

A unusual presentation of syringomyelia as neuropathic arthropathy of elbow joint and review of literature

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ABSTRACT

Neuropathic arthropathy (NA) is one of the less understood and challenging condition to treat in clinical practice. Jean Martin Charcot described neuropathic joint in 1868. He has shown occurrence of NA in cases of syringomyelia and tabes dorsalis. Since then NA has been described in various other conditions. Knee and ankle joints are most commonly affected. Occurrence of neuropathic joint in upper extremity is very rare. Elbow is less commonly affected joint. We describe two cases of NA of elbow joint with unusual presentation. One patient presented with 3 month history of elbow injury with gross destruction of elbow joint. Another patient presented with acute swelling of elbow joint mimicking septic arthritis with ulnar nerve involvement. We reviewed literature to discuss etiopathogenesis, clinical presentation, and treatment available for NA of elbow joint.

Keywords: Charcot joint, neuropathic arthropathy, syringomyelia

Introduction

Neuropathic arthropathy (NA) is one of the less understood and most challenging conditions to treat in orthopedics practice. Jean Martin Charcot^[1] described neuropathic joint in 1868. He has shown occurrence of NA in cases of syringomyelia and tabes dorsalis. Since then NA has been described in various other conditions. Even after so many years the exact mechanism behind the NA is still elusive.^[3,4]

NA occurs in various neurological and systemic disorders. Various central and peripheral lesions have been identified as causes of neuropathic joints.^[1-3] NA is seen in neurologic disorders like syringomyelia, tabes dorsalis, peripheral nerve injuries, meningomyelocoele, metabolic disorders like diabetic mellitus

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and systemic sclerosis. Various joints are involved in NA. Hip, knee, ankle, and foot joints are most commonly involved. Involvement of elbow joint is very rare. Syringomyelia and tabes dorsalis are among the most common causes of NA of the elbow. NA of elbow usually develops in patients with long standing disease.

We are reporting two cases of unusual presentation of syringomyelia as NA of elbow joint. A 24-year-old male presented as posttraumatic arthritis of elbow of short duration without any history of preexisting weakness. A 44-year-old female presented as acutely swollen elbow joint with involvement of ulnar nerve mimicking septic arthritis. Primary cause in both patients was cervicodorsal syringomyelia with Grade I Arnold Chiari malformation.

Case Reports

Case 1

A 30-year-old male patient presented in outpatient department with complaint of pain and stiffness in left elbow joint of 3 month duration, following fall from motorcycle. Treating physician then had advised him above elbow splint for 2 weeks. Splint was removed after 2 weeks. Since then he was complaining of stiffness of left elbow joint. Pain was insignificant. Two months back he started noticing swelling of his left elbow joint, which gradually

increased. There was no history of fever, loss of appetite, loss of weight, urethral discharge [Figure 1].

On general examination he had no fever, no pallor, no icterus, pulse regular 74/min, and blood pressure was 124/84 mmHg. The elbow was swollen (circumference 32 cm); overlying skin was stretched but no dilated veins or sinuses. Mild tenderness was present over joint line. The swelling was boggy, soft to firm in consistency. Three point relationship was disturbed. Motion was possible from 90-110 degree. No varus or valgus laxity or malalignment was present. Pain and temperature sensation over the involved extremity were abnormal. Touch and position sensations were normal. No motor deficit was seen in forearm and hand muscles. He had one old X-ray taken immediately after trauma which was not showing any bony injury. The X-ray taken on the day of admission showed destruction of radial head, loss of joint space, and soft tissue swelling. Radial head was dislocated. Irregular destruction and sclerosis was seen over distal end humerus.

The amount of destruction of joint was not correlating to posttraumatic arthritis. So, we made provisional diagnosis of neuropathic joint with probable differential diagnosis of syringomyelia, tabes dorsalis, and diabetes mellitus. Venereal disease research laboratory (VDRL) test for syphilis was negative. Sugar levels were normal. Magnetic resonance imaging (MRI) of cervicodorsal spine revealed a syrinx extending from cervical 3 to dorsal 8 vertebral levels with Grade I Arnold Chiari malformation [Figure 2].

Patient underwent foramen magnum foraminotomy and excision of arch of cervical first vertebra to decompress the cord. Procedure was uneventful. Four months after the surgery, patient demonstrated no neurological deterioration and function of elbow was maintained.

Important points to be noted in this case are that patient had presented with posttraumatic destruction of elbow joint within



Figure 1: Case 1- Neuropathic arthropathy of elbow

a short duration of 3 month without having any preexisting symptoms of neurological involvement.

Case 2

A 45-year-old female patient came with subacute onset pain and swelling in right elbow. Swelling has progressively increased over 3 months. She gave no history of any injury to elbow joint. She also complained of warmth over right forearm and elbow and inability to extend her two medial fingers since last 1 month. She took treatment from various physicians without relief. There was no history suggestive of diabetes mellitus or syphilis.

General examination revealed no positive finding. Right elbow was grossly swollen with healed scar mark present over anterior aspect of elbow. That was a burn injury due application of hot packs for elbow pain. Ulnar claw hand with wasting of hypothenar eminence was present. Local temperature was raised with tenderness over elbow joint. Elbow joint was in dislocated position with abnormal mobility [Figures 3 and 4].

Initial clinical diagnosis of a septic arthritis was done and blood investigations and X-ray examination was done. Total leukocyte count was 10,600 with 64% polymorphs and 24% lymphocytes. C-reactive protein was positive with 60 erythrocyte sedimentation rate (ESR). Fluid aspirated from the joint was cloudy and was suggestive of chronic inflammation. Dislocation of elbow joint with cupping of distal humerus, debris, and sclerosis of bone ends was noted on plain radiograph. Increased soft tissue making with multiple opacities was seen surrounding the elbow joint. Based on these findings, we made provisional diagnosis of neuropathic joint secondary diabetes mellitus and syringomyelia [Figure 5].

MRI of cervicodorsal spine showed presence of syrinx from C4-T3 and Grade II Arnold Chiari malformation. Neurosurgeon has planned for foramen magnum foraminotomy with excision of C1 arch [Figure 6].

Interestingly, this patient presented with nontraumatic elbow joint swelling with signs suggestive of septic arthritis with

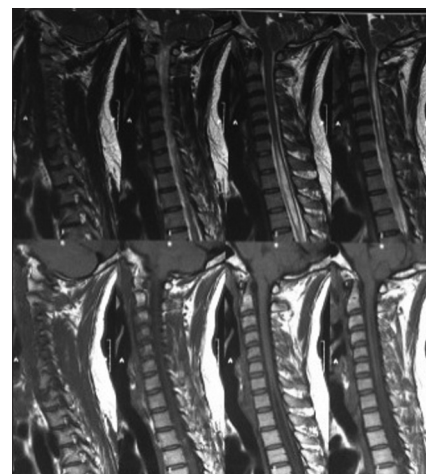


Figure 2: Magnetic resonance imaging showing cervical syrinx



Figure 3: Case 2- Neuropathic elbow with ulnar palsy



Figure 4: Ulnar claw hand



Figure 5: Radiograph showing joint destruction

involvement of ulnar nerve. Only on detailed examination we could diagnose the neuropathic joint.

Discussion

Many researchers have come up theories to explain NA. Two most accepted theories are neurotraumatic theory and neurovascular theory. Neurotraumatic theory suggested that NA results from accumulated microtrauma in joint lacking joint position and pain sensation. Allman and colleagues^[5] in 1988 suggested neurovascular theory for NA. They claimed that neural involvement produced a state of persistent hyperemia around the joint with resorption of bone through osteoclast activation. Most people agree that both theories have part in joint destruction seen in NA. The accumulated microtrauma in patients with insensate joint, leads to progressive destruction and fragmentation of articular cartilage of joint and subchondral bone. This debris accumulates inside the joint capsule with synovial effusion. Joint capsule expands and get stretched creating huge swelling.^[6] Advanced NA is radiologically and pathologically characterized by the 5Ds: Debris, increased Density (sclerosis), Destruction, Disorganization, and Dislocation. Radiologic findings can be classified as hypertrophic (productive) or atrophic (destructive).

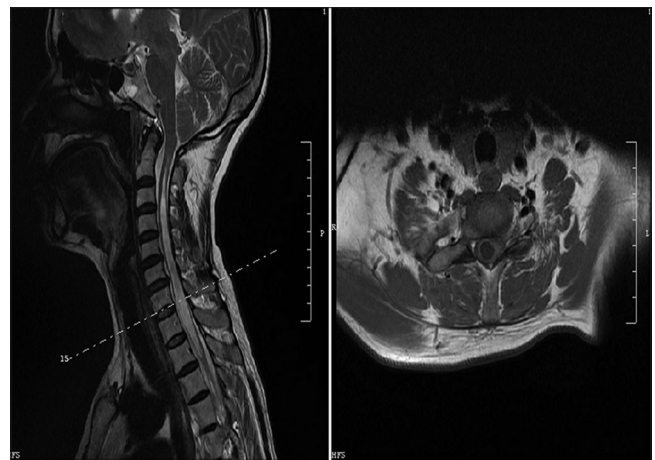


Figure 6: MRI showing cervical syrinx

Hypertrophic NA presents as disorganized joint with huge swelling. The atrophic NA is characterized by progressive destruction without much synovial collection.^[1,3]

Syringomyelia is disorder of spinal cord characterized by formation of a fluid-filled cavity in spinal cord leading progressive neurologic involvement. The sensory loss is dermatomal in distribution. Pain and temperature are more profoundly affected as compared to touch sensation and joint position sensation. Pyramidal signs are found in lower extremity (Finlayson^[2] 1988, Mancall^[4] 1984). Although the lesion is often congenital, symptoms usually appear in the 3rd or 4th decade However, 80% of joint disorders associated with syringomyelia usually present themselves in the upper extremity^[7]

Oldfield *et al.*,^[8] studied mechanism of development of syrinx in patient with Grade I Arnold Chiari malformation with anatomical and dynamic (phase-contrast and phase-contrast cine) MRI, and intraoperative ultrasonography to examine the anatomy and dynamics of movement of the cerebellar tonsils, the wall of the spinal cord surrounding the syrinx, and the movement of cerebrospinal fluid (CSF) and syrinx fluid at rest, during the

respiratory and cardiac cycles, and during Valsalva maneuver in seven affected patients.

Strike *et al.*,^[9] in 2011 reported a case of cervical syrinx presenting as septic arthritis of elbow. Detailed neurological examination revealed signs of cord compression seen in all four limbs with sensory motor deficit. The patient underwent drainage and debridement of joint. They found that downward movement of tonsils with each systole resulting into blockage of CSF flow at foramen magnum, constriction of spinal cord and syrinx. They achieved decompression of the foramen magnum via suboccipital craniectomy, laminectomy of C-1 and C-2, and dural grafting; leaving the arachnoid intact. Immediately after surgery, the pulsatile downward thrust of the tonsils and constriction of the spinal cord and syrinx disappeared. They reported resolution of syringomyelia within 1-6 months after surgery in all patients.

Ekim and Armağan^[10] reported syringomyelia presenting as NA of the joints in the upper limb involving shoulder, elbow, and metacarpophalangeal joint in the same upper limb. Atypical feature of this case was presence of shoulder subluxation and brachial plexopathy which was diagnosed by electrophysiologic studies.

Zimmermann *et al.*,^[11] described occurrence of neuropathic joint in upper extremity with myelomeningocele in a 5-year-old male child, youngest case of neuropathic joint. Treatment in this age group is very challenging.

Deirmengian *et al.*,^[12] have reported outcome of treatment in five patients with elbow neuropathic joint. They have recommended minimal surgical intervention with more emphasis on preservation of joint motion and stability through use of braces and physical therapy. They perform surgery in three patients for associated radial and ulnar nerve compression. One patient in their series required surgery for ulna nonunion. They have reported satisfactory outcome at average follow-up of 19 months.

NA can occur in various conditions and can have atypical presentation. Good clinical examination with judicious use of investigation can lead us to diagnosis of the underlying cause. Minimal surgical intervention and proper physical therapy would help to preserve joint function for long time.

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