Neuropathic Arthropathy of the Elbow: Two Case Reports

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Abstracts

Neuropathic arthropathy (NA) or Charcot joint, is a chronic and progressive degenerative arthropathy associated with an underlying neurologic disorder. NA of the elbow is a rare entity, with few cases reported in the literature.

We report the cases of 2 patients with NA of the elbow. In case 1, the probable etiology was an intramedullary lesion, most likely syringomyelia and likely diabetes mellitus. In case 2, the probable etiology was syringomyelia and likely diabetes mellitus.

The aim in NA treatment is to manage the underlying disease and reduce the rate of deformity to its lowest level. Management of NA is usually conservative. The diagnosis of NA should be considered in destructive cases without an evident pain report, and the underlying neurologic problem should be addressed.

Neuropathic arthropathy (NA), or Charcot joint, is a chronic and progressive degenerative arthropathy associated with an underlying neurologic disorder. Characteristically, patients with NA present with a swollen, erythematous joint with pain of varying degree, usually in the setting of a sensory neuropathy. The numerous causes of NA include diabetes mellitus, syringomyelia, tabes dorsalis, and peripheral nerve disorders.[1-7] The elbow is one of the sites least often affected by NA, and syringomyelia and tabes dorsalis are among the most common causes of NA of the elbow.[1,2,4]

Since Jean Martin Charcot first described NA in 1868,[8] there has been much discussion about the causes of this condition. On one hand, purely mechanical factors can produce the features of Charcot joints.[1-3] Charcot described NA in tabes dorsalis and NA has since been observed in a variety of conditions, including diabetes mellitus, syringomyelia, and peripheral nerve disorders. The joints most commonly affected are the ankles and feet in patients with diabetes mellitus, the hips and knees in patients with tabes dorsalis, and the shoulders in patients with syringomyelia.[1-7]

In this article, we report the cases of 2 patients with NA of the elbow. In case 1, the probable etiology was an intramedullary lesion, most likely syringomyelia. In case 2, the probable etiology was syringomyelia and likely diabetes mellitus. The patients provided written informed consent for print and electronic publication of their respective case reports.

Case Reports

Case 1

A 39-year-old man presented with diffuse swelling of the right upper limb and multiple painless ulcers with club-shaped fingers (Figure 1). He had a 25-year history of pain impairment in the right upper limb and a 20-year history of recurrent painless ulcers in the right hand. Painless swelling of the right elbow joint was noticed 2 months before presentation.

Specific investigations revealed weakness of grip and arm elevation on the right side and loss of pain and temperature sensations in the right hand. There was no involvement of the left upper limb, lower

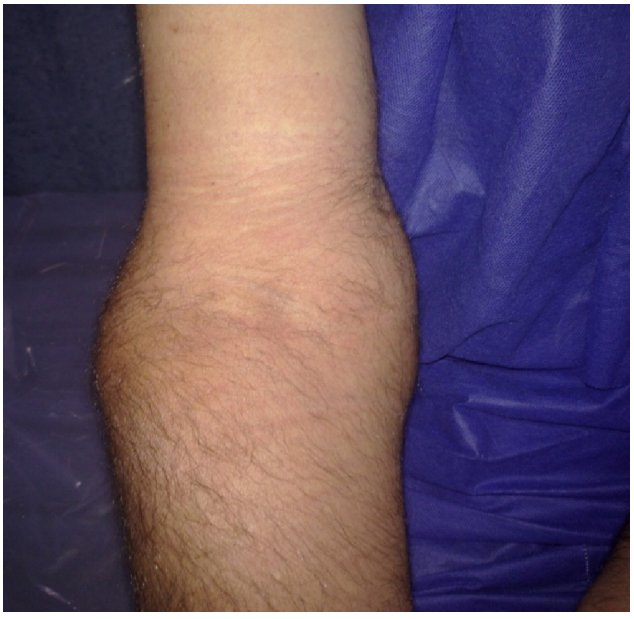

Figure 1. Case 1—39-year-old man with diffuse swelling of right elbow.

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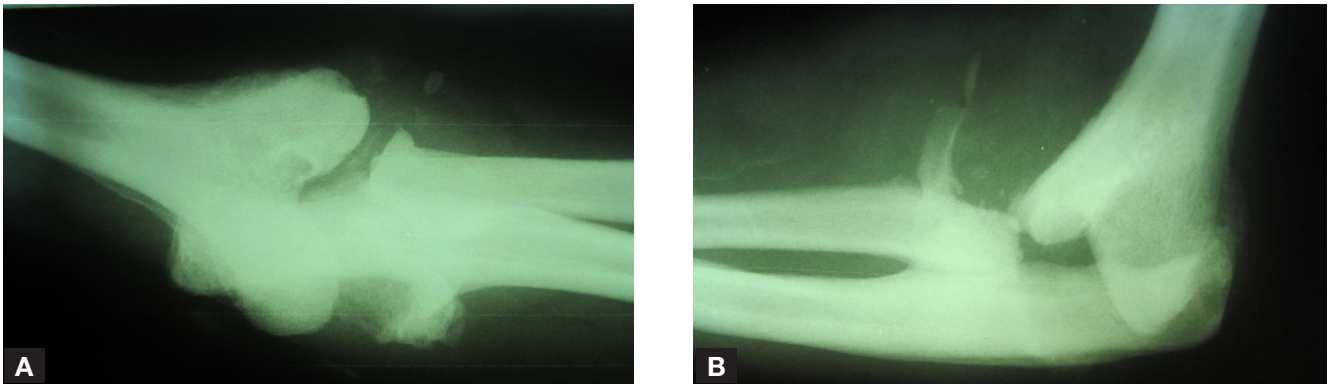


Figure 2. Case 1—plain anteroposterior (A) and lateral (B) radiographs of right elbow show "5D": debris, increased density (sclerosis), destruction, disorganization, and dislocation.

limbs, or sphincters. The patient did not have diabetes and gave no history of sexually transmitted diseases. Physical examination revealed diffuse swelling of the right upper limb with scars of old injuries. The right elbow joint was markedly swollen (47 cm in circumference), was painless on manipulation, and crepitated on passive flexion. All cranial nerves were normal. Grip was weak and there was wasting of the small muscles of the right hand. The marked swelling of the right elbow precluded proper elicitation of reflexes in the right upper limb. There was no neck pain. The sensory system demonstrated loss of pain and temperature sensations from the C2–T3 segmental level on the right, with preservation of touch. Vibration and position senses were preserved over the right upper limb. Deep tendon reflexes were hypoactive in the right limb. On the other side, reflexes were normal.

Electroneuromyography of the right upper limb showed a complete lesion in the upper and lower branches of the brachial plexus. Routine urine examination and blood counts were normal. Fasting blood glucose (FBG) was 86 mg/dL, and the blood *VDRL* (Venereal Disease

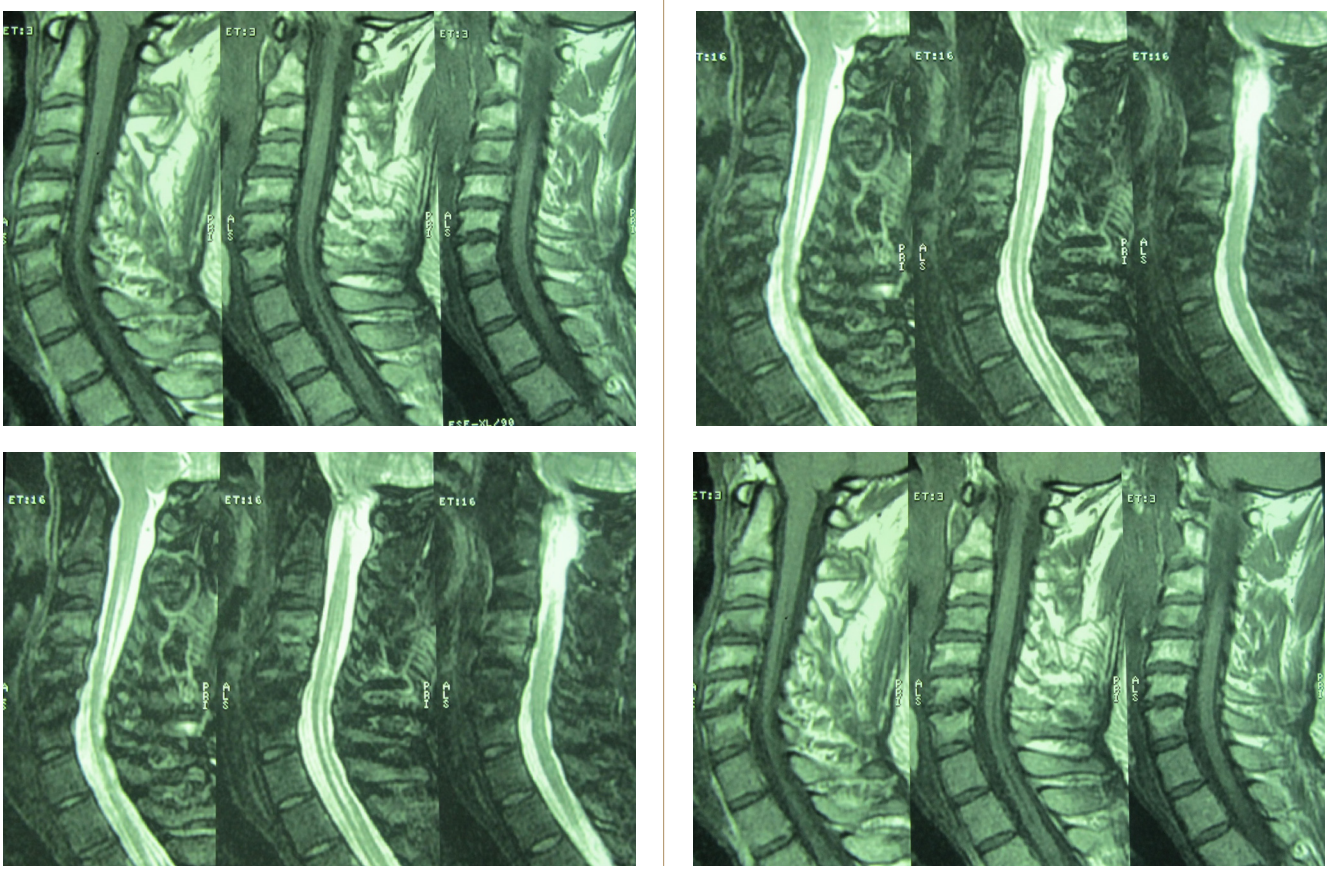


Figure 3. Case 1—cervicothoracic sagittal T_1- and T_2-weighted magnetic resonance imaging shows syringohydromyelic cavity extending from C2 to T3, with type I Arnold-Chiari malformation.

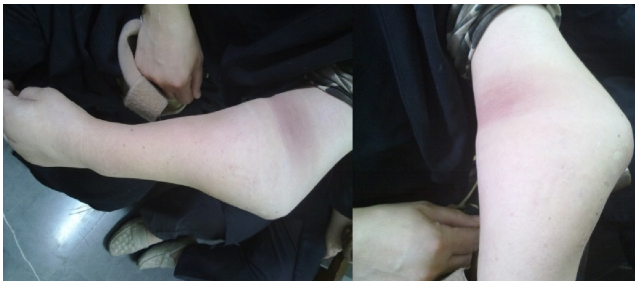
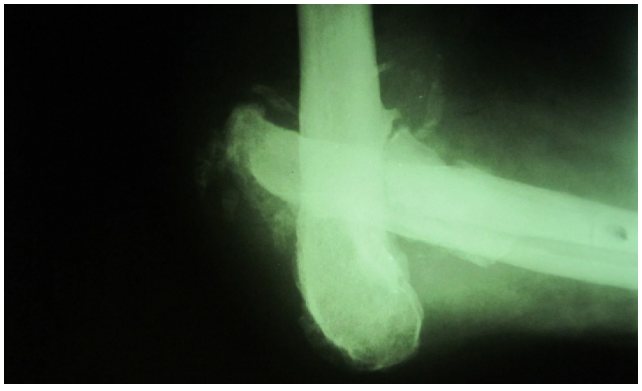


Figure 4. Case 2—48-year-old woman with limited range of motion and painless swelling of left elbow.



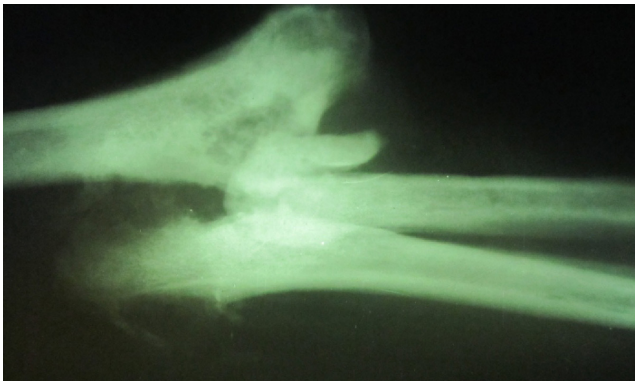
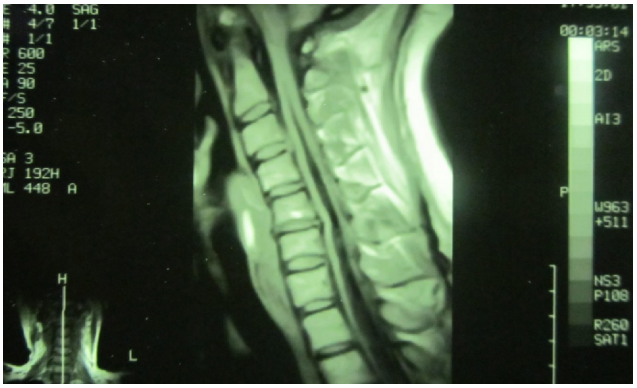
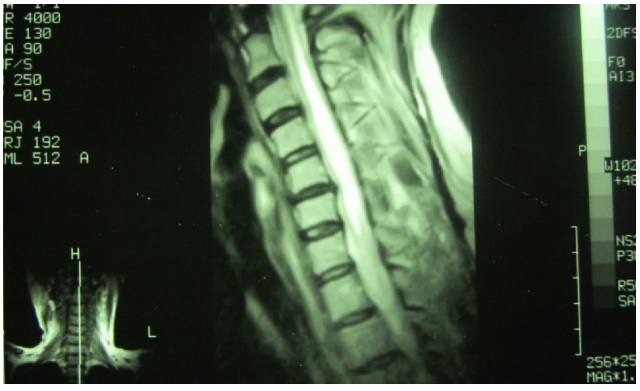


Figure 5. Case 2—plain anteroposterior radiograph of left elbow shows normal architecture of joint lost, bone fragmentation, and new heterotopic bone formation with dislocation.





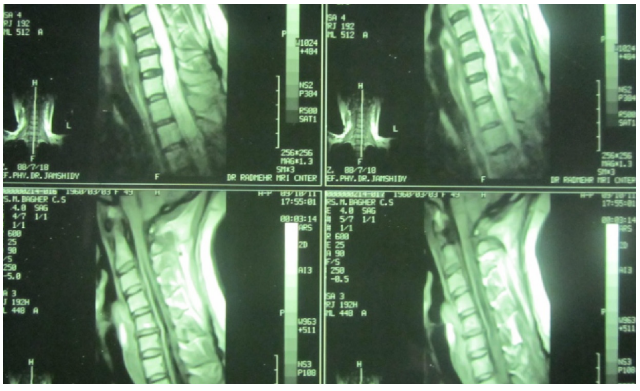


Figure 6. Case 2—cervicothoracic sagittal T_1- and T_2-weighted magnetic resonance imaging shows syringohydromyelic cavity extending from cervical region to thoracic region, with type I Arnold-Chiari malformation.

Research Laboratory) test was nonreactive. Complete blood cell (CBC) count, biochemical analysis, erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), rheumatoid factor, thyroid function tests, and tumor markers were normal. Venous Doppler ultrasonography of the right upper extremity was normal. Chest-and-cervical-spine radiograph was normal. Radiograph of the right elbow joint showed marked destruction of articular surfaces with dislocation of the bone ends, and new bone formation within the joint cavity and soft tissue around the elbow (Figure 2). Cervicothoracic magnetic resonance imaging (MRI) showed a syringohydromyelic cavity extending from C2 to T3 and a type I Arnold-Chiari malformation (Figure 3).

On the basis of these findings, we diagnosed our patient with NA in the joint of the right elbow. Probable etiology was an intramedullary lesion, most likely syringomyelia.

Case 2

A 48-year-old right-handed woman was admitted to our clinic with limited range of motion and painless swelling of the left elbow joint (Figure 4). This swelling was first presented 2 years earlier; over the past 6 months, the joint swelled rapidly.

There was weakness of grip on the left side, with loss of pain and temperature sensations in the left hand. The other limbs were not involved, and there was no history

of diabetes mellitus or sexually transmitted diseases. Physical examination revealed diffuse swelling of the left upper limb, but no trophic ulcers. Significantly reduced pain and temperature sensations were detected in the left upper extremities, but position and vibration senses were preserved.

CBC count, ESR, CRP, rheumatoid factor, thyroid function tests, and tumor markers were normal. Left upper extremity venous Doppler ultrasonography was normal. FBG was abnormal on first check (167 mg/dL) and on recheck (174 mg/dL). The patient had not consumed any antidiabetic agents. Significant destruction, sclerosis, debris, and dislocation of the articular surfaces were detected on anteroposterior radiographs of the left elbow (Figure 5). Cervicothoracic MRI showed a syringohydromyelic cavity involving the entire cervical and upper half thoracic cord levels (Figure 6).

In the light of the clinical, laboratory, and imaging findings, the case was diagnosed as NA of the elbow. Probable etiology was syringomyelia and likely diabetes mellitus.

Discussion

NA of the elbow is a rare entity, with few cases reported in the literature. Eichenholtz[9] noted only 3 neuropathic elbows in a series of 94 joints. NA of the elbow has most often been associated with syringomyelia and tabes dorsalis.[2,10,11] NA is usually seen in weight-bearing joints, such as the ankle, knee, and hip.[1,5,7] Central (upper motor neuron) and peripheral (lower motor neuron) lesions may lead to arthropathy. Central lesions (eg, tabes dorsalis, leprosy, syringomyelia, multiple sclerosis, peroneal muscular atrophy) and peripheral lesions (diabetes mellitus, alcoholism, amyloidosis, infection, familial sensory neuropathies, pernicious anemia, intra-articular or systemic corticosteroid use) may cause NA through sensory impairment.[1-3,12,13]

The pathogenesis of NA remains a controversial issue. According to one theory, joint changes result from damage to central nervous system "trophic centers," which control bone and joint nutrition. Another theory suggests that joint changes result from subclinical trauma that accumulates over years and goes unnoticed because of the insensitivity of the joint.[1-3,6,7] However, the main theories regarding the pathogenesis of NA are the neurotraumatic and neurovascular theories. The neurotraumatic theory, first described by Johnson in 1967,[14] involves repetitive trauma sustained by an insensate joint. The neurovascular theory, proposed by Allman and colleagues[15] in 1988, describes active bone resorption by osteoclasts secondary to sympathetic dysfunction and a neurally mediated persistent hyperemia.[1,3-6] Advanced NA is radiologically and pathologically characterized by the 5Ds: **D**ebris, increased **D**ensity (sclerosis), **D**estruction, **D**isorganization, and **D**islocation. Radiologic findings can be classified as hypertrophic (productive) or atrophic (destructive). Alpert and colleagues[6] noted that the ankle, knee, and elbow most often exhibit hypertrophic changes, whereas the foot, hip, and shoulder more commonly reveal atrophic changes. Other authors have suggested that the hypertrophic and atrophic forms of NA are only different stages in the natural progression of the disease.[2-5,10,11]

Syringomyelia, a fluid-filled cavity or syrinx within the spinal cord, commonly occurs in the lower cervical and upper thoracic segments; the distention may propagate proximally. Causes of syringomyelia may be congenital, traumatic, infectious, degenerative, vascular, or tumor related. MRI is the most effective modality for visualizing a syrinx. Syringomyelia develops in 75% to 85% of patients with a type I Arnold-Chiari malformation.[1-4,11] (Note: the male patient in our case 1 had type I Arnold-Chiari malformation and developed syringomyelia.)

The aim in NA treatment is to manage the underlying disease and reduce the rate of deformity to its lowest level. Management of NA is usually conservative. Numerous authors have noted that protecting the joint that is susceptible to trauma and immobilization is indispensable in the management of NA. Some authors do not immobilize the involved joint but instead encourage their patients to use it. The diagnosis of NA should be considered in destructive cases without an evident pain report, and the underlying neurologic problem should be addressed.[1,2]

Authors' Disclosure Statement

The authors report no actual or potential conflict of interest in relation to this article.

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