Imaging Findings in Chiari I Malformation with Syringomyelia in a Case of Charcot Shoulder

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ABSTRACT

Neuropathic arthropathy of the shoulder is reported in only 5% of cases. Here, we report a rare case of neuropathic arthropathy of the shoulder, secondary to Chiari malformation Type I with associated syringomyelia, that remained undetected for four years. A 38-year-old female presented to our Department with a swelling over the right shoulder that had persisted for four years. X-ray of the joint showed destruction of the head of the right humerus, with typical blunt amputated appearance of the bone and increased joint space. Magnetic resonance imaging showed destruction and lateral dislocation of the head of the humerus. Large amount of fluid collection was seen in and around the right shoulder joint. Neuropathic osteoarthropathy can be defined as bone and joint changes that occurs secondary to loss of sensation. In our case, neuropathic shoulder joint was secondary to syringomyelia associated with Chiari I malformation.

Key words: Charcot, Chiari malformation, shoulder, syringomyelia

INTRODUCTION

Neuropathic joints, often called Charcot joints, are a chronic form of degenerative arthropathy caused by a loss of sensation in the joint such that the joint is severely damaged and disrupted. Typical imaging features include joint destruction, disorganization, and effusion with osseous debris. Resorption of the ends of tubular bones and neuropathic fractures are also associated findings. Charcot joints are more commonly seen in the lower limb. Previously, Tabes dorsalis was a common cause, affecting the hip and knee joints; however, this is much rarer these days. The more common cause in the present times is diabetic neuropathy, which involves ankle and foot joints. In the upper limb the classical cause is syringomyelia. Neuropathic arthropathy of the shoulder is reported in only 5% of cases. Here we report a rare case of neuropathic arthropathy of shoulder, secondary to Arnold-Chiari Type I malformation with associated syringomyelia, that was undetected for four years.
CASE REPORT

A 38-year-old female with a swelling over the right shoulder was referred to the Department of Orthopaedics. The swelling had been present for four years and was attributed to be the result of a fall. On examination, shoulder movements were restricted. Swelling was firm to hard, non-tender, associated with mild pain, with normal temperature, and diminished sensation. The patient had no history of diabetes mellitus or any other long standing disease. The patient had been treated with massage and manipulation of the shoulder by a local doctor in the village.

Radiograph done four years ago after the trauma revealed dislocation of the shoulder. The patient had no other X-rays with her. In the meanwhile, she had been treated with massage and physiotherapy continuously for a period of two years in her village, as advised by a local doctor. The symptoms got aggravated and she was referred to the medical college for further management. A repeat X-ray was done that showed destruction of the head of the right humerus leading to its typical blunt amputated appearance and increased joint space [Figure 1]. The proximal right humerus and glenoid cavity were sclerosed and the proximal humerus was subluxated superiorly. Multiple bony fragments were seen in the soft tissues around the right shoulder joint with displacement of adjacent fat planes.

Magnetic resonance imaging (MRI) showed destruction and lateral dislocation of the head of the humerus [Figure 2]. Residual humerus was subluxated superiorly with sharp margins. A large quantity of fluid collection was seen in and around the right shoulder joint. Bony fragments and debris were seen within the collection. Tear and atrophy of muscles around the shoulder joint, namely the supraspinatus, infraspinatus, and subsapularis muscles were seen. Glenoid cavity was shallow and showed destructive changes.

MRI of cervico-dorsal spine showed syrinx formation with herniation of cerebellar tonsils (6mm) into foramen of magnum leading to foraminal stenosis consistent with the diagnosis of Chiari malformation Type I [Figure 3].

Treatment consisted of non-steroidal anti-inflammatory medication, passive motion exercises, and a protective sling. The patient with syringomyelia may experience sensory loss that prevents normal guarding, resulting in
repetitive trauma and eventual joint destruction. Further
the patient was referred to a neurosurgeon, who also
preferred conservative treatment as the patient was
not having any major neurological symptoms. Patient
education is equally important to minimize the stress and
inadverent trauma.

**DISCUSSION**

Neuropathic joint arthropathy, otherwise known as the
Charcot joint was initially described by Mitchell in 1831 and
later named after Jean-Martin Charcot in 1868.\(^2,3\)

Neuropathic osteoarthropathy can be defined as bone and
joint changes that occurs secondary to loss of sensation and
that accompanies a variety of disorders. The Charcot joint
is characterized by slow progression over many years (but
rapid progression occurring over months has also been
described).\(^4\)

Common causes of neuropathic arthropathy include
diabetes, use of steroids, alcoholism, trauma, infection,
amyloidosis, pernicious anaemia, syphilis, syringomyelia,
spina bifida, myelomeningocele, and leprosy. In diabetic
neuropathy, common sites of involvement are the
metatarsophalangeal, tarsometatarsal, and intertarsal
joints. In syringomyelia, neuropathic changes are relatively
more common in the shoulder joint, followed by the elbow
and wrist.\(^5\) The lower extremities can also be affected in
syringomyelia. Changes in the spine are most characteristic
in the cervical region. The joints of the lower extremity are
commonly affected in patients with Tabes dorsalis.

Although for long there has been, recognition of the
association between syringomyelia and neuropathic
arthropathy,\(^2,6\) the pathogenesis has remained controversial.
Charcot and Mitchell\(^6\) believed the underlying mechanism
to result from damage to the CNS trophic centres that
controlled bone and joint nutrition. This concept became
known as the "French theory". Volkman and Virchow
violently disputed this concept and espoused the "German
theory". Postulating that the neuropathic joint resulted
from the accumulation of years of subclinical trauma that
went unnoticed by the individual because of an insensate
joint.\(^2\) In 1981, Brower and Aliman\(^2\) studied 91 patients
with neuropathic joint arthropathy; 32% had joint pain
without any known neurologic disease. Four of their patients
were bedridden when neuropathic arthropathy developed.
They argued that the absence of weight bearing capacity
and repeated joint trauma in these immobilized patients
disputed a traumatic etiopathogenesis for neuropathic
arthropathy and postulated a neurovascular mechanism
(first espoused by Leriche in 1927).\(^2\) According to this, a
neurally mediated vascular reflex secondary to impairment
of sympathetic vascular modulation leads to increased
bone blood flow. Concomitant with this increased blood
flow is increased active reabsorption of bone by osteoclasts,
with fracture and joint damage occurring as a secondary
epiphenomenon, depending on the degree of weight
bearing and sensory impairment affecting the involved joint.

Neuropathic arthropathy comprises of both hypertrophic
and atrophic patterns which can be discerned radiologically.\(^7\)
The classically described hypertrophic joint is manifested
radiologically as joint destruction and fragmentation,
osses sclerosis, and osteophyte formation. The six
D's of neuropathic joint include distension of joint,
increased density, debris, destruction of articular surface,
dislocation, and disorganization of joint.\(^8\) Periarticular bone
fragments are as characteristic of neuropathic joints as is the propensity to rapid disintegration. Osteophytes formed in the setting of neuropathic arthropathy may differ from those of osteoarthritis on the basis of early production of ill-defined and rounded margins and later attainment of enormous size.

Atrophic form of neuroarthropathy has an appearance of osseous resorption that often gives the impression of surgical amputation. Joint disorganization and large persistent bloody joint effusion are features of both atrophic and hypertrophic types of neuroarthropathy.

Hans Chiari first described Chiari Type I malformations as elongation of the tonsils and medial divisions of the inferior lobules of the cerebellum into cone-shaped projections that accompany the medulla oblongata into the spinal canal. Its incidence has been found to be between 0.56%[6] - 0.77%[10] on MRI studies, and 0.62% in brain dissection studies.[11]

The radiological criteria for Chiari malformation in pre MRI era were based on myelography. MRI has now become the investigation of choice for evaluating such cases. Mikulis et al.[12] found that the cerebellar tonsils ascend with increasing age and suggested the following criteria for ectopia: First decade, 6 mm below the foramen magnum; second to third decades, 5 mm; fourth to eighth decades, 4 mm; and ninth decade, 3 mm below the foramen magnum. Narrowing or complete effacement of the CSF spaces of the foramen magnum and cisterna magna were documented in all symptomatic patients. In the above case with only 6mm tonsillar herniation, effacement of CSF cisterns and syringomyelia was responsible for emergence of symptoms.

Newer imaging techniques like cine phase contrast MRI have been introduced to diagnose CSF flow obstruction in patients with Chiari malformation. It was first reported in 1991 by Tominaga et al.[13] CSF flow analysis through foramen magnum helps distinguish symptomatic Chiari I from asymptomatic cerebellar ectopia and helps predict response to surgical decompression. In Chiari patients, cine phase-contrast MRI detected the abnormal pulsatile motion of the cerebellar tonsils, which produced a selective obstruction of CSF flow from the cranial cavity to the spine. The amplitude of the tonsillar pulsation and the severity of the arachnoid space reduction were associated with the symptoms.

In our case, neuropathic shoulder joint was secondary to syringomyelia associated with Arnold-Chiari malformation. This is a less common entity, reported in only 5% cases; it has been described in fewer than 60 patients worldwide.[11] Syrinxes interrupt the descussating fibers of the lateral spinthalamic tracts that mediate pain and temperature sense.[14,15]

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