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Pleural Desmoid Tumour: A Case Report

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Abstract **Case Report**

Desmoid tumours or fibromatoses are rare, benign fibroblast tumours of the soft tissues, characterised by invasive infiltration and a very high rate of local recurrence, but without metastatic potential. Pleural localisation is extremely rare. The diagnosis of certainty is histological. Imaging is not very specific. We describe the case of a patient with a pleural desmoid tumour.

Keywords: Desmoid Tumour.

INTRODUCTION

Desmoid tumours are fibroblastic tumours belonging to a group of aggressive benign tumours of fibrous tissue with a high potential for recurrence and locoregional invasion [1, 2]. They correspond to a monoclonal fibroblastic proliferation characterised by a variable and often unpredictable clinical course. They usually occur in the abdomen, the roots of the limbs and the chest wall. Pleural localization is rare [3, 4]. The definitive diagnosis is histological. Imaging, which is not very suggestive of the diagnosis, is used to evaluate the lesions and, above all, to detect recurrences. Treatment is ideally surgical whenever possible.

CASE REPORT

A 51-year-old patient with a history of treated pulmonary tuberculosis presented with a right thoracic parietal swelling for 6 months, evolving in a context of apyrexia and conservation of general condition. Clinical examination revealed a right thoracic bulge with no inflammatory signs. Standard radiography (topogram) showed an opacity over half the right thoracic hemichamber, concave superiorly and medially (Figure 1). Complementary CT revealed a poorly limited, spontaneously heterogeneous right pleural lesion process projecting from the middle lobe, with discretely hyperdense areas that were heterogeneously enhanced after injection of PDC, locally infiltrative, associated with a homolateral pleural effusion (Figures 2 and 3), pulmonary nodules and an osteolytic process of the middle arches from the 5th to the 8th homolateral rib (Figures 4 and 5), invading neighbouring muscles by contiguity. Bronchial fibroscopy revealed no signs of malignancy.

An anatomopathological study of a surgical biopsy taken from the thoracic lesion was in favour of a fibromyoblastic tumour, confirmed by immunohistochemistry. Trocar biopsies were taken from the pleural lesion. Histological and immunohistochemical studies were also in favour of a desmoid tumour (Figure 6). Given the lack of resectability and the size of the areas to be irradiated, which contraindicated radiotherapy, and given that the patient's general condition had to be preserved, simple monitoring with symptomatic treatment was opted for.

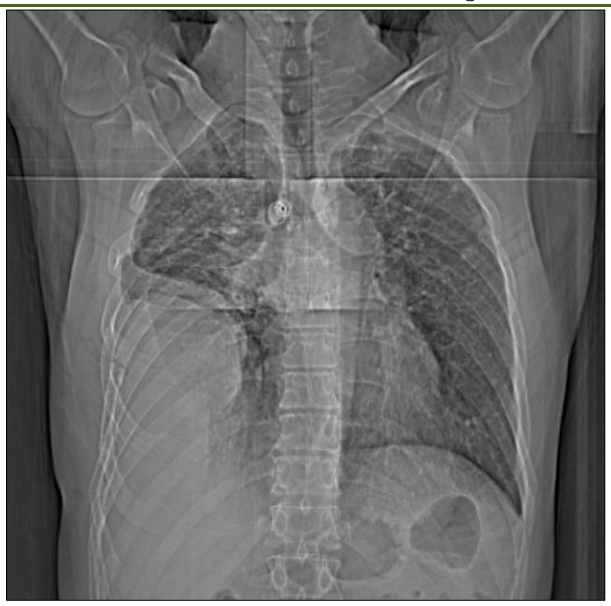

Figure 1: Standard radiography (topogram) showed an opacity over half of the right thoracic hemichamber, concave superiorly and medially.

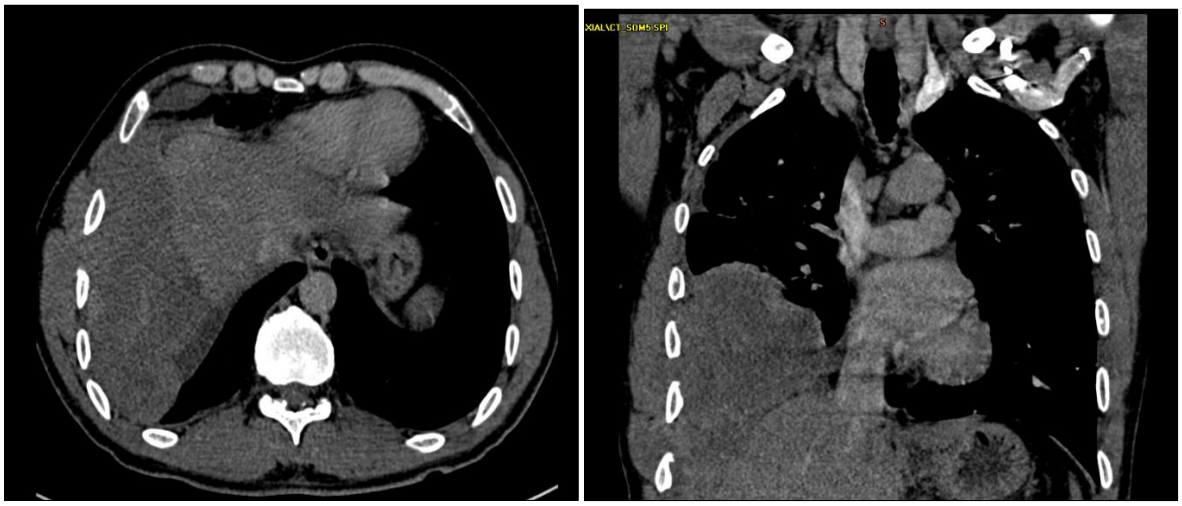

Figure 2 et 3: CT scan (axial and coronal sections) Locally infiltrating right pleural lesion associated with homolateral pleural effusion

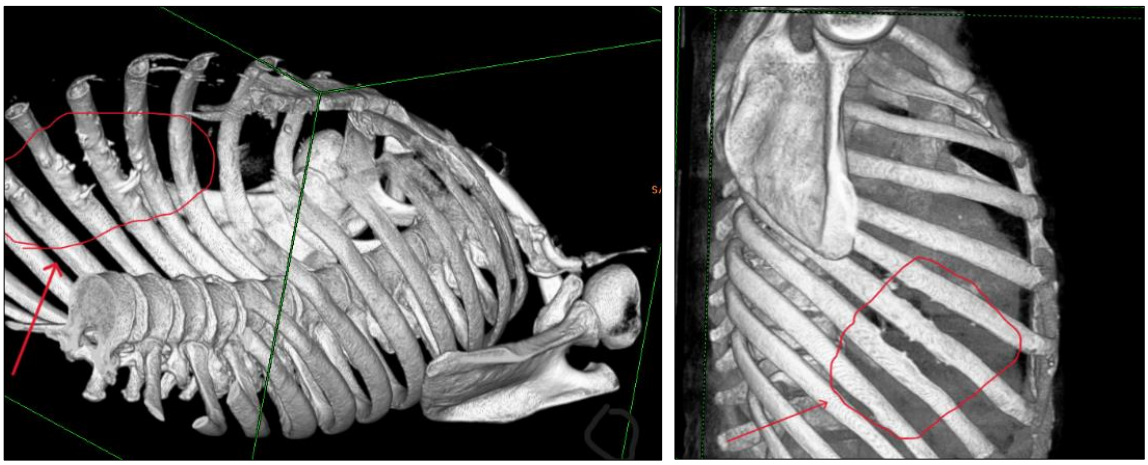

Figure 4 et 5: CT scan (3D reconstruction): osteolytic process of the middle arches from the 5th to the 8th rib on the right side

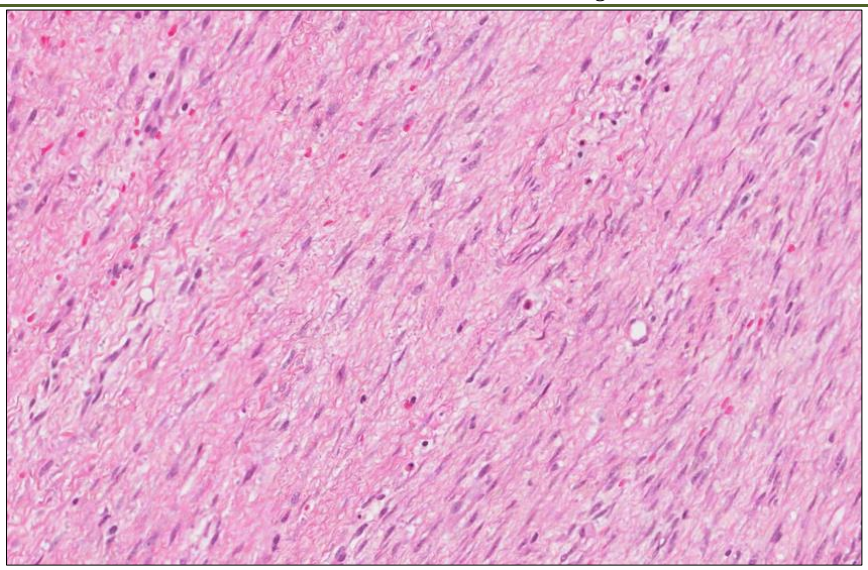

Figure 6: Microscopic appearance of the desmoid tumour, consisting of long thin cells called spindle cells

DISCUSSION

Desmoid tumours or aggressive fibromatoses are rare benign tumours that develop from the connective tissue of the musculoaponeurotic system [5]. They account for 3.5% of soft tissue tumours and 0.03% of all tumours [6]. They are most often found in the abdominal region. Extra-abdominal forms can be seen in the girdle, at the roots of the limbs, and rarely in the thorax. Their aetiology remains ambiguous. The predilection for young women between the ages of 25 and 35, and spontaneous regression after the menopause or ostrogen treatment, suggests a hormonal cause. Favouring factors have been identified, in particular a history of surgery, trauma or certain genetic factors [7]. Primary pleural localisation is rare, and most often occurs as an intra-thoracic extension of a parietal or cervical tumour [8]. The clinical picture is not very specific and depends on the location and infiltrative capacity of the tumour. There may be chest pain and/or signs of compression of neighbouring structures (vena cava syndrome, heart failure, etc). Imaging is non-specific and is not diagnostic. A CT scan reveals a more or less well-limited mass of tissue density, which may be slightly or slightly hypodense, with homogeneous enhancement, sometimes heterogeneous in large tumours with areas of necrosis and microhaemorrhages. MRI allows better tissue characterisation, better assessment of relationships with neighbouring structures (nerves, vessels, deep organs) and differentiation of postoperative changes or after medical treatment of a tumour recurrence [9]. It is often an ovoid or infiltrating mass, with generally lobulated or sometimes irregular borders, showing a homogeneous iso or hyposignal in T1-weighted sequences and a variable signal, often hypersignal, in T2 sequences, with intense and heterogeneous contrast after injection of gadolinium. The presence of hyposignal bands, associated with collagen bundles, on all sequences is very characteristic [10]. The diagnosis of certainty is anatomopathological, with evidence of fibroblastic or myofibroblastic cells, confirmed by immunohistochemical study. Treatment is essentially surgical, with wide resection where possible. In the event of non-resectability or contraindication, medical treatment involves hormone therapy and anti-inflammatories to stabilise or, more rarely, reduce the size of the tumour. There may also be courses of chemotherapy or radiotherapy.

CONCLUSION

Pleural desmoid tumours are a rare entity to be considered in the differential diagnosis of pleural masses, the diagnosis of certainty is histological, imaging is not very specific. The prognosis is unpredictable.

This case highlights the importance of close monitoring and a multidisciplinary approach to the management of desmoid tumours [10].

Conflict of Interest: The authors declare that they have no conflict of interest with this article.

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