Neuropathic arthropathy caused by syringomyelia in different joints and lesion of brachial plexus at right upper extremity: A case report

Ayşe Ekim*, Onur Armagan*

ÖZET
Siringomelye bağlı farklı eklemlerde nöropatik artropi ve sağ üst ekstremitede brakial pleksus lezyonu: Olgu sunumu
Anahtar kelimeler: Siringomyeli, nöropatik artropati, brakial pleksus

SUMMARY
Syringomyelia is characterized by slow progression; the joints involved most frequently are the shoulders and elbows. Neuropathic arthropathy of the hand is quite rare. Hereewith, we present a case of neuropathic arthropathy (NA) of the joints in the upper limb secondary to Syringomyelia. Atypical features of the case included NA of the shoulder, elbow and metacarpophalangeal joint in the same upper limb. These atypical features superimposed shoulder subluxation and brachial plexopathy which diagnosed by electrophysiologic studies. To the best of our knowledge, our study is the first of its kind that reports a patient who had all the abovementioned symptoms in the same upper limb.
Key words: Syringomyelia, Neuropathic arthropathy, Brachial plexus.

(*) Eskisehir Osmangazi Universitesi Tip Fakultesi, Fiziksel Tip ve Rehabilitasyon Anabilim Dalı, Eskisehir
Buşururu adresi:
Uzm. Dr. Ayşe Ekim, Eskisehir Osmangazi Universitesi Tip Fakultesi, Fiziksel Tip ve Rehabilitasyon Anabilim Dalı, Meşelik 26480 Eskisehir
Tel: (0 222) 2392979-2400  (0 222) 2392979-2400  e-posta: drayseekim@yahoo.com.tr

(*) Department of Physical Therapy and Rehabilitation, Eskisehir Osmangazi University Medical Faculty, Eskisehir
Correspondence to:
Ayşe Ekim MD, Department of Physical Therapy and Rehabilitation, Eskisehir Osmangazi University Medical Faculty, Eskisehir, TURKEY
Tel: (+90 222) 2392979-2400  (+90 222) 2392979-2400  e-mail: drayseekim@yahoo.com.tr

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Introduction

Neuropathic joint disease is a progressive degenerative arthritis with destructive and productive articular abnormalities and usually loss of a pain and prospective sensation (Williams 1990). There is a progressive disorganization of architecture in the insensitive joint, leading to painless joint swellings with radiological evidence of pronounced bone destruction as well as new bone formation in abnormal sites.

Since Charcot's original description of the disease in 1868, the list of its known causes has grown significantly, with syringomyelia one of the primary causes of neuropathic osteoarthropathy (Delano 1946). Syringomyelia is a degenerative disorder of the spinal cord characterized by an abnormal longitudinal cavitation (syrinx) filled by cerebrospinal fluid in the central canal. It has been described as a cause of Neuropathic arthropy (NA) secondary to syringomyelia often affects the upper limb joints (de Sausa Neves et al. 2005). In particular, it predominantly involves shoulders and elbows. However, involvement of the hand is quite uncommon.

We report a syringomyelia case with concurrent involvement of various joints in the same limb, apart from brachial plexopathy. His presenting signs were shoulders, elbows and metacarpopha langeal NA. He also had shoulder subluxation and in the same upper limb caused by syringomyelia. There are already known to be cases with neuropathic artropathy involving a single joint such as the shoulder and the elbow due to syringomyelia, apart from rare reports of shoulder subluxation, brachial plexopathy, and NA of the hand secondary to syringomyelia. However, we found no other case with the same aforementioned features concurrently seen in the same upper limb.

Case Report

A 54 year-old man referred to our clinic with limited movement of the right shoulder, elbow joint and painless swelling of the elbow joint and 3rd
metacarpophalangeal joint on right upper side. His history revealed that he had developed subluxation in the right shoulder due to lifting heavy loads two months prior to his admission. It was also determined that he had weakness in his right upper limb, accompanied by loss of pain and temperature sensations that had been continuing for the past 10 years. Also, he had been having a painless limited movement in the elbow joint over the last 3 years. His records showed that he was first examined in the department of neurosurgery in 1998. His magnetic resonance imaging (MRI) of the cervicothoracic spine revealed a syrinx extending from approximately cervical-2 (C-2) to thoracal-4 (T-4) vertebral levels (Figure 1). He was diagnosed with syringomyelia and was therefore operated on.

Investigations revealed that routine urine examination and blood counts were normal. Fasting blood sugar was 81 mg%, blood Venereal Disease Research Laboratory (VDRL) was non-reactive and the X-Ray chest normal. He was not a diabetic, and gave no history of exposure to sexually transmitted diseases.

On examination, the right shoulder joint active range of motion was limited; however, its passive range of motion was normal. The right elbow joint was markedly painless swelling and passive/active range of motion was limited. On sensory examination, the patient reported diminution of pain and touch of all the dermatomes on the right upper limb. The patient had marked weakness in his right shoulder muscles when compared with the other muscles in the left upper
extremity. Deep tendon reflexes were hypoactive in the right limb. On the other side, his reflexes were normal. His electroneuromyography of the right upper limb showed complete lesion in the upper and lower branches of the brachial plexus. However, his X-Ray of the right shoulder joint showed marked destruction of articular surfaces bone ends and severe anterior shoulder subluxation (Figure 2). Also, X-Ray of the right elbow joint established marked destruction of articular surfaces, diminution in joint space, condensation of subchondral bone with fragmentation and intra-articular calcification with new heterotopic bone formation and in X-ray 3rd metacarpophalangeal joint determined diminution and destruction in joint space, new bone formation within the joint cavity (Figure 3, 4).

Based on our radiological, electrophysiological and clinical findings, we diagnosed our patient as having developed neuropathic arthropathy in the joints of the right shoulder and elbow as well as in 3rd metacarpophalangeal joint secondary to syringomyelia, all coexisting in the same upper limb. He also had shoulder subluxation and brachial plexopathy. The patient was hospitalized in our clinic. He was given rehabilitation apart from being a physical therapy.

Discussion

Neuropathic arthropathy is a feature of chronic neurologic illness such as syringomyelia (25%), diabetes mellitus (0.16 to 2.5%) and tabes dorsalis (5 to 10%). Syringomyelia is a rare chronic disorder of the spinal cord or brainstem, usually characterized by slowly progressive brachial amyotrophy, dermatomal sensory loss in the shoulder or arms, and lower extremity pyramidal signs (Finlayson 1988, Mancall 1984). Although the
lesion is often congenital, symptoms usually appear in the third or four decades (Wisoff 1988, Tator and Briceno 1988). However, 80% of joint disorders associated with syringomyelia usually present themselves in the upper extremity (Bruckner and Howell 1972). No study has so far reported an interesting coincidence of neuropathic osteoarthropathy of the shoulder, elbow and metacarpophalangeal joint, apart from brachial plexopathy secondary to humeral subluxation associated with syringomyelia in the same upper limb.

It is well-documented that, in syringomyelia, the shoulder is the most commonly affected joint among the upper limb joints, the elbow being the next common (Barnett et al. 1973). However, only a few studies have reported involvement of the hand and the wrist involvement in syringomyelia. There are only two case reports reporting NA of the hand and the wrist secondary to syringomyelia. One of these cases had concurrent Rheumatoid Arthritis (RA), for which reason we think the hand involvement could partly be due to RA.

The pathogenesis of neuropathic joints has been the subject of discussion by several authors (Aggarwal et al. 1975, Basu 1972, Stendler 1931). The basic factor seems to be lack of appropriate sensory input from the joint. Abolition of proprioceptive and/or sensory impulses from the joint

Figure 4
leads to its exposure to unusual trauma for a prolonged time. Repetitive trauma results in fibrillation and fragmentation of the joint cartilage resulting in the so-called loose bodies. The joint capsule is often stretched beyond tolerance both by the hemarthrosis and by the stresses on the joint. Apart from this, there is hyperemic bone resorption and softening and the resultant atrophic bone is traumatized with ease. The result is a vicious circle which may go on until the joint gets completely destroyed (Bhaskaran et al. 1981). In this setting, the daily stresses of normal movement produce injury, malalignment and abnormal joint loading. Cumulative injury leads to progressive degeneration and disorganization of the articulation. In neuropathic arthropathy, major joint changes include marked bone destruction, soft tissue swelling, joint space narrowing, condensation of subchondral bone with fragmentation and intra-articular calcification with new heterotopic bone formation and severe joint subluxation (Drvaric et al. 1988).

Besides that, there was a clear shoulder subluxation and brachial plexopathy in our case. To date, there has been only one case reported with brachial plexus lesion and shoulder subluxation secondary to syringomyelia (Singer et al. 1992). We think that our patient, who already had shoulder NA, developed shoulder subluxation and brachial plexopathy most probably due to lifting heavy load, as was reported by the patient himself. Although the single case report in the literature has not stated any reason for shoulder subluxation and brachial plexopathy, we speculate that our case developed these pathologies due to loss of sense. It is well known that loss of the protective sensations of pain and proprioception in joint with neuropathic osteoarthropathy leads to relaxation of the supporting structures and chronic instability of the joint (Veilleux et al. 1987, Peioglou-Harmoussi et al. 1986).

Charcot arthropathy secondary to syringomyelia is common. However, concurrence of such pathologies as multi-joint involvement due to syringomyelia, in addition to shoulder subluxation and brachial plexopathy, in the same limb have not been reported up to the present time. As was true of our case, such patients are susceptible to traumas due to loss of sense. However, we believe that through some means of protection, prognosis of the disease could be delayed or prevented. We, therefore, suggest that patients with syringomyelia should be informed about possible traumas as part of their treatments.

References