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ARP Case Report Nº 20: Charcot Neuroarthropathy of upper Limb

Caso Clínico ARP Nº20: Neuroartropatia de Charcot do Membro Superior

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Case presentation

65-year-old patient with a personal history of rheumatoid arthritis diagnosed more than 30 years ago, undergoing treatment.
Four years ago, she was referred for an orthopaedic consultation for paresthesia and inability to extend the fingers of the left hand. There was no history of trauma.
An electromyography (EMG) was performed which revealed cubital neuropathy on the left, with a probable location in the elbow, and carpal tunnel syndrome.
During the evaluation by several Specialties (Neurology, Neurosurgery, Rheumatology, Physiatry), an MRI of the skull, cervical and dorsal spine was performed, which showed Arnold-Chiari I malformation and cervico-dorsal syringomyelia(Fig. 5). The development of pain and swelling of the shoulder, associated with peripheral neuropathies of the left upper limb, prompted the request for an imaging evaluation of the shoulder, with the information of "an expansive lesion of the shoulder with nervous involvement and paresis of the left upper limb".
After radiographs, CT and MRI of the shoulder (Figures 1, 2 and 3 respectively) and radiographs of the elbow(Fig. 4) were performed, the presence of extensive gleno-humeral joint disorganization, with effusion, fragmentation and chronic rupture of the rotator cuff, associated with the presence of similar changes in the elbow on the same side and the existence of syringomyelia and Chiari malformation (Fig. 5), led to the diagnosis of chronic Charcot's neuroarthropathy.

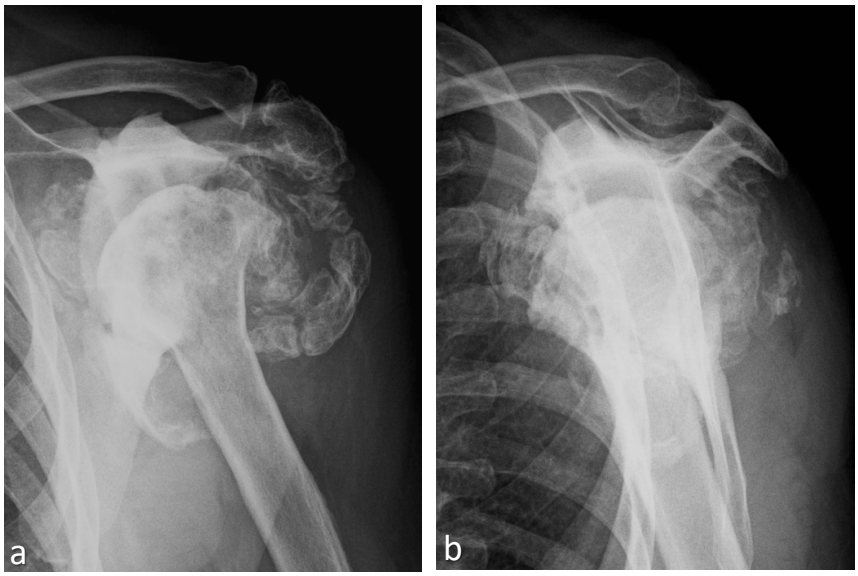

Figure 1 – Radiographic study of the left shoulder in two views (a, b). Radiographs show glenohumeral joint deformity with sclerosis and multiple peri-articular bone fragments.

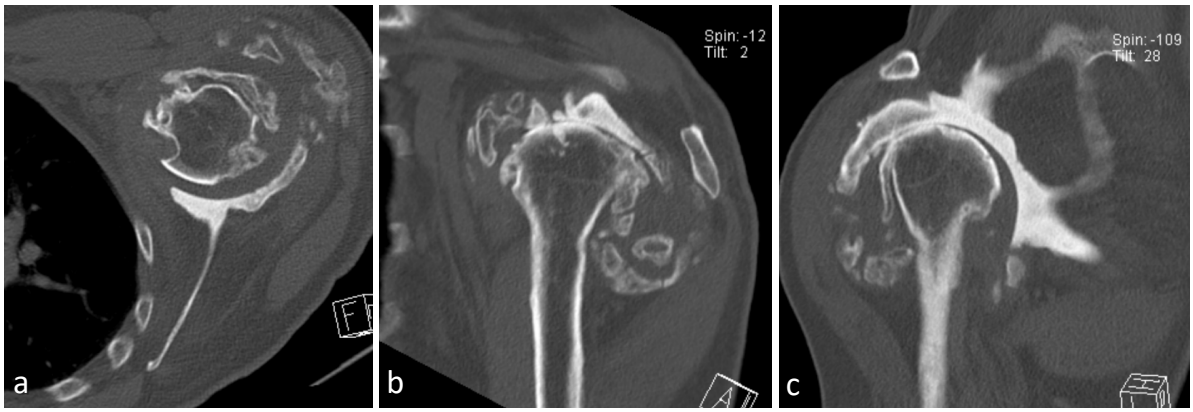

Figure 2 – Axial (a), sagittal (b) and coronal (c) reconstructions of left shoulder CT. Joint disorganization is evident, with anterior glenohumeral subluxation and marked bone deformities, with emphasis on the exuberant enlargement of the glenoid, associated with productive bony changes, with sclerosis, osteophytosis and extensive fragmentation.

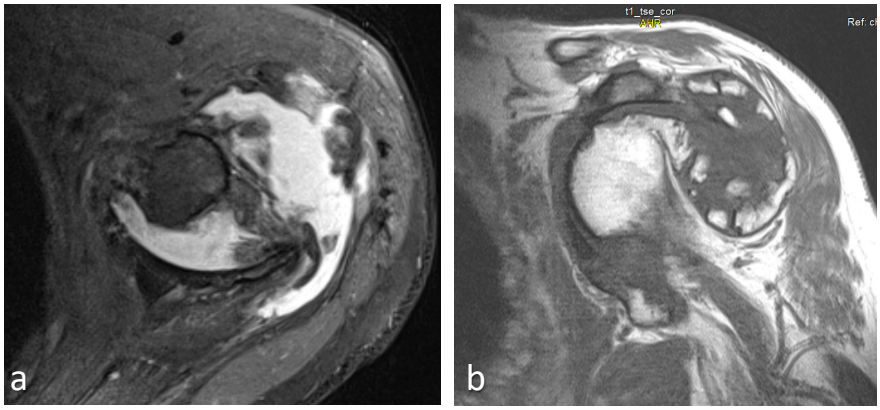

Figure 3 – Left shoulder MRI, with axial PD sequences with fat saturation (a) and coronal T1 (b), which demonstrate an extensive joint effusion and the intra-articular location of multiple bone fragments.

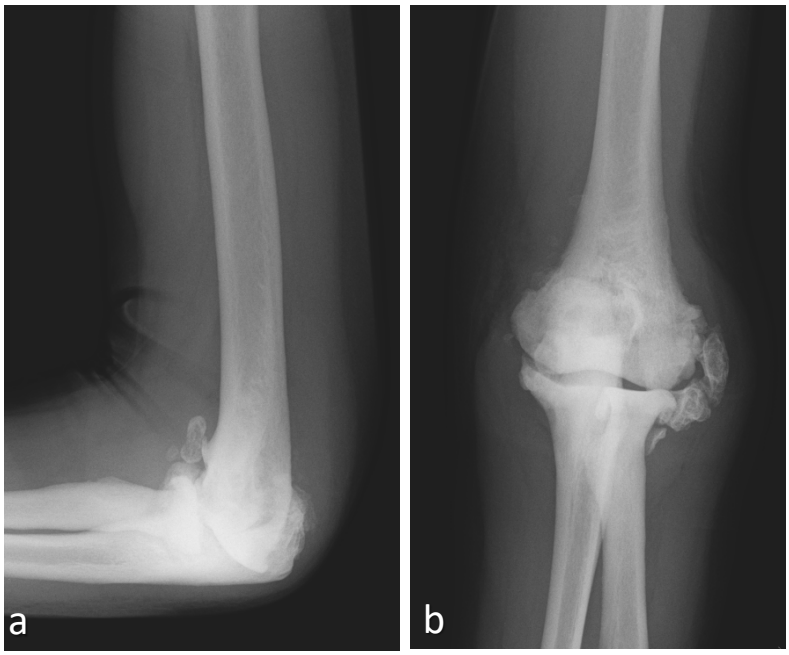

Figure 4 – Radiographic study of the left elbow in two views. Radiographs show marked joint deformity with sclerosis and fragmentation, associated with thickening and densification of soft tissues. These aspects represent hypertrophic neuroarthropathy of the elbow.

Discussion

Neuropathic arthropathy or Charcot's neuroarthropathy is a generally progressive and chronic degenerative condition, which causes destruction of one or more joints. It is associated with a neuro-sensorial deficit, the etiology of which includes several entities, the most frequent being diabetes mellitus, syringomyelia, tabes dorsalis and other peripheral and central neuropathies. Early identification of the cause of this neurosensory deficit is essential to prevent the progression of the arthropathy.

Regarding the physiopathology of this entity, there are two commonly accepted theories: the neurotraumatic theory and the neurovascular theory.

The neurotraumatic theory advocates that in the genesis of joint destruction there are repeated microtraumas, resulting from the loss of peripheral sensitivity and proprioceptive sensitivity.

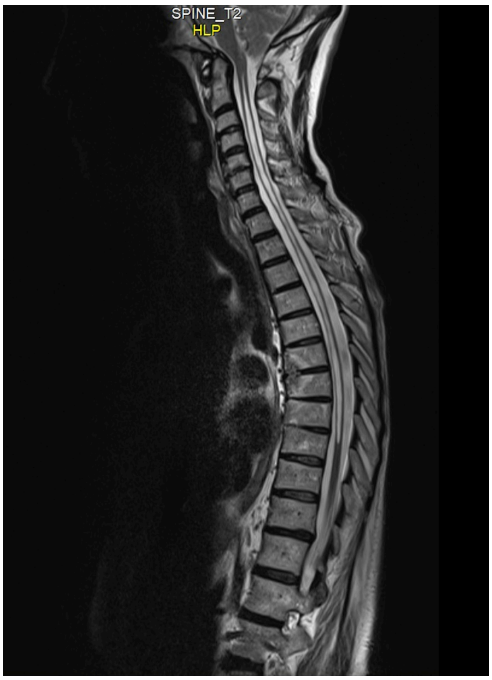

Figure 5 – MRI of the cervical and dorsal spine, with a T2-weighted sagittal image, showing a hydrosyringomyelia cavity from C1 to D9, as well as a low position of the cerebellar tonsils, below the plane of the foramen magnum, representing Chiari I malformation.

The neurovascular theory suggests that the peripheral neuropathy leads to increased bone blood perfusion, which in turn increases osteoclastic activity and results in osteopenia. The fragile bone is prone to pathological fractures, causing important destruction and joint deformation.

The anatomical location of the bone and joint findings, the patient's age and clinical history provide clues for etiological investigation.

In the upper limb, the most frequent etiology of Charcot arthropathy is syringomyelia, with 20-25% of patients with syringomyelia developing Charcot arthropathy.

In the genesis of syringomyelia, several conditions can be found, such as posterior fossa neoplasms, vertebra-medulla injuries, subarachnoid hemorrhages, arachnoiditis of the basal cisterns and malformations of the cranio-vertebral junction, namely the Arnold-Chiari malformation.

Syringomyelia can present a wide variety of neurological symptoms, depending on its exact location, although traditionally there is loss of thermal and pain sensitivity in a "cap" along the cervico-dorsal region and upper limbs. As the volume of syringomyelia increases, there may be abolition of myotactic reflexes, reduction / loss of strength and muscle atrophy, with changes typically more expressive in the upper than in the lower limbs.

In Charcot's arthropathy caused by syringomyelia there may be joint involvement in the shoulders and elbows, and rarely in the hands, the monoarticular form being the most common.

In the specific case of Charcot's neuropathy of the shoulder, the most frequent initial clinical manifestation is an increase in volume, followed by pain, stiffness and reduced joint amplitude.

The imaging manifestations of neuropathic osteoarthropathy vary over a spectrum that includes the atrophic pattern and the hypertrophic pattern (the two may coexist), the first being more frequent in upper extremity joints, which do not support the body load. According to some authors, these two patterns, which share joint disorganization, represent different stages of the natural history of the disease.

Traditionally, in the hypertrophic pattern there is joint destruction with fragmentation, bone sclerosis and formation of osteophytes, which are sometimes difficult to distinguish from advanced degenerative changes.

In the atrophic form there is massive bone resorptionwhose appearance is often similar to a surgical amputation, whileat other times it has similar characteristics to septic arthritis.

Although traditionally syringomyelia is associated with the atrophic form of shoulder neuroarthropathy, the exuberant bony productive changes in this case indicate the hypertrophic form of this entity.

The mass effect of arthropathies of the upper limb, particularly at the level of the elbow, may cause cubital neuropathy due to the compressive effect of bone / synovial hypertrophy on the nerve at the level of the epitrochleo-olecranon gutter.

Fractures are another possible manifestation which may happen spontaneously or as a result of minor injuries. They often go unnoticed and are later diagnosed by the visualization of an exuberant bone callus.

Several entities can be included in the differential diagnosis of the clinical case in question.

Rheumatoid arthritis, which the patient had for decades, does not explain the present findings, since it is essentially characterized by destructive bony changes (e.g. erosions, osteopenia), with productive changes (e.g. ankylosis, secondary degenerative joint disease) much more subtle and with a different imaging pattern than the one found.

Degenerative arthropathy, namely the one which develops in chronic rupture of the rotator cuff, as well as the so-called Milwaukee shoulder, can present with instability and massive effusion, but the bony productive changes are generally less exuberant, without the degree of disruption and fragmentation observed in the present case.

Myositis ossificans and tumoral calcinosis are associated with calcifications that are sometimes exuberant in soft tissues, but do not present with bone or articular involvement. Additionally, the calcifications are extra-articular and have a distinct pattern.

Synovial osteochondromatosis is another entity that may present with multiple ossified intra-articular loose bodies.

The degree of joint disorganization and the exuberance of degenerative changes in Charcot's arthropathy are, once again, key factors in the differential diagnosis.

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