

**CASE REPORT**

# Non Surgical Desmoid Tumor with Pulmonary and Scapular Localization: A Case Report

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## ABSTRACT

**Background :** Desmoid tumors are rare and represent 0.03% of all solid tumors. Their histology is benign but they are willingly aggressive with significant locoregional invasions and frequent recurrences.

**Case presentation :** We report the case of a 52-year-old patient who presented with a scapular and pulmonary localization of this tumor, but stable over time. In view of potentially decaying surgery and stability of the lesions, simple monitoring was decided. The lesions are currently stable, two years after diagnosis. We discuss the clinical, radiological and therapeutic characteristics of this type of tumor, in the light of data from the literature.

**Conclusion :** Desmoid tumors are rare, benign but aggressive. Surgery is the main treatment but is often decaying. Simple monitoring can be scheduled in case of less aggressive tumors.

**KEYWORDS :** Desmoid tumor, surgery, pulmonary localization, scapular localization.

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## INTRODUCTION

Desmoid tumors are rare tumors of musculoaponeurotic tissue. Their frequency is estimated at 0.03% of solid tumors [1]. They usually develop between 15 and 60 years of age and are more common in females [2]. They are localized mainly in abdomen but thoracic lesions are not uncommon. They are potentially aggressive and often relapse locally without metastasis [3]. They also can remain stable for a long time without treatment, or heal spontaneously [4]. We describe the case of a patient with a non surgical dual-site desmoid tumor.

## CASE PRESENTATION

A military 52-year-old patient, former moderate smoker at 6 pack-years, was admitted for basi-thoracic and right scapular pain. He reported the occurrence of the same thoracic pain 6 years before for which a chest x-ray had been performed showing a right paracardiac opacity. We note in his history a closed chest trauma that had happened 23 years ago on a mine glare.

The clinical examination showed a painful swelling next to the right scapula. The patient was hemodynamically and respiratory stable. Cardiopulmonary examination was normal. The rest of the clinical examination was unremarkable.

The chest X-ray showed a stable right para-cardiac opacity compared to the old photograph (Fig 1). A thoracic CT scan was performed objectifying a necrotic, multi-lobed, right para-cardiac, pulmonary tissue mass with two pulmonary nodules on the right lower lobe as well as a heterogeneous tissue mass infiltrating the right scapula (Fig 2).

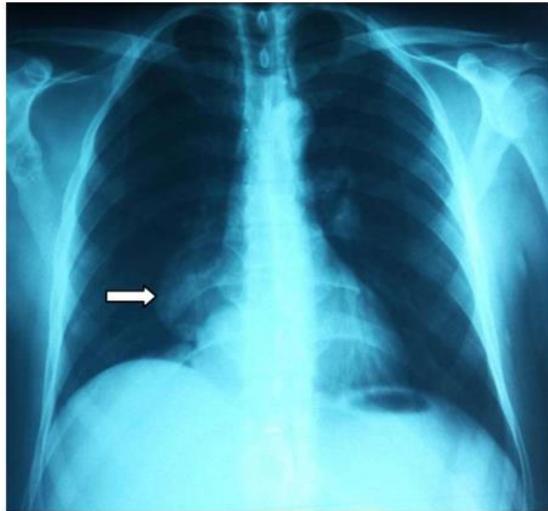
An MRI was performed to assess mass relationship with the heart chambers and showed infiltration of the right atrium (Fig 3).

The biological assessment was normal with no inflammatory syndrome or hematological abnormalities.

The spirometry showed a moderate restrictive ventilatory disorder at 2.54 liters of FEV1 (67% of theoretical).

A biopsy of the scapular and mediastinal mass was performed showing a fibroblastic tumor proliferation

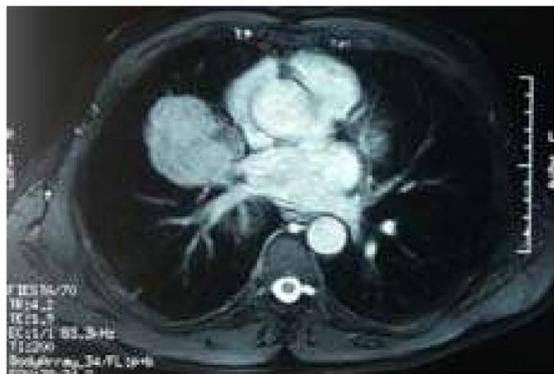
made of intertwined bundles of cells with regular nuclei and fine chromatin. The immunohistochemical study showed an expression of anti-AML and  $\beta$ -catenin antibodies, and a negativity of desmin, PS100 and CD34. Ki67 was expressed by less than 1% of tumor cells.



**Figure 1 :** Frontal chest x-ray showing right paracardiac opacity.



**Figure 2 :** Chest CT scan showing a bulky well limited mediastinal posterior mass, with two pulmonary nodules of the lower right lobe (green arrow) and a thoracic wall mass invading the right scapula (red arrow)



**Figure 3 :** Chest MRI showing invasion of the right atrium by the tumor mass.

In total, the morphological aspect and the immunohistochemical profile were in favor of a desmoid tumor.

Given the stability of the lesions and the difficulty of the surgical approach, simple monitoring was decided. The lesions are currently stable, two years after diagnosis.

## DISCUSSION

Desmoid tumors, also called desmoid fibromatosis, aggressive fibromatosis or low-grade sarcomas [5], were first described in 1832 by Mac Farlane [1].

They represent 0.03% of solid tumors and 3.6% of soft tissue tumors [1,6]. The incidence is estimated at 2 to 4

per million inhabitants [7,8]. They occur mainly in adolescents and young adults with a female predilection and an age group between 15 and 60 years [2]. Occurrence in children is rare [9].

Despite its low grade of malignancy, this tumor is aggressive with very frequent local recurrences but without metastases.

The locations are very variable [10-12]: abdominal wall, intra-abdominal (mesenteric, retroperitoneal, pelvic), extremities, belts, chest wall (scars), intra-thoracic (exceptional), cervico-facial .

No etiology has been identified for these tumors, but pathophysiological hypotheses have been advanced. The appearance of these lesions after a trauma, especially on scars as in our patient, supposes a fibrous transformation of a hematoma [13]. The frequent association with familial adenomatous polyposis (Gardner syndrome) in 5 to 10% raises the question of genetic predisposition [13, 14]. And finally, a rapid growth of the tumor during pregnancy and the use of oral contraceptives and its possible regression after menopause raises the hypothesis of a hormonal influence [13,15].

Recent cytogenetic studies have shown a lack of phosphorylation of  $\beta$ -catenin (an adhesion molecule of mesenchymal cells) which may be at the origin of the disease [6]. The search for these abnormalities within tumors would allow the detection of potentially aggressive lesions requiring surgery.

Clinically, this tumor is rarely painful except in case of nerve compression. It results in a simple hard, bumpy, painless swelling of the soft tissues without evident skin changes [16].

Radiologically, this is often a heterogeneous, sometimes necrotic, compressive lesion with possible erosion of bone structures [17]. In the case of intra-thoracic localization, MRI makes it possible to study the relationship with the adjacent structures before surgery [6]. In the case of our patient, the tumor infiltrated the right atrium, making surgery difficult.

The diagnosis is based on histological analysis of the surgical specimen. When resection is not considered, a simple biopsy of the lesion can be performed, sometimes by mediastinoscopy, thoracoscopy or even thoracotomy for intrathoracic tumors. Transparietal biopsies under thoracic CT are not contributory, due to the small size of the samples.

The histological analysis finds a clonal, sometimes multifocal proliferation of fibroblasts in inter-muscular bundles, along the fascia without cellular atypia. Collagen tissue is often abundant [18]. The differential diagnosis arises with fibrosarcoma, which has cellular atypia and less collagenous tissue. The sarcoma immunohistochemical markers are usually negative in the desmoid tumors.

Surgical excision is not indicated as first-line treatment. Surgery is decided in multidisciplinary consultation according to the evolution of the tumor [6].

The prognosis is fairly favorable: progression-free survival of 50% at 5 years and 20 to 30% of spontaneous cures, simple monitoring can therefore be considered in

chosen patients (Watchful and Waiting) with clinical and radiological re-evaluation after 8 to 12 weeks then quarterly for 1 to 2 years [4].

The treatment is mainly surgical, often decaying with wide excisions, requiring autologous or prosthetic wall reconstructions [19-21]. Surgery must take into account the size of the tumor, its evolution, its extension, its anatomical location, any loss of function and the possibilities of reconstruction.

Radiotherapy (56 Gy in photons) can be considered in inoperable patients or in addition to surgery in case of incomplete resection [22, 4]. Other treatments have been tested with varying efficacy: NSAIDs, tamoxifen, methotrexate, vinblastine / vinorelbine, anthracyclines, tyrosine kinase inhibitors (imatinib, nilotinib or sorafenib) [23, 4].

Our patient was not operated due to the stability of the lesions over 6 years, the deep infiltration of the scapular lesion and the invasion of the right atrium. There is no evidence of scalability two years after diagnosis.

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#### AUTHORS' CONTRIBUTIONS

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Recurrences are frequent and occur in half of the cases [3]. They occur mainly in young people under 30, women and in case of incomplete resection [23, 4].

The scalability of the tumor is linked to its aggressive potential. The challenge is to detect weakly aggressive tumors that can be monitored, in order to avoid decaying surgery with frequent recurrences. Further progress in cytogenetic may be very helpful to highlight such tumors.

#### CONCLUSION

The desmoid tumor is a rare, benign but aggressive tumor with frequent recurrences. Surgery is not systematic and should be reserved for aggressive lesions. The aggressive potential can be determined by a cytogenetic study. Surgery when considered should be as broad as possible. Radiation therapy is possible in case of incomplete resection or inoperable patients. We report the case of a patient with non-operable dual scapular and pulmonary localization for whom simple monitoring has been scheduled showing lesion stability after two years.

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#### COMPETING INTERESTS

The authors declare no competing interests.

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