JMSCR Vol||05||Issue||11||Page 30458-30460||November 2017

www.jmscr.igmpublication.org
Impact Factor 5.84
Index Copernicus Value: 71.58
ISSN (e)-2347-176x ISSN (p) 2455-0450
crossref DOI: https://dx.doi.org/10.18535/jmscr/v5i11.110

IGM Publication
Journal Of Medical Science And Clinical Research
An Official Publication Of IGM Publication

Syringomyelia with Charcot's Shoulder- A Rare Case Report

Authors
Ginu P. George[1], Udayamma K. P.[2]
[1]Junior Resident, Department of General Medicine, Government T D Medical College, Alappuzha
[2]Associate Professor, Department of General Medicine, Government T D Medical College, Alappuzha

Introduction

Syringomyelia is a developmental cavity of the cervical cord that may enlarge and produce progressive myelopathy or may remain asymptomatic. It is a potential cause of neuropathic osteoarthropathy like charcot's joint.

Case Report

A 52 year old gentleman with no significant past medical or surgical history presented with Progressive weakness of both upper and lower limbs involving proximal and distal muscles for last 6 years and loss of pain and temperature sensation over left side of body including face for 3 yrs. Patient had sustained painless burns on his left arm 3 yrs back. Weakness started in the right upper limb, then involved the left upper limb followed by left lower limb and then right lower limb over a period of 4 years. It is then remaining static over a period of 2 years. There is history of bladder involvement in the form of urgency and precipitancy of urine.

General examination revealed burn scars over left arm and forearm and Scoliosis with convexity to right side. Neck length was normal. On examination of nervous system, there was involvement of left fifth cranial nerve in the form of loss of pain and temperature sensation over left side of face and absence of corneal and conjunctival reflex. There was Loss of contour of right shoulder with wasting of the nar prominence of right hand. Hypotonia was noted in both upper limbs with spasticity of both lower limbs. There was restriction of range of movements of both shoulders. Abduction was limited to 30 degrees bilaterally. Patient had only grade 4 power of both upper limbs and grade 4+ power of both lower limbs. Superficial abdominal and cremasteric reflex were absent. Plantar response was bilaterally extensor. Deep tendon reflexes were absent in the upper limb and exaggerated in the lower limb. Pain and temperature sensation was impaired on the left side with preservation of touch. There was loss of vibration and joint position sense on left side and right upper limb. Other system examination did not reveal any abnormality.

Investigation revealed normal routine blood results. HbA1C and viral markers were normal. MRI Brain with whole spine revealed diffuse syringomyelia of spinal cord extending from cervico medullary junction to T11 level with moderate atrophy of cord. X Rays and MRI of shoulder joint was taken. MRI revealed dislocated gleno humeral joint with deformity and destruction of glenoid and humeral head. There was gross joint effusion with multiple intra articular loose bodies and Complete resorption of

glenoid labrum and articular cartilage. The findings were consistent with charcot's joint.

In view of clinical findings and imaging results, we arrived at the diagnosis of syringomyelia with secondary charcot's arthropathy.

Fig 1- Burn scars over left arm

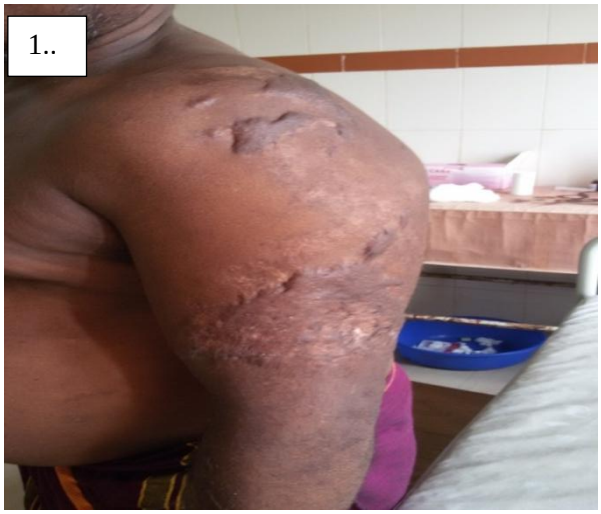
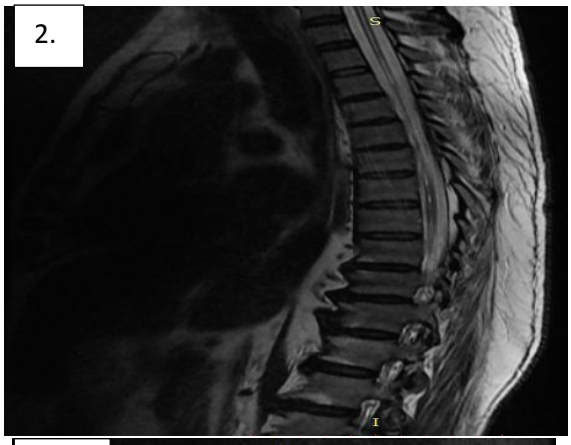


Fig 2, 3- MRI Cervical spine showing syringomyelia



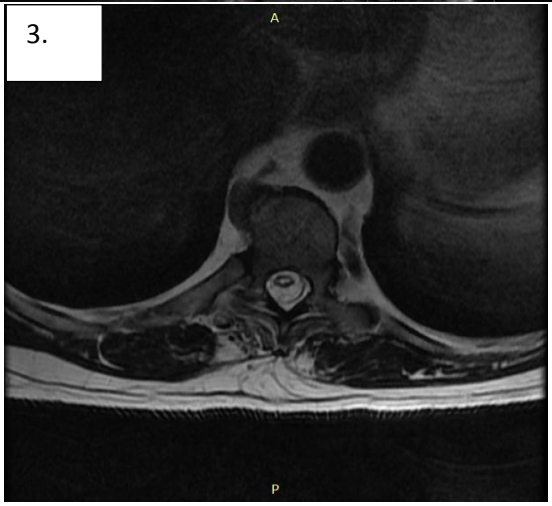


Fig 4- X Ray left shoulder showing dislocated glenohumeral joint, destruction of glenoid and humeral head, multiple intra articular loose bodies

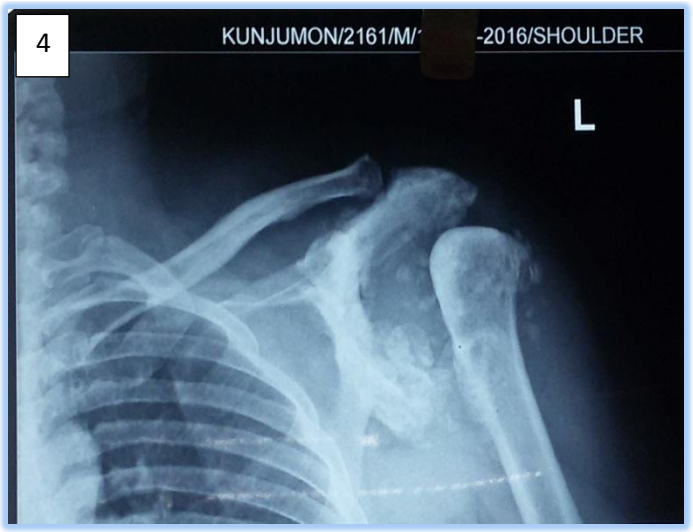
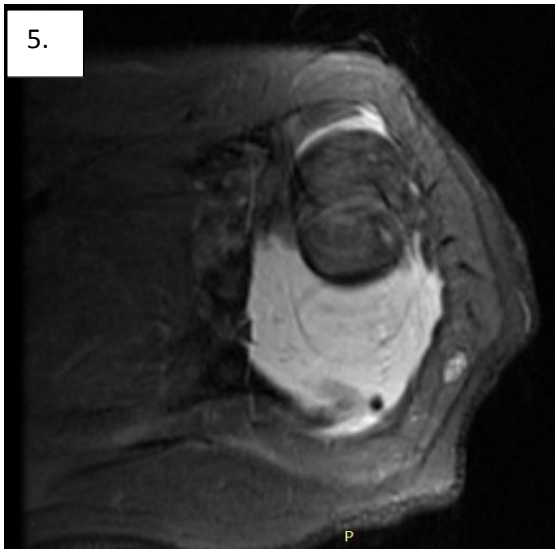
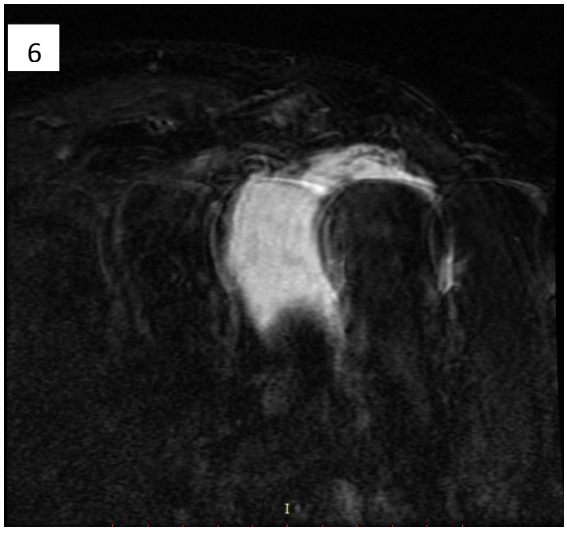


Fig 5, 6- MRI left shoulder showing features of Charcot's arthropathy





Discussion

The term "syringomyelia" was devised by ollivier in 1837 from the two Greek words "to become hollow" and "marrow". It is a developmental cavity of the cervical cord that may enlarge and produce progressive myelopathy. Symptoms begin insidiously in adolescence or early adulthood and progress irregularly. It may undergo spontaneous arrest for several years. Many young patients acquire a cervical-thoracic scoliosis. More than half of all cases are associated with Chiari type 1 malformations. The pathophysiology of syrinx expansion is controversial, but some interference with the normal flow of CSF seems likely, perhaps by the Chiari malformation. Acquired cavitations of the cord in areas of necrosis due to trauma, infections are also termed syrinxcavities. Syringomyelia that develops without trauma, spinal tumor or craniocervical or intracerebral pathology is defined as idiopathic syringomyelia Patients present with regional dissociated sensory loss (loss of pain and temperature sensation with sparing of touch and vibration) and are flexic weakness in the upper limbs. “Suspended” sensory deficit over the nape of the neck, shoulders, and upper arms (cape distribution) or in the hands is also a characteristic finding. Most cases begin asymmetrically with unilateral sensory loss in the hands, leading to injuries and burns that are not appreciated by the patient. Muscle wasting in the lower neck, shoulders, arms, and hands may be seen. Asymmetric or absent reflexes in the arms reflect expansion of the cavity in the gray matter of the cord. As cavity enlarges and compresses the long tracts, spasticity and weakness of the legs, bladder and bowel dysfunction, and Horner’s syndrome appear.

Some patients develop facial numbness and sensory loss from damage to the descending tract of the trigeminal nerve (C2 level or above). In cases with Chiari malformations, cough-induced headache and neck, arm, or facial pain may be reported.

MRI accurately identifies developmental and acquired syrinx cavities and their associated spinal cord enlargement. Images of the brain and the entire spinal cord should be obtained to delineate the full longitudinal extent of the syrinx, assess posterior fossa structures for the Chiari malform-ation, and determine whether hydrocephalus is present.

Charcot’s shoulder is a rare rapid destruction of the proximal humerus and glenoid related to neuropathic disease. There is decrease in both active and passive ROM. There are 2 theories describing the pathogenesis of neuropathic osteoarthropathy. These are the neurotraumatic and neurovascular theories. The neurotraumatic theory, first described by Johnson in 1967, involves repetitive trauma sustained by an insensate joint. The neurovascular theory, proposed by Allman and colleagues, describes active bone resorption by osteoclasts secondary to sympathetic dysfunction and a neurally mediated persistent hyperemia.

Identifying the cause of syrinx is important in deciding treatment. Patients with few symptoms and signs from the syrinx do not require surgery and are followed by serial clinical and imaging examinations. Surgical decompression is an option in selected cases. Neuro rehabilitative care helps to preserve remaining neurological functions.

Conclusion

Syringomyelia is a potential cause of neuropathic osteoarthropathy. Symptoms can mimic other neuromuscular disorders. Charcot arthropathy involving shoulder joint is an uncommon disorder, with less than 70 patients reported in the English literature. Among them, bilateral involvement was observed only in three cases. Prompt diagnosis with magnetic resonance imaging is important in establishing diagnosis.