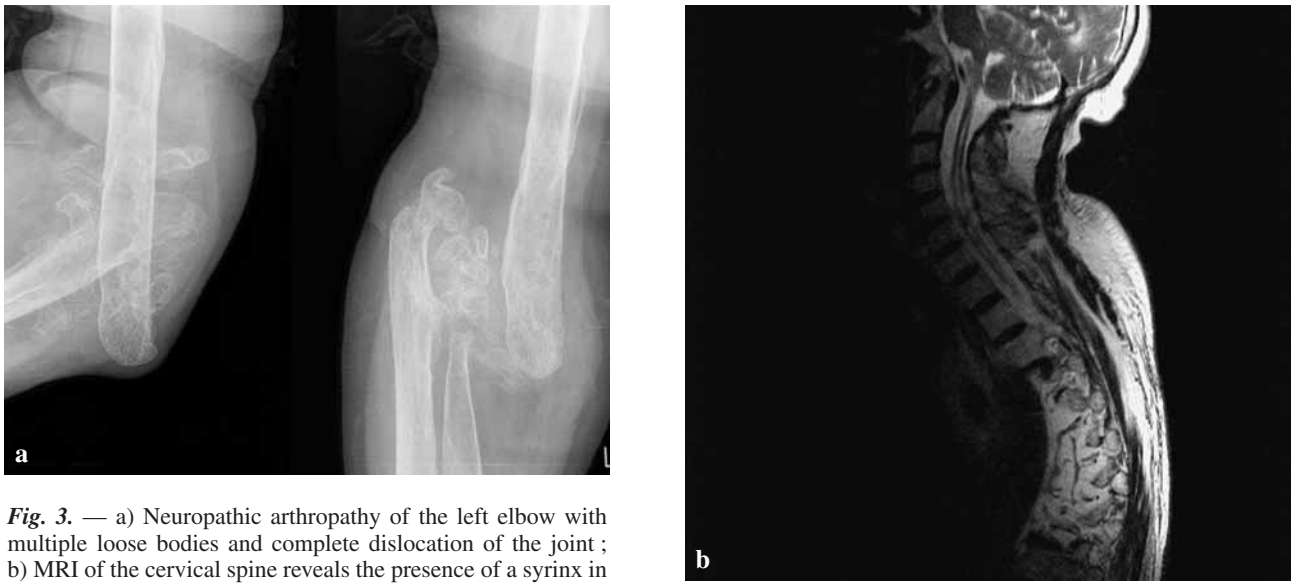

Fig. 3. — a) Neuropathic arthropathy of the left elbow with multiple loose bodies and complete dislocation of the joint ; b) MRI of the cervical spine reveals the presence of a syrinx in the spinal cord.

extremity. Radiographs revealed minimal degenerative changes in the glenohumeral joint. A tentative diagnosis of a beginning neuropathic arthropathy was made and a protective rehabilitation program was started. The shoulder pain subsided and mobility improved. Radiographs of the elbow revealed total destruction of the joint with fragmentation of the articular surfaces and the presence of intra-articular loose bodies (fig 3a). A tonsillography was performed and the presence of a Chiari type I malformation was diagnosed, and a decompression of the posterior fossa was performed. An MRI scan revealed the presence of a syrinx in the cervical spine (fig 3b). The elbow was treated

conservatively with the aim to maintain the range of motion. Subsequently the patient had several episodes of septic arthritis of the left elbow. Multiple debridements were performed and an Ilizarov frame was used to immobilise the elbow. In this way we were able to eradicate the infection.

DISCUSSION

We report three cases of neuropathic changes in the upper limb secondary to syringomyelia. Up to 25% of cases of neuropathic arthropathy are caused by syringomyelia but the combination of shoulder and elbow involvement in the same patient is very rare (*10, 16*). The syrinx interrupts the decussating fibers of the lateral spinothalamic tract that mediates pain and thermal sense while sparing fibers that mediate the sensation of deep touch. This causes a dissociated segmental anaesthesia over the neck, shoulder and arm in a cape or hemicape pattern. Loss of deep tendon reflexes and weakness associated with or without atrophy of the upper limb musculature can also be present. In cases with extensive cavitation, weakness, ataxia and loss of proprioception of the legs may develop (*9*). A neuropathic joint disease may develop early or late in the course of syringomyelia. Most often, there is involvement of the shoulders and elbows ; arthropathy of the hand, wrist or cervical spine is noted less frequently (*5, 6, 14*). Clinically, the neuropathic joints are characterised by swelling, pain and deformation together with a limited range of motion (*4, 5*). Early in the course of the disease, joints can be warm and erythematous, showing the existing hyperaemia and possibly difficult to differentiate from a septic arthritis. Less frequently, instability of the shoulder can be the presenting symptom of syringomyelia. Although neuropathic joints have classically been described as being painless, up to one third of the patients can have pain and remarkably, the pain is less intense than one should suspect based on clinical and radiographic findings. Often, a neuropathic arthropathy of the shoulder or elbow progresses slowly, however rapid progression occurring over months or even weeks has been described (*13, 15*).

Joint effusion, soft-tissue swelling, narrowing of the joint space with subchondral sclerosis and osteophytes are possible early radiographic findings of a neuropathic arthropathy and thus a differentiation between a primary osteoarthrosis and neuropathic arthropathy is often difficult, especially in the early stages. A neuropathic arthropathy can be classified as atrophic or hypertrophic (*1*). The atrophic form is characterised by a massive bone resorption accompanied by total disintegration of the joint which causes instability. The hypertrophic form is characterised by severe joint destruction, periarticular new bone formation, osteophytes, sclerosis, fractures and osseous debris which finally leads to a loss of normal joint architecture. Some authors suggest that both forms are only different stages in the natural course of the disease (*11*). Especially in syringomyelia, atrophic changes in the shoulder co-exist with hypertrophic changes in the elbow.

Several theories are postulated regarding the exact pathogenesis of neuropathic arthropathy. Two main theories have been put forward (*3*). Initial damage to the joint is a result of damage to the autonomic nerves and subsequent disruption of the normal neurovascular reflexes around the joint. This results in hyperaemia and activation of osteoclasts. The resulting bone resorption makes the joint prone to pathological fractures (‘neurovascular theory’). Alternatively, the loss of protective somatic muscular reflexes can lead to repetitive trauma and ultimately to joint destruction (‘neurotraumatic theory’). Jeffcoate *et al* (*7*) state that an initial trauma may be sufficient to activate an inflammatory cascade resulting in an activation of osteoclasts leading to bone resorption and fractures.

The primary goal of conservative treatment is reducing further articular damage by prevention of repetitive trauma. Aspiration of large effusions will prevent further ligamentous laxity. Recently, several studies have demonstrated a beneficial effect of bisphosphonates in the treatment of neuropathic joints. Bisphosphonates reduce disease activity and bone turnover as a result of inhibition or apoptosis of activated osteoclasts (*2, 12*). When conservative treatment is not successful, surgical interventions

can be considered. Treatment options include prosthetic replacements, resection arthroplasty or arthrodesis. Performing an arthroplasty of the neuropathic joint is not without risks due to the lack of protective pain sensation and reflexes, the presence of osteopenic bone and the weakness of the surrounding ligamentous and muscular tissues. This causes high stresses on the implanted components and as a consequence there is a high failure rate caused by septic loosening and periprosthetic fractures. Despite the occasional good result after surgical treatment a non-operative treatment with the use of braces is probably the best solution for long-term management of these cases.

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