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Neuropathic arthropathy of the shoulder associated with syringomyelia: a report of six cases

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Here, we report a series of 5 patients (6 shoulders) diagnosed with neuropathic arthropathy of the shoulder joint in our clinic between 2005 and 2008. Initial diagnosis, previous treatment, and radiological and clinical follow-up findings were reviewed. The mean age at diagnosis was 44.2 years. Four patients had unilateral and 1 patient had bilateral involvement. The presenting symptoms were pain, swelling, and loss in range of motion. Active forward flexion and abduction ranged from 0° to 90°. Hypoesthesia and loss of temperature sense was evident in 3 patients. Radiographs showed massive osteolysis of humeral head and glenoid process, and magnetic resonance imaging showed periarticular fluid collection, and degeneration at the rotator cuff and shoulder joint, resembling chronic septic arthritis or sarcoma. Biopsy was performed in 4 patients before definitive diagnosis, and synovial hypertrophy and necrotic bone was found. Two patients had a history of operated cervical syringomyelia, and the remaining 3 patients were later diagnosed to have syringomyelia and referred to neurosurgery clinic, where 2 of those were operated. Four patients were followed-up with symptomatic therapy, and 1 patient underwent an unsuccessful shoulder arthroplasty in another clinic. As a conclusion, neuropathic arthropathy of the shoulder is rare, and correct diagnosis is possible by careful physical and neurological examination and pathologic evaluation when needed.

Key words: Neuropathic arthropathy; shoulder joint; syringomyelia.

Neuropathic arthropathy is a type of chronic degenerative arthropathy which is associated with sensory loss at the involved joint. Neuropathic arthropathy can be seen in many diseases which lead to neural damage, most commonly in diabetes mellitus, syringomelia, and tabes dorsalis. In diabetes mellitus, foot and ankle joints are most commonly involved, whereas in syringomyelia, upper extremity involvement is seen, with shoulder joint being the most commonly involved site. Neuropathic arthropathy is seen in 25% of all cases with syringomyelia, and 80% of those occur in the upper extremity.[1-3] Overall, the shoulder is involved in 5-6% of patients with neuropathic arthropathy.[4]

The clinical findings in neuropathic shoulder arthropathy are variable. Usually, symptoms related with shoulder manifest earlier than neurological symptoms, and patients first present to orthopedic clinics.[1]

Our aim in this study was to present six shoulders of five patients, who had the diagnosis of neuropathic arthropathy of the shoulder at our clinic, and point out important points that should be kept in mind for diagnosis, treatment, and follow-up.

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Case reports

Six shoulder joints of 5 patients (3 females and 2 males) were diagnosed with neuropathic arthropathy of the shoulder in our clinic between 2005 and 2008 (Table 1). A retrospective analysis of the patients was carried out by the help of archive data; in addition, the patients were called for last follow-up physical examinations, and control radiographs and shoulder and cervical spinal magnetic resonance imaging (MRI) were obtained. The mean age was 44.2 years (range 35-54 years). Initial complaint was pain and swelling at the shoulder in 2 patients, shoulder pain in 2 patients, and swelling at the shoulder in 1 patient. The mean time period between the initiation of symptoms and detection of osteolysis at the shoulder was 4.3 weeks (range 1 week-3 months). The patients were followed up for a mean duration of 26 months (range 12-37 months).

Case 1

A 39-year-old male presented with pain at the left shoulder and swelling in the left forearm after a strain of the left shoulder. He was examined by state and university hospitals at his hometown, and a preliminary diagnosis was malignant mass at the shoulder after MRI and Tc-99 m bone scintigraphy studies. A tru-cut biopsy was performed, and compact bone spicules showing ischemic and reactive changes were detected. The patient was referred to our clinic one month after initial presentation; on physical examination, active forward flexion at the left shoulder was 45°, internal rotation was at the gluteal region, and there was no active external rotation. On neurological examination of the upper extremity, no motor or sensory deficit was detected. Deep tendon reflexes in the lower extremities were hyperactive bilaterally, and there was clonus at the Achilles tendon. On radiography, osteolysis at the humeral head and glenoid were noted (Fig. 1). The preliminary diagnosis at our clinic was neuropathic arthropathy of the shoulder. Cervical MRI was obtained to search for the etiology (Fig. 2). MRI revealed Chiari type I malformation and syringohydromyelic cavity at all cervical segments down to T8 (Fig. 3). To rule out other diagnostic possibilities, an open biopsy was performed at the shoulder joint, and atrophic synovium, focal chronic inflammation, and chronic synovitis were found. No mycobacterial or other organisms were isolated from cultures. After assessment of all findings, the patient was diagnosed with neuropathic shoulder arthropathy and was referred to the neurosurgery clinic for the treatment of syringomyelia, where cervical laminectomy and decompression was performed. The neuropathic shoulder joint was followed conservatively. At 9-month follow-up, there was total osteolysis of the humeral head. At 2-year year follow-up, the patient was referred to the neurosurgery clinic with a new MRI for check-up. On neurosurgical assessment, it was found that syringomelia persisted, but there was circulation of cerebrospinal fluid (CSF), the extent of the cavity was smaller, and there was no need for a new operation. At the 30-month follow-up, it was found that osteolysis had not progressed (Fig. 4). On physical examination, passive forward flexion and abduction was 90°, and internal rotation was at the infrascapular region. A shoulder arthrodesis was decided for the patient at that time. At 3-year follow-up, forward flexion was 120°, abduction was 120°, external rotation was 20°, internal rotation was at lower lumbar region, and there was no pain at the shoulder joint (Fig. 5). As the patient's complaints had decreased and he preferred a mobile shoulder joint for work, plans for the operation were cancelled, and conservative follow-up was continued.

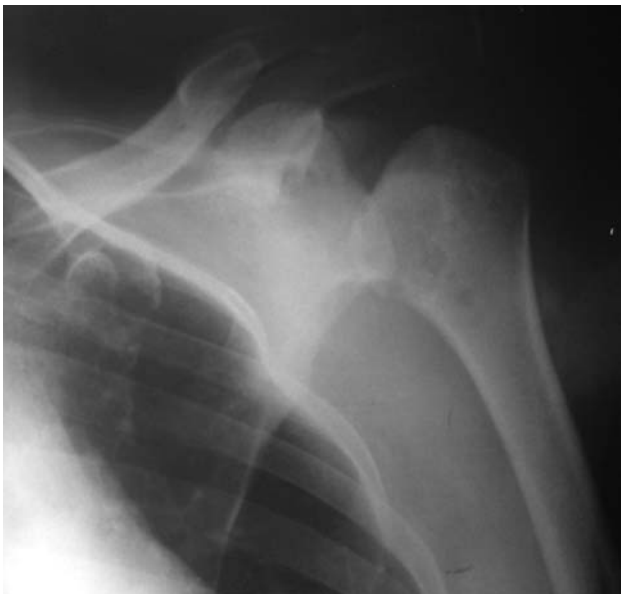

Fig. 1. Radiograph of case 1 at initial presentation.

Table 1
Mechanisms of injury according to activities

Patient no	Age	Sex	Side	Initial symptoms	Physical examination at presentation	Neurological findings at presentation	Previous procedures	Biopsy findings	Treatment	Follow-up	Physical examination at last follow-up
1	39	Male	Left	Pain, forearm swelling	Forward flexion 45°, no external rotation, internal rotation at the gluteal region	None	Tru-cut and open biopsy	Focal chronical inflammation at atrophic synovium, compact bone spicules showing ischemic and reactive changes	Suboccipital craniectomy C1 laminectomy, decompression	37 months	Forward flexion: 120°, external rotation 20°, internal rotation at lumbar region
2	47	Female	Left + right	Pain at right shoulder, swelling at right elbow	Right shoulder: forward flexion 95°, external rotation 30°, internal rotation at gluteal region; left shoulder: full range of motion	Hypoesthesia at right arm and shoulder, diminished temperature sense, bilateral claw hand	Tru-cut biopsy	Fat and fibrohyalin connective tissue with margins showing myxoid changes	Decompression (20 years ago)	12 months	Right shoulder: forward flexion 100°, external rotation 40°; internal rotation at lumbar region; full range of motion at the left shoulder
3	46	Male	Right	Pain	Forward flexion 30°, no external rotation, no internal rotation	None	Tru-cut biopsy, debridement, resection arthroplasty, tumor prosthesis	Necrotic bone lamellae neovascular-ization	None	31 months	Forward flexion: 20°, no external rotation, no internal rotation
4	35	Female	Left	Pain, swelling	Forward flexion 30°, no external rotation, internal rotation at gluteal region	Hypoesthesia at the shoulder, muscle strength around the shoulder 2/5	Tru-cut and open biopsy, resection	Villous hyperplasia at synovium, neovascularization, bone and cartilage islets, fibrinoid necrosis	Suboccipital craniectomy, C4 laminectomy, posterior fossa decompression, zoledronic acid	37 months	Forward flexion: 80°, no external rotation, internal rotation at lower lumbar region
5	54	Female	Left	Pain, loss in range of motion	Forward flexion 10°, no external rotation, no internal rotation	Hypoesthesia at the shoulder, muscle strength 3/5	None	-	Posterior fossa decompression, T12-L1 laminectomy	13 months	Forward flexion 30°, no external rotation, internal rotation at lower lumbar region

Case 2

A 47-year-old female patient presented to the orthopedics clinic complaining of swelling at the right elbow and decreased range of motion at the right shoulder for 2 weeks. Past medical history revealed that she was operated for cervical syringomyelia 20 years ago. On physical examination of the right shoulder, abduction was 80°, forward flexion was 95°, internal rotation was at the gluteal region, and external rotation was 30°. There was full range of motion at the left shoulder. Neurological examination showed hypoesthesia and loss of temperature sense at the right shoulder and arm region. Flexion contracture was present at the fingers of both hands. Plain radiographs showed osteolysis at the right humeral head and glenoid and early signs of osteolysis at the left shoulder. MRI of the right shoulder revealed marked fluid collection, synovial thickening, and a large lobulated, septate cystic formation. Tc-99 m whole body bone scan displayed increased osteoblastic activity at both shoulders, with the right shoulder having a marked activity. At first visit to the hospital, a Tru-cut biopsy was performed, which displayed lipid and fibrohyalin connective tissue with a structure showing mixoid changes at the margins. Cervical MRI revealed Chiari type I malformation and cervical syringomyelia. The combined results of radiological findings, physical examination, and past medical history led to the diagnosis of bilateral neuropathic arthropathy of the shoulder due to syringomyelia. The patient started conservative follow-up. Swelling improved with antiinflammatory treatment. At 12-month follow-up, active forward flexion of the right shoulder was 100°, internal rotation was at lumbar region, and external rotation was 40°.

Case 3

A 46-year-old male patient presented to our clinic with right shoulder pain which had been present for one week. At physical examination, active forward flexion was 30°, and there was no active internal or external rotatation. Plain radiographs showed osteolysis at the humeral head. A Tru-cut biopsy was performed to evaluate the initial diagnostic impression of a malignant mass, but the only finding was necrotic bone lamellae and neovascularization. The diagnosis of neuropathic arthropathy of the shoulder was considered; MRI of the cervical region was obtained, which showed a syringohydromyelic cavity at the cervicothoracal region, between C1- T6 levels. The patient was referred to the neurosurgery department, but surgical treatment was not recommended. The patient began conservative follow-up for neuropathic shoulder arthropathy, but was lost to

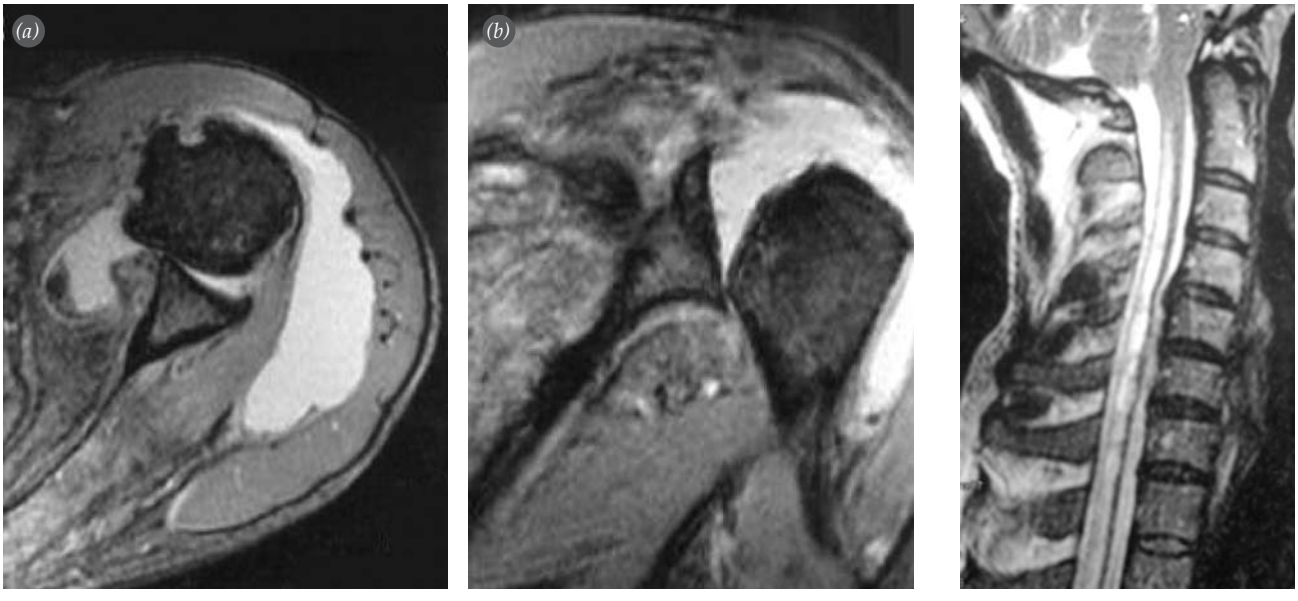

Fig. 2. (a, b) Shoulder MRI sections of case 1 at initial presentation.

Fig. 3. MRI section of case 1 showing syringomyelia.

follow-up shortly thereafter. Meanwhile the patient was seen at another center and received a shoulder debridement and resection arthroplasty. Following this surgery, the patient was seen at yet another clinic, and a tumor prosthesis was inserted to the right shoulder. At first week postoperatively, the patient suffered wound detachment, followed by infection of the prosthesis; thus, the prosthesis was removed 10 months postoperatively. Three years after initial presentation and one year after the shoulder prosthesis was first inserted, the patient was seen again in our clinic. On physical examination, active forward flexion at the right shoulder was 20°, abduction was 40°, and there was no active internal or external rotation. Deep tendon reflexes at the right upper extremity were hypoactive, and there was loss of temperature sense at both shoulders. The patient continues follow-up.

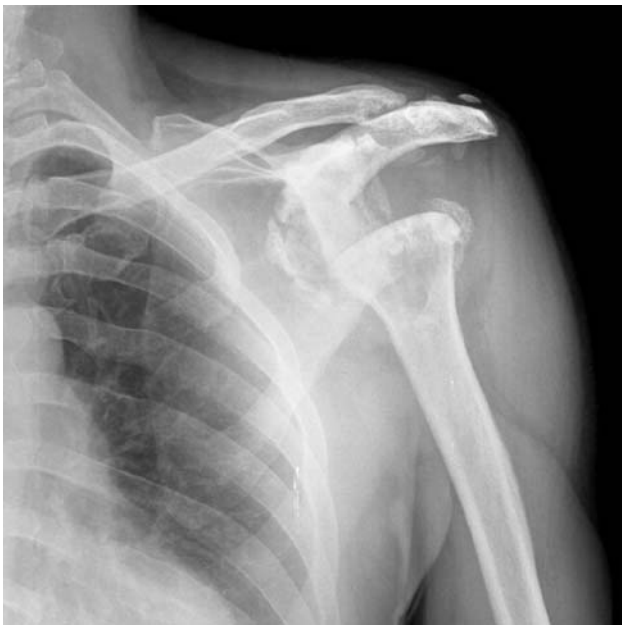

Fig. 4. Radiograph of case 1 at the third year follow-up.

Case 4

A 35-year-old female patient presented to our clinic with the complaint of swelling and pain at the left shoulder for one month. Physical examination showed that active forward flexion at the left shoulder was 30°, abduction was 40°, and there was no active internal or external rotation. Shoulder motion was painful, and there was swelling and hypoesthesia over the shoulder. Muscle strength around the left shoulder joint was found to be 2-3/5. An MRI had been ordered by the first clinic in which she was seen; destruction was detected at the humeral head. Subsequently, the patient had been referred to our clinic with an initial diagnosis of a malignant mass. Plain radiographs and MRI showed resorption at the left humeral head and glenoid (Fig. 6 and 7). A Tru-cut biopsy was carried out for suspected malignancy, and pathologic work-up showed atrophic bone lamellae. The pathologic finding was found to be inconclusive, so an open biopsy was performed, together with mass resection. Open biopsy findings were villous hyperplasia at synovium, neovascularization, bone and cartilage islets, and fibrinoid necrosis. After the assessment of the clinical findings and diagnostic work-up results, a diagnosis of neuropathic arthropathy of the left shoulder was considered, and a cervical MRI was obtained, which showed syrinx cavity between C2-T9 levels (Fig. 8). The patient was referred to the neurosurgery department, where she underwent laminectomy and poste-

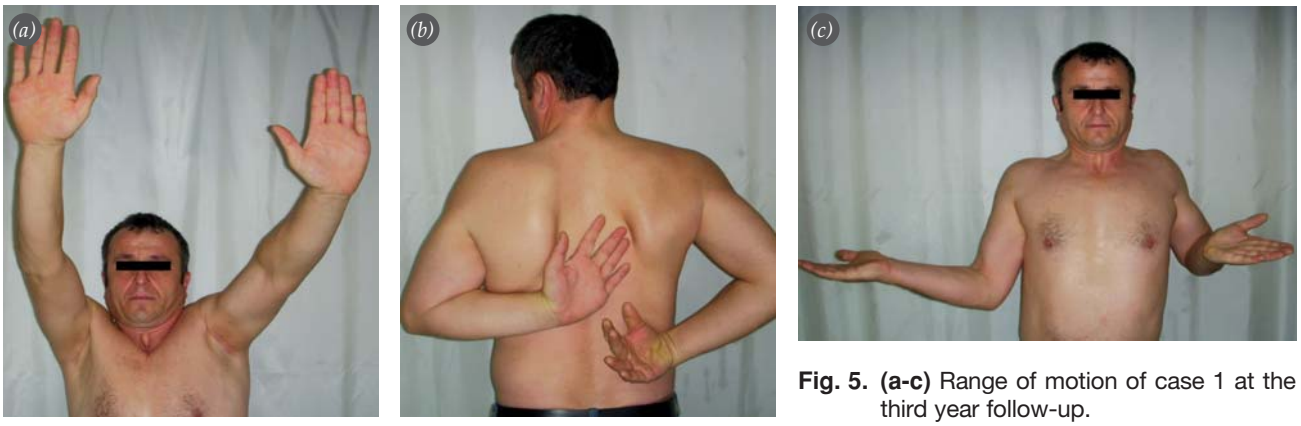

Fig. 5. (a-c) Range of motion of case 1 at the third year follow-up.

rior fossa decompression surgery. Following the operation, zoledronic acid treatment was commenced in order to prevent resorbtion at the shoulder joint. Following one year of zoledronic acid treatment, however, resorbtion persisted. Therefore, treatment was ended and conservative follow-up continued. At the end of the first year of follow-up, there was hypoesthesia at C4 dermatome with a cape-like distribution and loss of muscle strength around the shoulder joint, so the patient was referred once more to the neurosurgery department, where she was reoperated. At 18-month follow-up, it was found that resorbtion had stopped and there was sclerosis at the proximal humerus (Fig. 9a). The pain gradually lessened in intensity. At 3-year follow-up, there was a loss of thermal sense and hypoesthesia over the left deltoid region. Muscle strength around the shoulder was 4/5. Active shoulder abduction was 60°, forward flexion was 80°, internal rotation was at the lower lumbar region, and there was no active external rotation. The patient complained of pain at the left arm and shoulder during rest and with palpation, and swelling after motion. Radiographs showed a continued slow osteolytic process (Fig. 9b). Three-phase bone scan showed an increase in osteoblastic activity. The patient was reevaluated by the neurosurgery clinic with a new cervical MRI, but no operation was indicated. At the end of 3 years, the patient continues follow-up.

Case 5

A 54-year-old female patient presented to our clinic, complaining of a 3-week history of pain and decreased range of motion in the left shoulder. Past medical history revealed that she had been seen by a doctor 12 years ago with complaints of numbness and swelling of left hand and leg, and had been diagnosed with syringomyelia between C2-L1 levels. Laminectomy and decompression had been performed twice by the neurosurgery department, followed by insertion of a syringoepidural tube and Baclofen treatment; her complaints had improved after 2 days of Baclofen treatment. Physical examination showed hypoalgesia-hypoesthesia at the left shoulder with a cape-like distribution; muscle strength around the shoulder was 3-4/5; and range of motion was limited. Erosion and pathologic fracture was detected at left proximal humerus with plain radiographs. Considering patient history and clinical and radiological findings, the patient was diagnosed

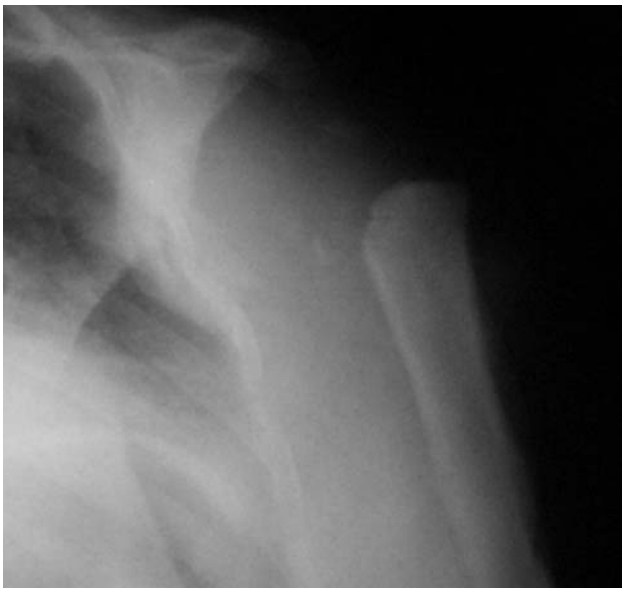

Fig. 6. Radiograph of case 4 at initial presentation.

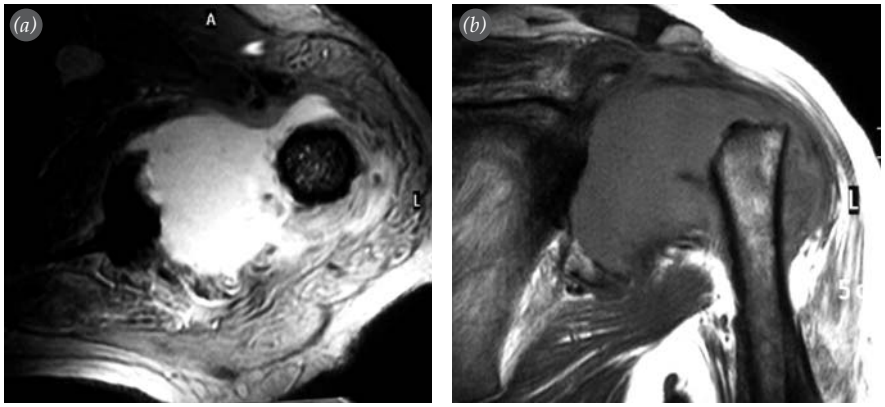

Fig. 7. (a, b) Shoulder MRI sections of case 4 at initial presentation.

with neuropathic shoulder arthropathy. Shoulder MRI showed erosion at the head and neck of left humerus, marked intraarticular synovial hypertrophy, and fluid collection. The patient was followed in a conservative manner. At 5-month follow-up, erosion was greater, and the humeral head and neck had eroded completely. Bone scan at 8 months showed marked osteoblastic activity at the left shoulder. At 13-month follow-up, active forward flexion at left shoulder was 30°, internal rotation was at lower lumbar region, and there was no active external rotation. Following antiinflammatory treatment, complaints of shoulder pain and swelling improved significantly; the patient is still followed with antiinflammatory treatment.

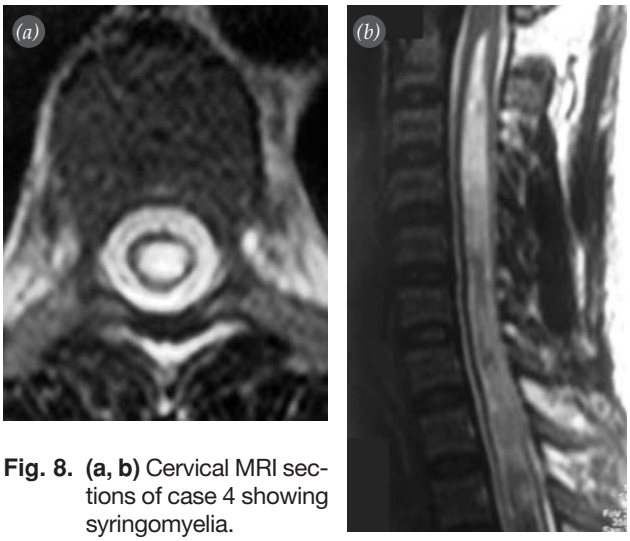

Fig. 8. **(a, b)** Cervical MRI sections of case 4 showing syringomyelia.

Discussion

Neuropathic arthropathy of the shoulder is rare when compared with other sites of neuropathic arthropathy. In a study by Brower and Alman in 1981, 23 cases of shoulder arthropathy were detected in a series of 91 neuropathic arthropathy cases. A later study found only 10 patients with neuropathic arthropathy of the shoulder in a series of 163 neuropathic arthropathy cases.[4,5]

Tabes dorsalis, diabetes mellitus, intraarticular steroid injection, peripheral neuropathy, leprosy, multiple sclerosis, myelodysplasia, meningomyelocele, congenital insensitivity to pain, amyloidosis, chronic alcoholism, and syringomyelia are amongst the factors which cause neuropathic arthropathy.[3] Considering neuropathic arthropathy of the shoulder, the etiologic factor is cervical syringomyelia in 75% of the cases;[2] frequent intraarticular steroid injections,[6] chronic alcoholism,[1] congenital insensitivity to pain[7] and diabetes mellitus are also reported to cause neuropathic arthropathy of the shoulder.

Syringomyelia is a chronic and slowly progressive disease of the spinal cord in which there is a fluid-containing cavity (syrinx) inside the spinal cord. The disease can be congenital or can occur due to trauma, infection, degeneration, vascular problem, or tumor.[8,9] Gray and white matter around the syrinx are the first structures to be damaged, which results in the disruption of pain and temperature pathways that cross the midline at the cervical level; as a result, a condition called ‘dissociative anesthesia’ occurs, in which proprioception and motor function are preserved while pain and temperature senses are lost.[8,10] As the syrinx grows bigger, areflexia, loss of muscle strength, and atrophy can be seen because of the damage to the dorsal column and anterior horn.

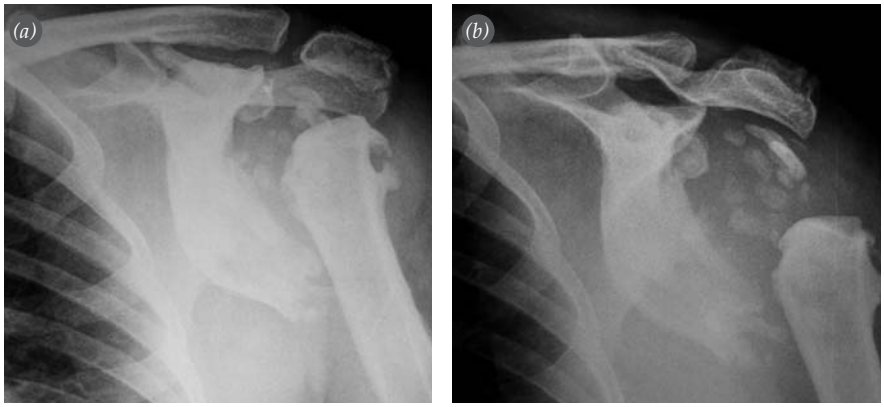

Fig. 9. Radiograph of case 4 at **(a)** 18 months and at **(b)** the third year follow-up.

Neuropathic arthropathy occurs in 20-25% of all cases of syringomyelia.[1,2] Amongst these cases, the upper extremity is affected in 80%, and the shoulder is the most common site of involvement; arthropathy can occur at an early or late period.[11] The symptoms of neuropatic shoulder arthropathy may mask the symptoms of syringomyelia; so patients may first be seen for othopedic evaluation.[1]

Consistent with the findings in our series, the most common initial symptoms of neuropathic arthropathy of the shoulder are swelling, pain, and loss in trhe range of motion; there can also be instability.[1] The differential diagnoses to be considered are septic arthritis, neoplasia (soft tissue sarcoma), synovial chondromatosis, tumoral calcinosis, and idiopathic osteolysis (hereditary multicentric osteolysis, nonhereditary multicentric osteolysis and nephropathy, Gorham disease–massive osteolysis, or Winchester syndrome).[2,12,13]

There are many theories considering the pathogenesis of neuropathic arthropathy. According to the French theory, which was developed in the 19th century and was represented by Mitchell and Charcot, damage of central nervous system trophic centers disrupts the nutrition of bone and joint and causes osteolysis. According to the German theory, represented by Volkman and Virchow, neuropathic arthropathy is caused by years of trauma, which continues subclinically for years after a loss in pain sense. Neurovascular and neurotraumatic theories were developed in the later years. According to the neurovascular theory, sensation loss disrupts normal neurovascular reflexes at joint level and leads to hyperemia and activation of osteoclasts; in the end, active bone resorption occurs. According to the neurotraumatic theory, loss of somatic muscle reflexes which protect the joint from exceeding certain limits of range of motion leads to recurrent traumas, which cause joint destruction. However, because not all reported cases have a history of trauma, and because neuropathic arthropathy is also seen in paraplegic, bedridden patients, the most widely accepted theory today is that osteolysis starts because of neurovascular processes and continues due to neurotraumatic processes. Trauma hastens disease progression.[3,5]

In patients presenting to the orthopedic clinic with complaints of swelling, pain, and loss in range of motion at the shoulder, a differential diagnosis of neuropathic arthropathy of the shoulder should be considered, and neurological examination should be carried out carefully. If a diagnosis of neuropathic shoulder arthropathy is considered to be likely after the evaluation of radiographs and MRI, a cervical MRI should be ordered to rule out syringomyelia, which is the most common underlying disease. In our series of 5 patients, syringomyelia was diagnosed after the cervical MRI study ordered by the orthopedics clinic in 3 patients, who were subsequently referred to the neurosurgery department for treatment. The progression of neuropathic joint destruction was stopped by neurosurgical decompression, and patients complaints improved at long term. Although the diagnosis of joint destruction caused by irreversible and progressive nerve damage due to syringomyelia is made by orthopedists, treatment is neurosurgery. As seen in one of our cases, orthopedic reconstructive operations applied without knowledge of the natural course of the disease and before the treatment of the underlying disease may lead to catastrophic results. Literature review also shows that arthrodesis, hemiarthroplasty, and resurfacing operations, which were tried before for the treatment of neuropathic shoulder arthropathy, have led to unsatisfactory results, although the reported follow-up periods were not sufficient.[6,14] Before a surgical treatment such as arthrodesis is considered for the shoulder, syringomyelia or other underlying diseases should be treated, and it should be documented that the osteolytic process is not progressing.

Alendronate treatment has been tried for the treatment of Charcot arthropathy caused by diabetes mellitus, and successful results have been reported in short term.[15] Although there is no similar study related with neuropathic arthropathy of the shoulder, zoledronic acid treatment was tried in one patient in our series, but was stopped after one year because there was no effect in the prevention of osteolysis.

In conclusion, neuropathic arthropathy of the shoulder is a rare disease; early diagnosis may lead to diagnosis and treatment of the underlying disease, including asymptomatic syringomyelia. In order not to miss the diagnosis, the orthopedist must be aware of characteristic clinical and radiological findings.

The only proven treatment of neuropathic shoulder arthropathy is surgical decompression of syringomyelia, if that is the cause. Reconstruction should only be considered after the osteolytic process is shown to have stopped, but bad experience in reconstructive treatment and patient expectations should be kept in mind. The pathogenesis of the disease is not fully understood; for example, it is not known why it usually occurs monolaterally. Future studies regarding pathogenesis may help in the development of new types of treatment.

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