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Desmoid Tumor of the Abdominal Wall: About 3 Cases

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Abstract **Case Report**

Desmoid tumours of the abdominal wall are a rare entity that presents a therapeutic challenge, particularly for large tumours. MRI is the preferred radiological examination for suggesting the diagnosis, detailing the depth of extension, and defining the excision margins of the tumour. We present three cases of desmoid tumours of the abdominal wall in patients who underwent tumour resection followed by abdominal wall reconstruction with a bifacial mesh. The postoperative course was uncomplicated.

Keywords: Desmoid Tumor, Abdomen, Parietoplasty.

INTRODUCTION

Abdominal desmoid tumours, also known as desmoid fibromatoses, are rare, benign (non-cancerous) tumours that develop in connective tissue. Despite their benign nature, they can exhibit locally aggressive behaviour and invade surrounding structures, complicating their treatment. They account for approximately 3% of soft tissue tumours, with an estimated incidence of 2 to 4 cases per million people [1].

Accurate diagnosis is challenging, and therapeutic management is complex, requiring a multidisciplinary approach involving surgeons, radiologists, and oncologists due to the unpredictable progression and functional consequences of the disease [2]. Treatment often necessitates radical surgery with wide, full-thickness excision, sometimes referred to as "transfixing."

PATIENTS AND OBSERVATIONS

Observation 1

This is a 34-year-old patient with a history of childbirth one year ago, presenting with left abdominal swelling for the past 3 months. Clinical examination upon admission revealed a mass in the left hypochondrium measuring 15 x 10 cm (Figure 1). The remainder of the examination was unremarkable.

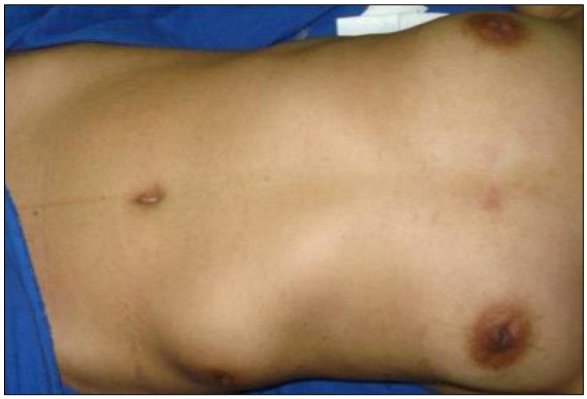

Fig. 1: clinical appearance of the mass

The patient underwent a biopsy of the mass, and the histological analysis confirmed a desmoid tumour. The CT scan revealed a tumour mass affecting the left anterolateral wall in its upper third (Figure 2). The assessment for tumour extension was negative.

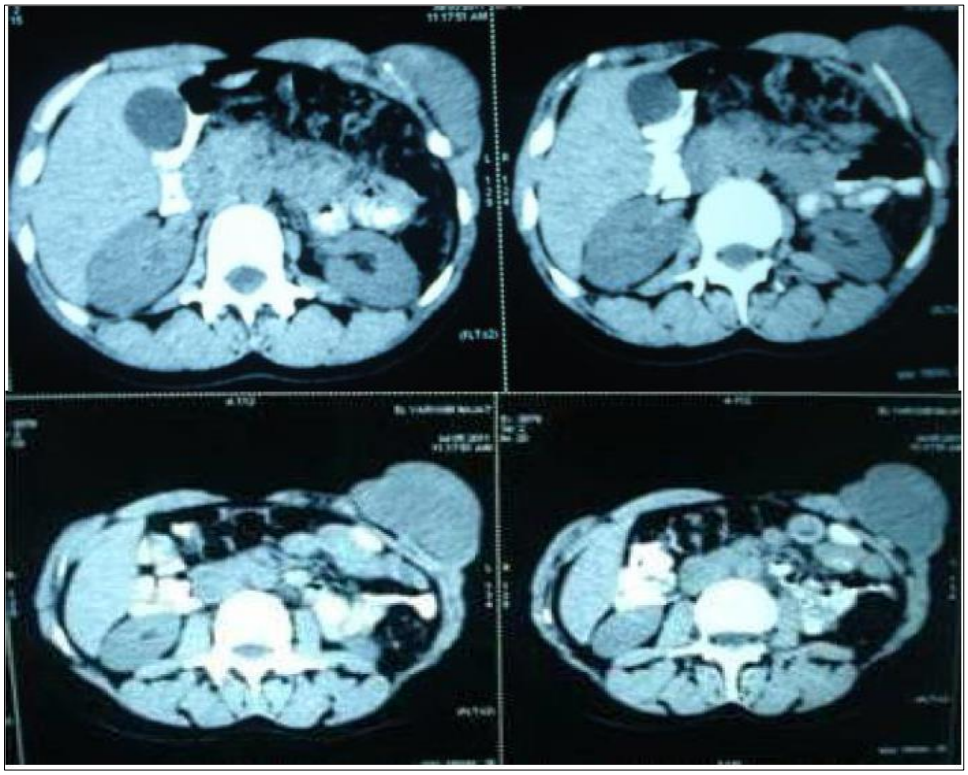

Fig. 2: Axial CT scan sections revealed a tissue mass in the abdominal wall at the level of the left hypochondrium (HCG). The mass was isodense with the rectus and oblique abdominal muscles and did not enhance following contrast administration

A one-piece excision of the tumor was carried out while removing surface a skin spindle including the biopsy scar. Then the repair of the deep plane was carried out by a bifacial mesh with translation of the remaining rectus abdominis and direct closure of the cutaneous plane by simple sliding (Figure 3). An MRI was done after 1 year, came back normal

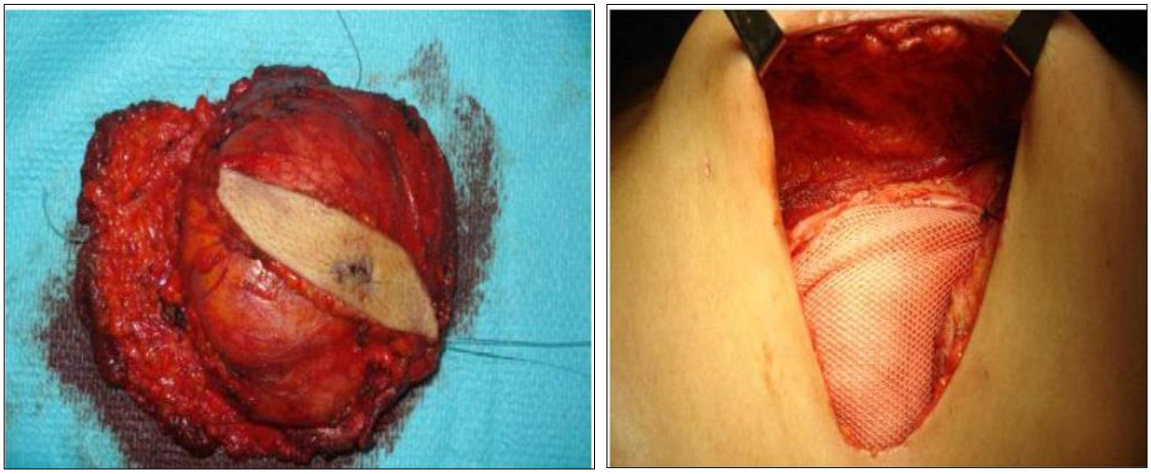

Fig. 3: Excision of the tumor and repair of defect by bifacial mesh

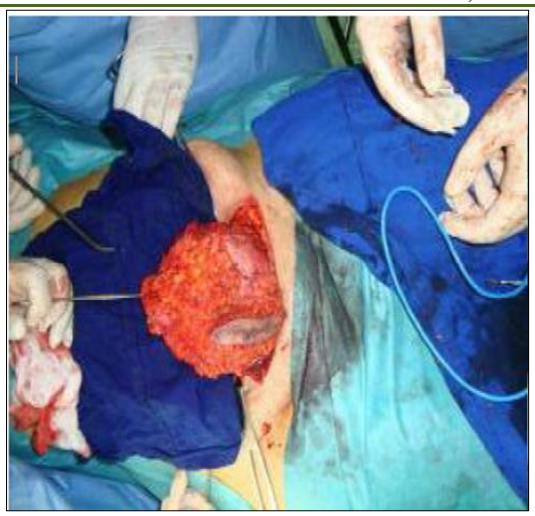

Fig. 4: Monoblock transfixing exeresis of the tumor mass

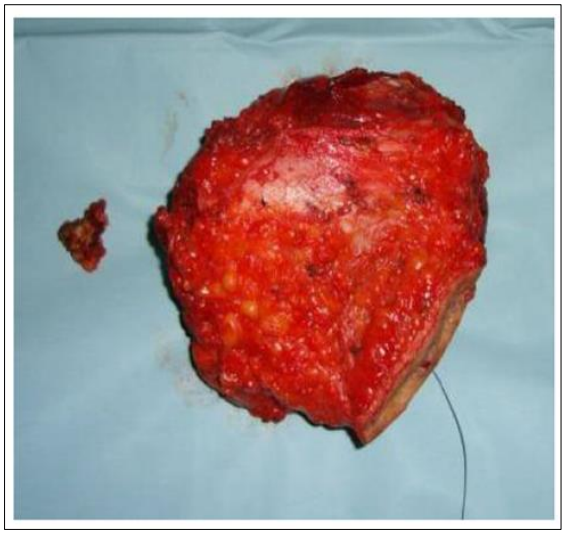

Fig. 5: Operating room

Observation 2

This was a 34-year-old patient with no notable pathological history, who presented 1 year after vaginal delivery with a desmoid tumor of the abdominal wall at the expense of the right iliac fossa reaching contact with the iliac bone.

MRI with preoperative assessment were carried out and returned in favor of a desmoid tumor. The expansion assessment: negative.

with general anesthesia: transfixing excision at a 3 cm margin removing the peritoneum and periosteum on the pubis (Figure 4). The histological study of the surgical specimen (Figure 5) confirmed the diagnosis of aggressive fibromatosis. The consequences were simple. A 6-month clinical and CT scan showed no signs of local recurrence.

Observation 3

This is a 46-year-old patient, with no notable pathological history, who consulted in June 2020 for an abdominal mass that had been present for more than a year.

The examination showed a mass in the left false iliac bone, non-painful and mobile in relation to the deep plane. Abdominal MRI showed a tissue mass at the expense of the rectus muscle of the left abdomen, roughly oval with lobulated contours measuring 78X54 mm, slightly hypodense which takes up the contrast after injection of gadolinium (Figure 6). Faced with this picture, a fibromatous desmoid tumor was suggested.

The patient benefited from a single-piece resection of this mass, removing the cutaneous plane, subcutaneous plane, the invaded muscular part as well as its aponeuroses and the adjacent peritoneal plane with a minimum macroscopic surgical safety margin of 1 cm lateral and in depth. Wall closure required a reconstruction procedure using a bifacial synthetic prosthesis (Figure 7). The histological study of the surgical specimen confirmed the diagnosis of aggressive

fibromatosis. A 6-month clinical and CT scan showed no signs of local recurrence.

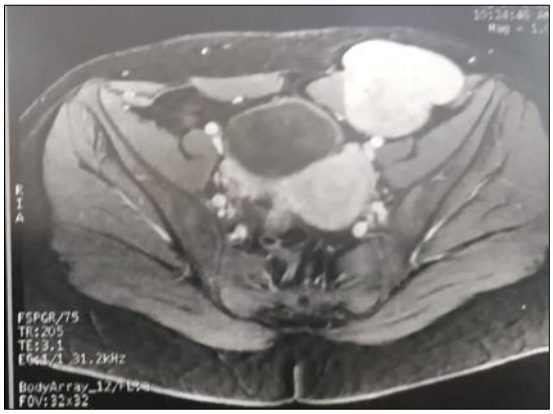

Fig. 6: MRI showed a tissue mass at the expense of the rectus muscle of the left

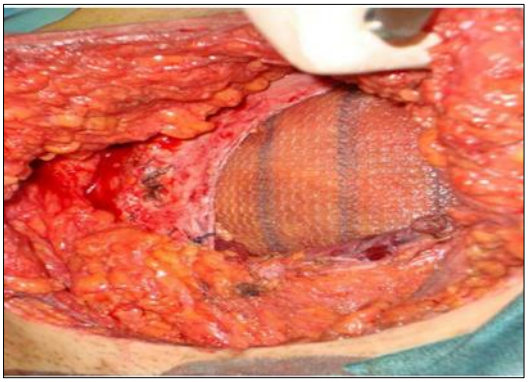

Fig. 7: reconstruction procedure using a bifacial synthetic prosthesis

DISCUSSION

Desmoid tumor (TD), or aggressive fibromatosis or musculoaponeurotic fibromatosis, is a rare entity with an annual incidence of 2 to 4 new cases per million inhabitants. It represents 0.03% of all neoplasias and 3% of fibrous tumors [3, 4]. It can occur at any age with a peak between 30 and 40 years old. It affects both sexes with a female predilection and a sex ratio varying from 2F/1M to 5F/1M depending on the different series [5]. In our series the average age is 35.3 years and the sex ratio is 2 F/1M, consistent with the literature data.

The exact causes of desmoid tumors remain unknown, although several risk factors have been identified:

1. Genetic: Mutation of the APC (Adenomatous Polyposis Coli) gene is associated with familial adenomatous polyposis (FAP), a condition that increases the risk of developing desmoid tumors.
2. Trauma: Injuries or surgical procedures, particularly in the abdomen, can trigger the formation of these tumors.
3. Hormones: Estrogens appear to play a role in the development of desmoid tumors, which would explain the higher prevalence in women of childbearing age [3].

In 40% of cases, these tumors are abdominal, 85% of which are located parietal, with a musculo-aponeurotic origin, often at the expense of the rectus abdominis or internal oblique muscle of the abdomen, only 15% are located deep: mesenteric, pelvic or retro-peritoneal [6].

Intra-abdominal forms are revealed by abdominal pain, a progressive increase in abdominal volume or during a complication: intestinal obstruction, entero-mesenteric infarction or uretero-hydronephrosis [3].

Extra-abdominal forms predominate in the chest wall, shoulder and pelvic girdles, extremities and cervical region [4]. In our series, the 3 tumors described are of parietal abdominal location.

CT reveals a more or less well-defined mass of tissue density, iso or discreetly hypodense compared to the surrounding muscle, enhancing in a homogeneous, sometimes heterogeneous manner in large tumors with areas of necrosis, microhemorrhages and metaplasia [4, 5].

MRI allows better tissue characterization, better study of relationships with adjacent structures (nerves, vessels, deep organs) and differentiation of postoperative changes or after medical treatment of tumor recurrence [8].

Imaging mainly uses CT and magnetic resonance imaging (MRI) which are useful for evaluating the size and extension of desmoid tumors, studying surrounding structures and evaluating tumor recurrence.

Wide surgical excision with curative intent, when possible, is the treatment of choice despite the high risk of recurrence. Indeed, when the tumor is resectable, R0 surgery must be performed [8].

When surgery is not possible or in the event of recurrence, several therapeutic options can be considered:

- Nonsteroidal anti-inflammatory drugs (NSAIDs): Like sulindac, can slow tumor progression.
- Hormonal therapies: Antiestrogens such as tamoxifen.
- Chemotherapy: Cytotoxic agents such as doxorubicin or dacarbazine for refractory cases.
- Targeted therapies: Tyrosine kinase inhibitors such as imatinib [4-9].

Radiotherapy is used in some cases when surgery is not possible or for recurrent tumors. Radiation therapy can help control tumor growth.

In our series, all our patients benefited from a wide excision passing through healthy tissue with parietoplasty performed using a bifacial plate in the 2 women. The postoperative course was simple and no adjuvant treatment was used.

The histological study reveals a monoclonal proliferation of spindle-shaped fibroblastic cells surrounded and separated from each other by collagen fibers arranged in bundles [4].

In our series, the diagnosis of desmoid tumor was confirmed by the pathological study. Post-therapeutic monitoring is based on clinical examination and imaging, particularly MRI, which allows local recurrences to be better visualized. These recurrences appear within five years following surgery in 33 to 75% of cases, and depend on the quality of excision, the growth phase at the time of surgical treatment, the site and extension of the tumor [3].

All our patients benefited from clinical and scan monitoring over variable periods ranging from 8 months to 4 years with an average follow-up of 2 years and no locoregional recurrence was observed.

Desmoid tumors do not metastasize, but they can be locally aggressive and recurrent. The prognosis depends on the location, size of the tumor, and the ability to achieve complete resection. Optimal management often requires a multidisciplinary approach involving surgeons, oncologists, radiologists and pathologists.

CONCLUSION

desmoid tumors are aggressive fibromatoses due to their infiltrative nature with a high potential for recurrence. The diagnosis must be considered in the face of any abdominal parietal tissue mass. MRI makes it possible to make the diagnosis, guide therapeutic management and monitor progress. Confirmation of the diagnosis is histological. Treatment currently relies on an initial active surveillance strategy looking for signs of complications or progression.

Conflict of interest: The authors declare no conflict of interest.

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