Inflammatory pseudotumor involving sternoclavicular articulation

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Abstract

Inflammatory pseudotumor is a rare kind of tumor of unknown etiology, occurring more frequently in the lung and the orbit, but it can involve other anatomic site while emulating both clinical and radiologically a malignant condition, representing a diagnosis dilemma.

We report the case of a patient with inflammatory pseudotumor involving the left sternoclavicular articulation.

Key words: inflammatory pseudotumor, sternoclavicular articulation.

Introduction

The inflammatory pseudotumor was first observed in the lung and described by Brunn in 1939, having been named by Umiker et al. in 1954, as it is prone to emulate both clinic and radiologically a malignant condition.1

This rare type of tumor has an unknown etiology, occurring more often in the lung and orbit, but there are already two described cases of attaining several anatomic locations, as the nervous central system or the gastrointestinal tract.[2]

The inflammatory pseudotumor is histologically characterized by the presence of acute and chronic inflammatory cells with a variable fibrosis degree.[3,4]

It is described the clinical case of a patient with an inflammatory pseudotumor involving the left sternoclavicular joint.

Clinical case

Female patient 80 years of age, white race, born and residing in Penela, without a relevant personal history, with a pain in the left shoulder not related with effort or trauma, of inflammatory characteristics, progressing for two months, associated to the tumefaction of inflammatory signs on the left supraclavicular fossa and parasternal region. She has been medicated with non-steroid anti-inflammatories and antibiotics but showing no complaints improvement.

From the objective exam, it is to be highlighted the existence of a tumefaction around 3cm [1.2 inches], hard, low mobility, painful with inflammatory signs, in the left supraclavicular fossa and left parasternal region. There were no palpable peripheral adenopathies or other changes in the reminder exam.

Analytically, there was only a change on the sedimentation rate (65 mm/1st hour) and CRP (5.8 mg/dL) without any other changes in the hemogram, coagulation, biochemistry or protein gram. Hemoculture and blood serum for Brucella, Salmonella, Rickettsia, Chlamydia, Coxiella and EBV were negative. Tumor markers s Ca 125, Ca 15.3, CEA, AFP and Ca 19.9 were within the normal range of values.

The left sternoclavicular joint radiography has shown erosive alterations at internal extremity level that can correspond to an arthritic condition.

A skin and superficial structures ultrasound has detected, in the left sternoclavicular joint level, a solid expansive lesion measuring 7.2x5.0x3.3 cm [2.8x1.9x1.3 inch], heterogeneous with hyper reflected foci, conditioning a shadow cone which seemed to correspond to a bone metastasis or a bone destruction conditioned by the mass. The lesion was independent from the thyroid and it did not seem associated to adenopathies.

The left sternoclavicular joint CAT scan has shown an increased thickness of the peri-articular soft tissues and destruction of the joint bone, being these changes compatible with an intense inflammatory

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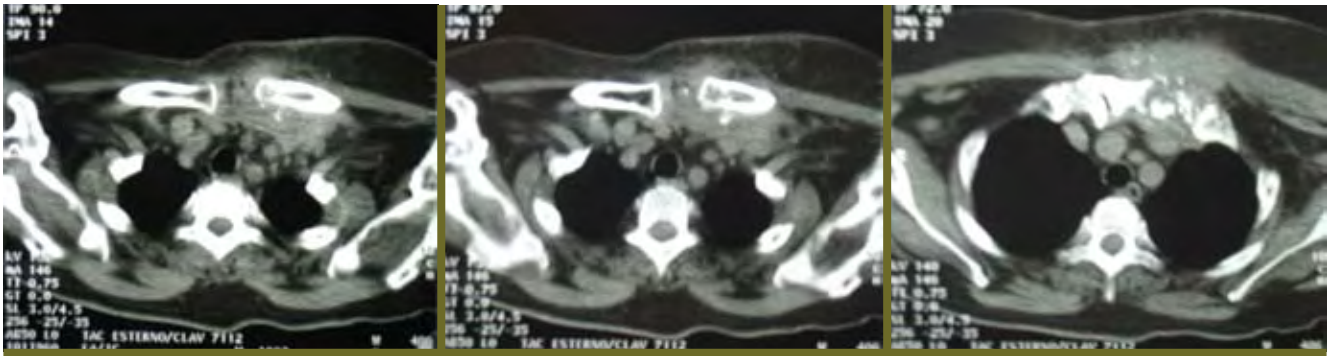
Received for publication on the 10th October 2006
Accepted for publication on the 31st October 2008



Sternoclavicular CAT Scan.

FIG. 1

or infectious condition, other etiologies cannot be excluded, namely neoplasm *(Fig. 1)*.

The left sternoclavicular joint biopsy showed an inflammatory tissue with collagen stroma, permeated by lymphocytes, plasmocytes and some giant cells of foreign body, following a neocapillary proliferation, compatible with subacute to chronic inflammatory reaction of the region.

On the sternoclavicular joints NMR it could be seen a left sternoclavicular joint image with 5.0x5.2x5.8 cm [1.9x2.0x2.3 inch], spreading posteriorly to the anterior mediastine (prevascular space) and anteriorly to the subcutaneous plans of the anterior thoracic wall. This image showed imprecise limits with heterogenous hyposignal in T_1 and heterogeneous hypersignal in T_2 and STIR, spreading to the involving soft tissues and to the clavicular medullar and sternal manubrium, meaning edema. The described aspects were suggestive of a local inflammatory process *(Fig. 2)*.

Before such clinical condition and as the patient kept the symptomatology in spite of the implemented anti-inflammatory therapy, it was decided the lesion exeresis, and the patient was transferred to the Coimbra University Hospitals (HUC) Cardiothoracic Surgery Service.

Posteriorly the histology of the surgical piece has revealed acute panniculits and synovites characterized by the formation of abscesses with polymorphonuclear neutrophils and fibrin replacing the sternoclavicular, with fibroblast proliferating, angiogenesis and hyperplasia in the nervous structures. The diagnosis of inflammatory pseudotumor involving the sternoclavicular joint was therefore confirmed.

After the surgery the patient progressed favorably with symptoms disappearing. At present, there is no recurrence history and the patient has been asymptomatic for the last year.

Discussion

Inflammatory pseudotumors are etiologically enigmatic, nosologically confusing, and often biologically unpredictable. They are histologically characterized by the presence of acute and chronic inflammatory cells with a variable fibrotic response.[3,4]

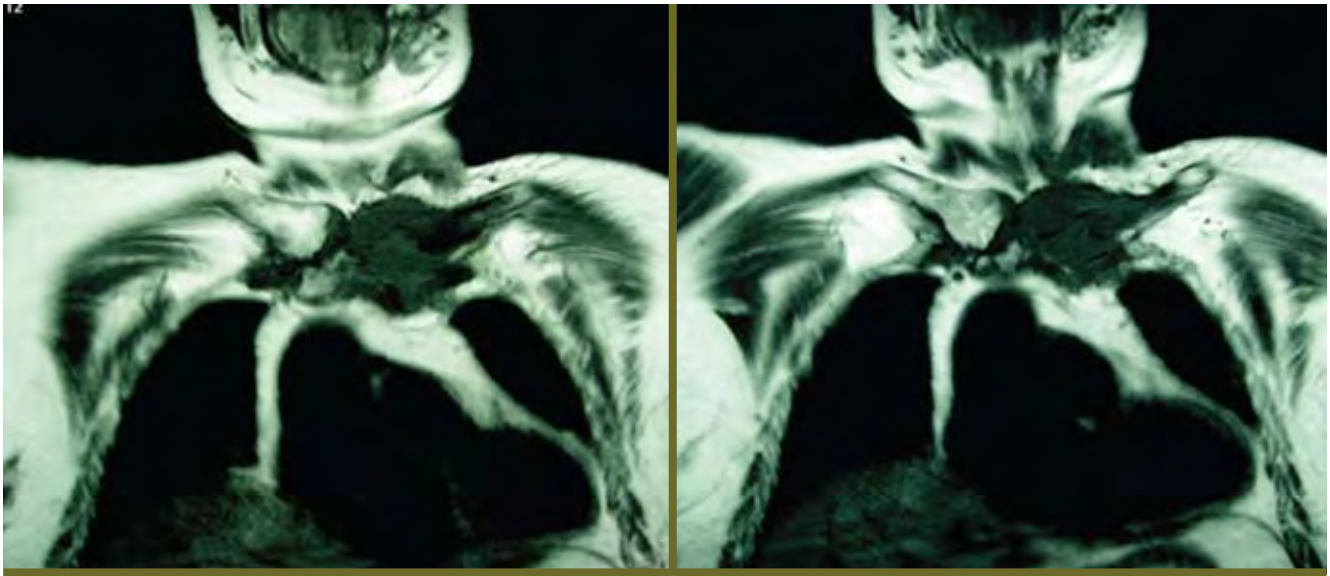
Inflammatory pseudotumors causes are unknown. Some authors believe this is a low grade fibrosarcoma tumor with inflammatory cells (lymphomatosis). Inflammatory tumors tend to be locally aggressive, sometimes multifocal, and to progress occasionally to a malignant tumor supporting such view.[5-9]

In some cases, it is thought the inflammatory pseudotumor is the inflammatory byproduct of a small trauma or surgery or might be associated to another neoplasm.[10,11]

There seems to be a subtype of inflammatory pseudotumors occurring after an infection. The living organism found associated to these tumors include Mycobacteria, EBV, Actinomycetes, Nocardia and Mycoplasma.[7]

In the current case there was no history of previous trauma or surgery in the affected area and the patient had no signs of infection either, both cultures and serology were negative.

Clinically, patients with inflammatory pseudotumors are prone to several fever degrees, delayed growth, anemia by lack of iron, thrombocytosis and hypergammaglobulinemia.[11,12] In the case of our



Sternoclavicular joints NMRI.

FIG. 2

patient, we have only found pain with inflammatory characteristics and an increase on inflammatory markers ESR and CPR.

According to previous descriptions, it is very difficult whether clinically or radiologically, to decide when a lesion involving an infiltrating mass of soft tissues, with bone destruction, is a pseudotumor or a malignant neoplasm.[4,13-17]

In the clinic case presented the diagnostic hypotheses were of bone malignant neoplasm, mediastine neoplasm or metastatic lesion, which were not confirmed after biopsy and were excluded after the tumor resection with histology revealing only inflammatory aspects.

The biology potential of inflammatory pseudotumors is highly changeable, but usually it has an innocuous progression with local recurrence in about 25% of cases. Rare cases of metastasis[8] and spontaneous remissions have been described.

The tumor surgical resection, if possible, is the treatment of choice for most inflammatory pseudotumors, excepting orbit tumors.[18-21] In the described case, the patient had a clinical improvement after the surgery and one year after, remains asymptomatic.

To conclude, the inflammatory pseudotumor, in the current case, involving the sternoclavicular joint is an extreme rare lesion emulating malignant lesions and might show different clinical manifestations. However, this kind of tumor is benign, might be surgically curable, although requiring a prolonged follow-up in all patients due to the risk of recurrence. ■

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