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Case report

Chest wall Desmoid tumor…A multidisciplinary care

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Abstract

Desmoid tumors, though rare, can present challenges, especially when occurring in the chest. This case involves a 44-year-old woman with a parietal swelling, initially painless but progressively growing. "Café au lait" spots were noted, along with a left paraspinal mass. Imaging revealed an infiltrating parasternal mass extending to adjacent structures. An ultrasound-guided biopsy confirmed the diagnosis of desmoid type fibromatosis. After initial monitoring, surgery became indicated due to tumor growth. The patient had a total en bloc resection of the tumor process taking out the anterior arches of the 3rd to the 5th left rib with their intercostal spaces, the lower and left part of the sternal body as well as the xiphoid appendage. A polypropylene plate was used to construct the parietal defect. In order to cover the loss of soft tissue, a myoplasty with a pure and pedicle left pectoralis major muscle flap was performed. The final anatomo-pathological examination of the specimen concluded to a complete resection of the tumor with healthy margins. The patient's postoperative course was uncomplicated with no local recurrence after 12 months. Despite the benignity of desmoid tumors, they do represent a real local danger considering their aggressiveness. The tumor resection must be complete with adequate margins in order to decrease the risk of local recurrence.

Keywords: Desmoid Tumor; Case report; Chest wall; Surgery; Multidisciplinary care

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1. Introduction

Desmoid tumors, also known as aggressive fibromatoses, account for 3.5% of all fibrous tumors [1]. These rare tumors are highly invasive and recurrent but without metastatic potential. Uncommon in the chest, they pose diagnostic and therapeutic challenges [1]. Diagnosis relies on anatomo-pathological examination, guided by thoracic CT and MRI.

The management of this type of tumor represents a challenge. Indeed, the way in which these tumors are treated has changed over the years. While "Active monitoring" is favored [2], surgery is considered for locally advanced cases. In these cases, resection is often extensive and complex parietal reconstruction may be necessary. Adjuvant radiotherapy and non-steroidal anti-inflammatory treatments are other options.

2. Case report

We report the case of a 44-year-old woman, without any notable pathological history, who presented with growing parietal swelling. It was a left basithoracic swelling that has rapidly increased in size. The examination showed multiple "café au lait" spots (Fig.1) spread over the whole body with a left anterior paraspinal mass, solid, fixed to the deep plane, painless, without signs of inflammation. The thoracic MRI showed a four-centimeter left anterior paraspinal thoracic infiltrating mass with extension to the anterior arches of the 4th rib (Fig.2).

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An ultrasound-guided biopsy was performed and the anatomo-pathological study of the specimens concluded to a desmoid type fibromatosis. The decision was to initially monitor the patient, but the evolution was marked by an increase in size and the mass became increasingly painful.

One month later, the patient was operated on. Through an elective incision (Fig.3), she had a total en bloc resection of the tumor taking out the anterior arches of the 3rd to the 5th left rib with their intercostal spaces, the lower and left part of the sternal body as well as the xiphoid appendage (Fig.4).

A polypropylene plate was used to construct the parietal defect (Fig.5A). In order to cover the soft tissue loss, a myoplasty with a pure and pedicle left pectoralis major muscle flap was performed (Fig.5B). The postoperative course was uneventful and the patient was discharged at 8 days postoperatively.

The final anatomo-pathological examination of the specimen concluded to a complete resection of the tumor with healthy margins. At 12-month follow-up, no local recurrence has been noted.

3. Discussion

Desmoid tumors, first described in 1832,are locally aggressive [3]. Macroscopically, desmoid tumors are hard and firm, gray or whitish in color. Microscopically, the cells are characterized by heterogeneous, ill-defined and uniform proliferation of spindle cells reflecting myofibroblasts enveloped in an abundant collagen stroma and a capsule-free vascular network. There is no atypia, necrosis or mitosis [3].

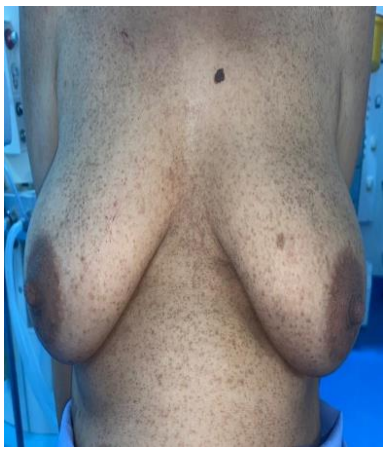

Fig. 1. 'Café au lait' spots

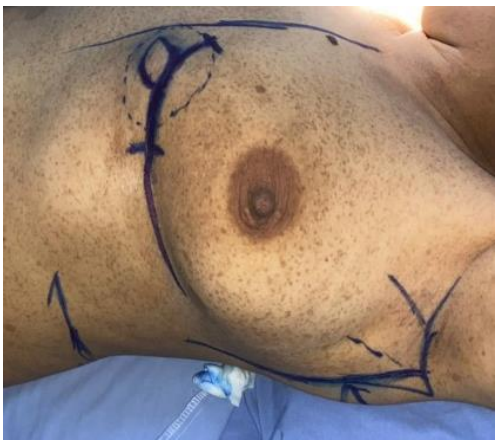

Fig. 3. Limits of the surgical incision

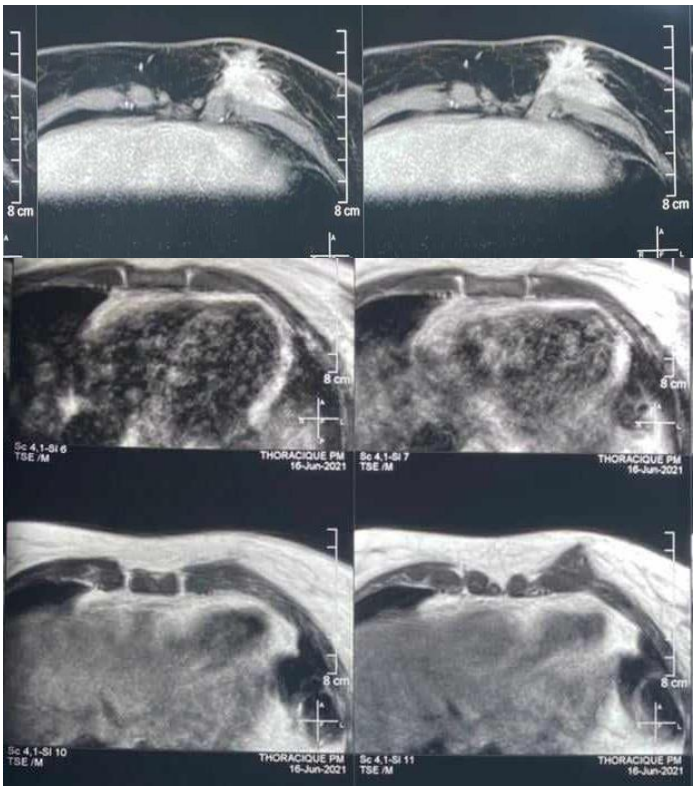

Fig. 2. The thoracic MRI showing the tumor mass

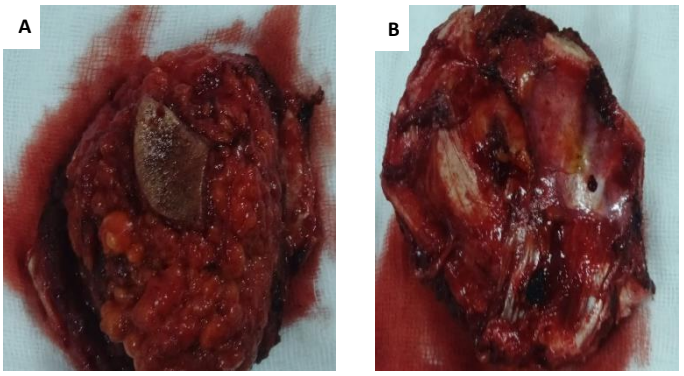

Fig. 4. Photo of the specimen (A: Anterior view/ B: posterior view)

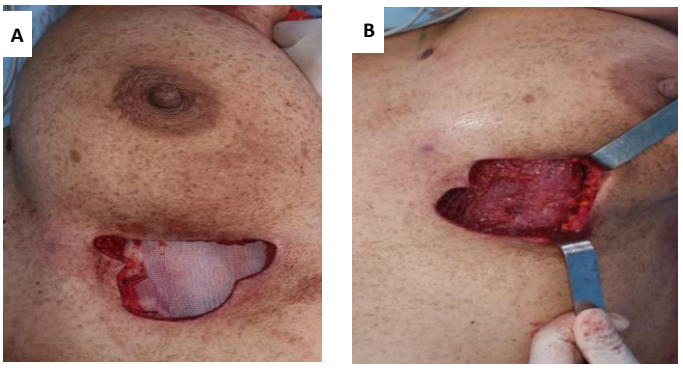

Fig. 5. (A) The coverage of the parietal defect with a polypropylene plate. (B) The coverage of the soft tissue defect with a left pectoralis major muscle flap

These tumors are predominantly seen in young females, with an average age ranging between 30 and 40 [4]. As they can be asymptomatic, desmoid tumors can be disabling. This depends on their location [3].

Radiological investigations, such as thoracic CT and thoracic MRI, are crucial for diagnosis and follow-up [3,5]. The diagnosis of desmoid tumor can only be confirmed by pathological examination of a transparietal or open biopsy. Needle biopsy seems to be the most commonly used method [6,7].

The evolution of desmoid tumors is unpredictable. As they may regress spontaneously, they may become locally aggressive. Surgical treatment has long been considered the reference and 1st-line treatment for these tumors. However, in recent years, the therapeutic strategy has changed, and surgery is no longer the 1st-line treatment. Today, surgical treatment is reserved for invasive forms or localizations that could jeopardize vital or functional prognosis [2,6-9].

In this case, surgery can only be performed when it is not risky and when complete resection with healthy margins is guaranteed [1,7,10-12]. Desmoid tumorsare characterized by a local recurrence rate of 20 to 65% within 5 years [1,13,14]. Thus, local recurrence remains a concern. The three golden rules for surgery of desmoid tumors are: a complete and carcinologically satisfactory resection, an adequate parietal repair to restore normal respiratory function and a coverage by good quality of flaps [15-18].

For resections with healthy margins, adjuvant radiotherapy has shown no difference in the control of local recurrences compared to surgery alone [2,6,13]. Hence, for the patients operated on for desmoid tumor with R0 resection, regular monitoring is all that's needed [1]. However, for R1 and R2 resections, radiotherapy is essential. It is recommended at a dose of 50-56 Gy. It is also

recommended in the event of local recurrence [19]. As for chemotherapy, it is indicated for tumors with rapid evolution and for tumors of critical location [19]. Moreover, some non-steroidal anti-inflammatory treatments have shown efficacy in the treatment of desmoid tumors [1,20].

In conclusion, despite their benign nature desmoid tumors pose a local threat and require careful management. In fact, they are known by their slow progressive growth, their local invasion as well as their local recurrence after excision with no metastatic potential. The 'Wait and see' approach is recommended, and if surgery is indicated, complete and satisfying resection is crucial to minimize local recurrence risk.

Consent of patient

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

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Conflict of Interest Disclosures

All authors declare that they have no conflict of interest.

Authors' contributions

The patient was operated by TC, NM and FS. Drafting of the manuscript was done by FS. The manuscript was corrected by IM, CK and SJ. All authors have read and agreed to the published version of the manuscript.

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