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Shoulder Charcot- a Presage of Cervical Syrinx

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Abstract
Neuropathic arthropathy, otherwise called as charcot joint is a range of destructive disease process involving the bone and joints , resulting secondary to neurosensory deficit. It was first described by Jean-Martin Charcot in 1868 in a syphilis patient. It is not common in non weight bearing joints of upper limb. Radiological investigations play a major role in diagnosis of neuropathic joints and in eliciting the underlying cause in shoulder disease, syringomyelia being the most common etiology.

Introduction

57 year old female patient presented with complaints of gradually increasing swelling of left shoulder and restriction of movement for past 3 months. No history of fever, pain, trauma or past history of tuberculosis was there. AP Radiograph of left shoulder showed resorption of the glenoid fossa of the scapula with adjacent area of osteosclerosis, widened joint space with few small bone debri within, few subchondral foci of sclerosis and lucencies in humeral head and anterio dislocation of the gleno-humeral joint [Figure 1].

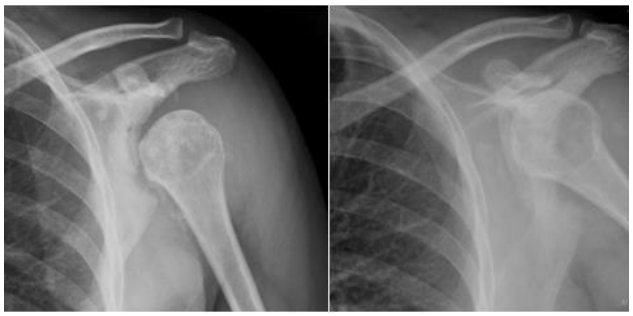


Figure 1a,b - AP radiograph of shoulder in neutral position and internal rotation showing glenoid fossa resorption, adjacent scapular sclerosis, small bone debri in distended joint cavity, few subchondral foci of sclerosis and lucencies in humeral head and anterior dislocation of gleno-humeral joint.

Magnetic resonance imaging shoulder confirmed the resorption of the glenoid, adjacent scapular osteosclerosis, large joint effusion with few bone debri, few subchondral foci of sclerosis and lucencies in humeral head and dislocated joint. Minimal marrow edema of humeral head was evident as hyperintense signal on STIR images. Screening cervical Magnetic resonance imaging spine showed a linear fluid density lesion in the center of the spinal cord extending from C2 vertabral level to upper thoracic level representing syringomyelia.

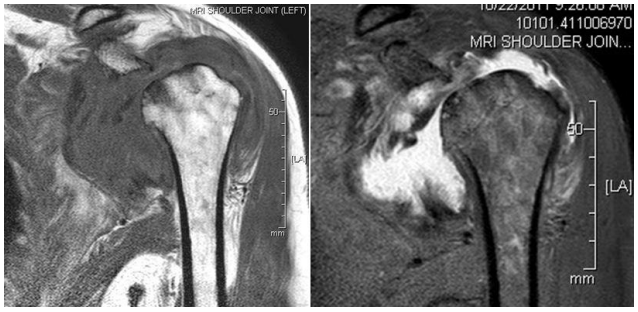
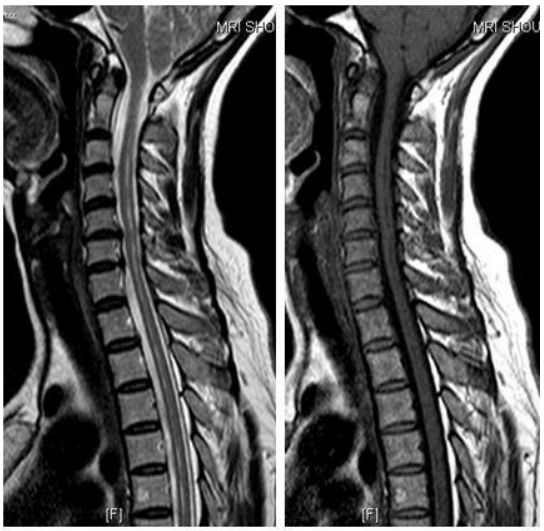


Figure 2a,b- MRI Shoulder - Coronal T1 W and STIR images showing glenoid fossa resorption, adjacent scapular sclerosis, small bone debri in large joint effusion, minimal edema of humeral head and anterior dislocation of gleno-humeral joint.



MRI Cervical spine – T1 and T2 WIS showing thin tubular cental cord T1 hypointense and T2 hyperintense lesion extending from C2 vertebra above till upper thoracic level.

Discussion

Neuropathic arthropathy was first described by Jean-Martin Charcot in 1868 in a syphilis patient. Neuropathic arthropathy, otherwise called as charcot joint is a range of destructive disease process involving the bone and joints , resulting secondary to neurosensory deficit, namely the pain and proprioception sense. The level of neurosensory deficit could be central as in syringomyelia or myelomengocele or peripheral as in diabetes mellitus and syphilis. Most common cause in Western countries is diabetes [1].

Pathogenesis

Pathogenesis behind this disease process is attributed by two theories. First is Neuro traumatic theory, according to it, the cause given is unattended multiple minor trauma due to lack of normal protective sensory reflux, resulting in progressive inflammation and osteolysis[1]. Second theory is neurovascular theory, which holds that vascular change that is associated with the neurosensory deficit is the underlying cause. There are few studies which has shown that when neural stimulus in the limb is lost, there is loss of sympathetic tone of the limb resulting in vasodilatation and hyperemia, that in turn results in resorption of the bone. The debilitated subchondral bone ends in neuropathic changes [2].

Types

Neuropathic joint is of two types: atrophic and hypertrophic. It is commonly unilateral and rarely bilateral (20%) [1]. Atrophic type is the majority one [3], commonly involving the upper limb joints, which are non weight bearing. This type is marked by articular end resorption of the affected bone, acutely progressive nature, destruction of the joint with fragment resorption and lack of osteosclerosis and osteophyte formation [4]. In Hypertrophic type sensory nerves are only involved. It is characterized by insidious progression of the disease, destruction of the joint with bone debri, widening of the joint space in the beginning and joint space narrowing subseque-

ntly, existence of osteosclerosis, formation of osteophytes [3] and lack of osteoporosis, except when there is superadded infection[4].

Etiology

The causes of neuropathic joint are diabetes (most common),leprosy, syringomyelia, spinal cord injury, spina bifida, scleroderma, multiple sclerosis, poliomyelitis, rheumatoid arthritis, tertiary syphilis and steroid use. The location of the affected joint gives clue to the underlying etiology as following: shoulder - syringomyelia, hip: syphilis, knee: syphilis, congenital insensitiveity to pain and ankle and joints of foot: diabetes

Imaging features

Radiograph and CT – the common mnemonic which reminds us the general imaging features of charcoat joint is six Ds , namely – increased density bone (subchondral sclerosis) , destruction of articular ends, debris of bone, dislocation / subluxation of joint, distension of joint with fluid and disorganization.

MRI – all general features of charcot joint could be appreciated well on MRI. Sclerosis is seen as hypointense signal on both T1, T2 WIs. Fluid distending the joint is seen as hypointense collection on T1WIs and as hyperintense collection on T2WIs. Major advantage of MRI is, early identification of complications like superadded osteomyelitis and exact assessment of the disease extent. Early evidence of the osteomyelitis/ soft tissue infection could be seen as hyperintense signal on STIR images and other FAT SAT images. On gadolinium based contrast study infected tissue will show enhancement.

Differential Diagnosis

Common differential diagnosis of neuropathic joints on imaging includes: Osteomyelitis in advanced stage, osteomyelitis can co-exist and complicate the neuropathic joint. Inflammatory arthritis, in spine - tuberculous spondylitis and osteoarthritis in hypertrophic type.

In our patient the differentials considered were, primary and metastatic malignant tumor, tuberculous and other microbial infection. Clinically due to the presence of gradual onset joint distension and on radiographically due to the presence of bone debri, in the joint space, suspicion of neoplastic etiology was there, However identification of joint centered disease with involvement of bones on either side of the joint ,large joint effusion with bone debri, helped us to differentiate the neuropahic joint from primary or metastaic neoplasms which commonly involves the bone on one side of the joint and by the absence of joint effusion. In addition absence of chondroid matrix in the involved bones ruled out the possibility of chondrosarcoma.

The features which favored the diagnosis of neuropathic joint instead of tuberculous arthritis was the disproportionate minimal amount of humeral head edema in the presence of significant bone destruction and the co- existent cervical and upper thoracic syringomyelia.

Final Diagnosis

Made was Neuropathic joint of the shoulder – predominantly atrophic type with cervical syringomyelia as the underlying etiology.

Though leprosy and diabetes could be the underlying cause for shoulder charcot, cervical syringomyelia is a commonest causative factor. Syringomyelia is a fluid collection in the spinal cord, the causes being congenital, traumatic, infective, degenerative disease, vascular origin and neoplasm related[5]. In syringomyelia, the adjacent gray and white matter are disrupted, pain and thermal sensory fibers being involved primarily contributing to the neuropathic joint disease of shoulder. Most sensitive radiological modality to identify syrinx is MRI.

Whenever charcot shoulder is suspected, co-existence of underlying causes like leprosy, diabetes and spinal pathologies to be looked for with the help of clinical history, examination, blood sugar estimation, and cervical MRI screening respectively in accordance to the

patient's presentation. Neoplastic etiology can be ruled out radiologically. Fluid aspiration and cytological examination either add to the image based differentiation of cahrcot joint from infective cause or support the presence of superadded osteomyelitis complicating charcot if suggested by imaging.

In our patient the syrinx is of congenital in origin for which the patient did not had any treatment. It progressed with time and resulted in charcot shoulder. Surgical management of the syrinx and surgical shoulder reconstruction surgery were suggested for the patient.

Conclusion

Radiologically, precise diagnosis of neuropathic joint of shoulder and its differentiation from infection or tumor is possible. Syringomyelia being the most common possible underlying etiology of shoulder Charcot, it is necessary to investigate for it, to provide comprehensive management of the patient condition.

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