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# Case Report of a Desmoid Tumor of the Abdominal Wall with Eventration: A Challenge for Resection and Repair

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## Article History

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**Abstract:** **Case Report**

Desmoid tumors are rare tumors with slow growth but with high local aggressiveness and a high risk of recurrence. These tumors can develop in the abdominal wall, intra-abdominally, or extra-abdominally. Their treatment is becoming increasingly codified with surgical indications becoming more and more limited. The challenge in surgery remains to perform an R0 resection in order to limit the risk of recurrence and to repair the abdominal wall in the event of a loss of substance. A case of a desmoid tumor of the anterior abdominal wall adherent to the rectus sheath associated with an incisional hernia at a McBurney scar was reported, presenting a surgical challenge of repairing the abdominal wall after tumor resection.

**Keywords:** Desmoid Tumor, Local Aggressiveness, R0 Resection, Abdominal Wall Reconstruction, Incisional Hernia.



## INTRODUCTION

According to the dictionary of the French Academy of Medicine, a desmoid tumor is a fibromatous tumor, named "desmoid" by Müller in 1838 due to its resemblance to the structure of a tendon, mainly observed in adults with a preference for women during their reproductive years, developing from the aponeurotic sheath, most often that of the rectus abdominis, and thus generally located in the abdominal wall.

Desmoid tumors of the abdominal wall present as palpable masses, most often in the rectus and external oblique muscles of the abdomen. Their size varies from 3 to 10 cm in diameter. They evolve gradually and slowly without ever crossing the midline [1].

They most often occur between puberty and age 40, with the incidence peaking between 25 and 35 years. Women are more often affected than men [2, 3].

## OBSERVATIONS

Ms. B. M. Age 43, with a history of a Mac Burney appendectomy in 2021, presents with an abdominal mass that has been evolving for 9 months with no other associated signs, including no bowel movement issues and a preserved general condition.

Clinical examination: found a healthy patient with an OMS score of 0, a Mac Burney laparotomy scar, an abdominal mass 4X7 cm below and laterally to the umbilicus, firm in consistency, well-defined, not painful, and fixed to the posterior plane. Presence of a uncomplicated ventral hernia at the Mac Burney scar.

**Diagnostic Approach**

**Abdominal and pelvic CT:** a tumor mass is found in the anterior abdominal wall, lateral and sub umbilical on the right, with no clear border with the rectus abdominis muscle, with regular contours measuring 58X78 mm vs 45X53mm (scan performed 4 months prior), with tissue density and a slight infiltration of the surrounding fat.

Evidence of a right uterine mass that is roughly oval in shape, measuring 53 x 93 mm, with cystic density and thin partitions that come inward in close contact with the small intestine and outward from the lateral abdominal wall without a separating interface.

Eventration at the level of the right iliac fossa with a 7.5 cm parietal defect containing intestinal and omental contents.

**Surgical biopsy:** The tumor lesion that suggested a desmoid tumor was confirmed by a surgical biopsy.

**Surgical Intervention**

The patient underwent surgery with a monobloc excision of the tumor lesion, including the underlying aponeurotic and muscular plane, via a transverse oval incision with umbilical preservation, and repair of the abdominal wall reinforced by the placement of a non-absorbable polypropylene plate in front of the posterior layer of the rectus abdominis muscle covered by the aponeurosis of the external oblique muscles and the rectus abdominis muscle, with multiple release incisions. The intra-abdominal lesion was a false cyst filled with clear liquid with no signs of malignancy on cytology.

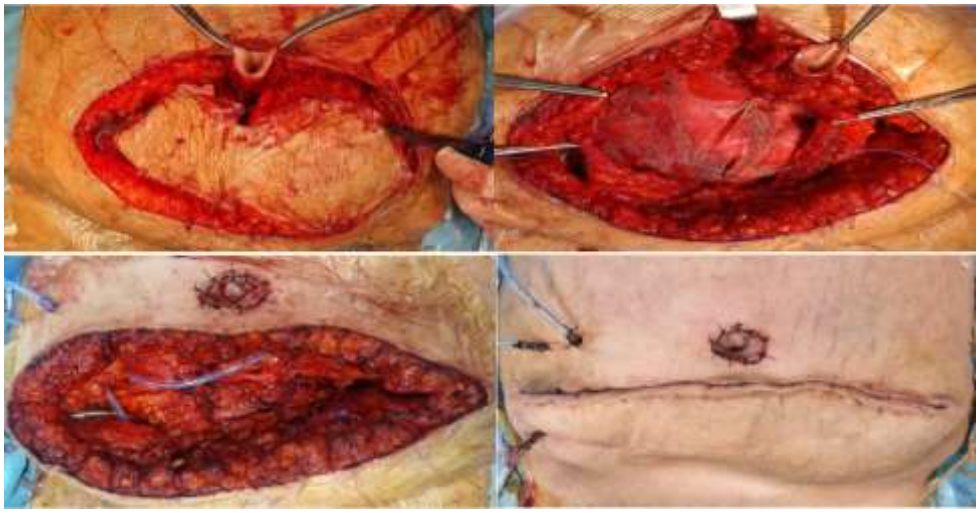

**Image 1: Per-operative images of the mass and repair of abdominal wall**

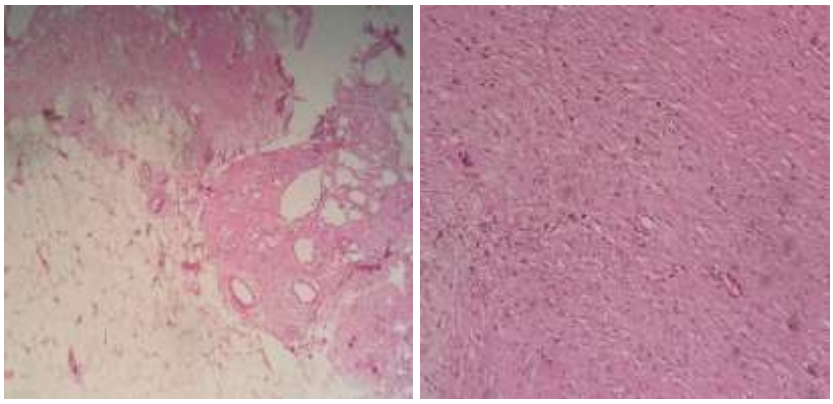

**Image 2: HE Staining X20 Poorly defined fibroblastic proliferation encompassing a few adipose foci**

## DISCUSSION

Desmoid tumors are rare benign tumors that were first described by John Mac Farlane in 1832 [4], they can be:

- Sporadic: the most common, 85 to 90% of cases associated with a mutation in the coding pathway for beta-catenin CTNNB1.
- Part of a tumor predisposition syndrome associated with APC (adenomatous polyposis coli) gene mutation in patients with familial adenomatous polyposis (FAP) [5, 6].

At present, no etiological hypothesis has been confirmed. Three etiopathogenic factors take center stage: traumatic factors, based on the relative frequency of TD on surgical scars. Other genetic and hormonal factors appear to be involved, given the marked incidence of this type of tumor in patients with familial polyposis, or in women taking estrogen-progestin [1], in our case, we mention the traumatic cause due to the appendectomy preceding the appearance of the tumor.

Magnetic resonance imaging (MRI) is the preferred method for imaging desmoid tumors. A paradigm shift in the management of these lesions has occurred in recent years, with a shift towards active surveillance first, followed by systemic or local symptomatic treatment in the case of an aggressive tumor. Therefore, once the diagnosis is established, MRI plays an important role in the monitoring protocol with a direct impact on the management and evaluation of treatment efficacy [7], however the definitive diagnosis is histological based on an anatomical pathological study of a percutaneous biopsy or the surgical specimen.

Managing desmoid tumors remains difficult. They are benign tumors, but their unpredictable clinical evolution makes them similar to low-grade fibrosarcoma; in 10% of cases, they regress spontaneously (especially after menopause); one-third alternate between phases of growth and regression; approximately 50% remain stable; and 10% have rapid growth [1]. Depending on the location and size of the tumor, as well as other factors, the risk of recurrence after surgical treatment can be high [8, 9].

The terms of care have evolved over the past ten years. For most patients, surgery is no longer the preferred primary treatment and has been replaced by active surveillance or alternative treatments, such as systemic therapy [8].

The NCCN (National Comprehensive Cancer Network) and DTWG (Desmoid Tumor Working Group) guidelines (Figure 1-3) recommend an initial period of active surveillance with ongoing follow-up for asymptomatic tumors that do not cause functional limitations. Because surgery or radiation therapy in patients with progressive, morbid, or symptomatic tumors may be associated with poor long-term functional outcomes [10], systemic therapy (chemotherapy or tyrosine kinase inhibitors) is recommended in these cases.

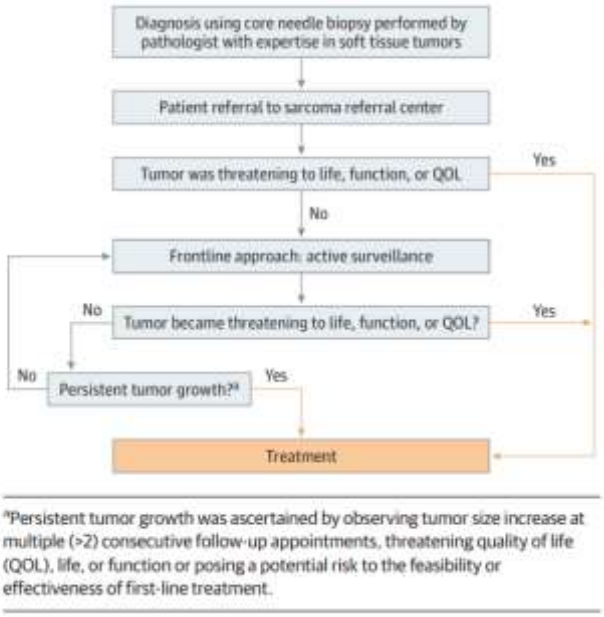


**Figure 1: DTWG guidelines for management of desmoid tumors**

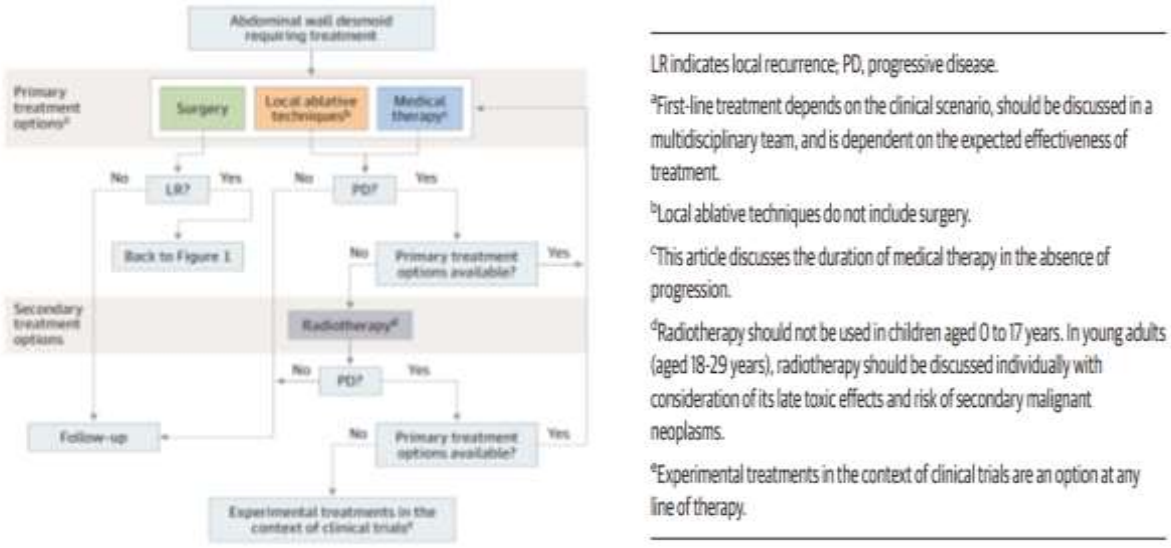


**Figure 2: DTWG guidelines for management of abdominal wall desmoid tumors**

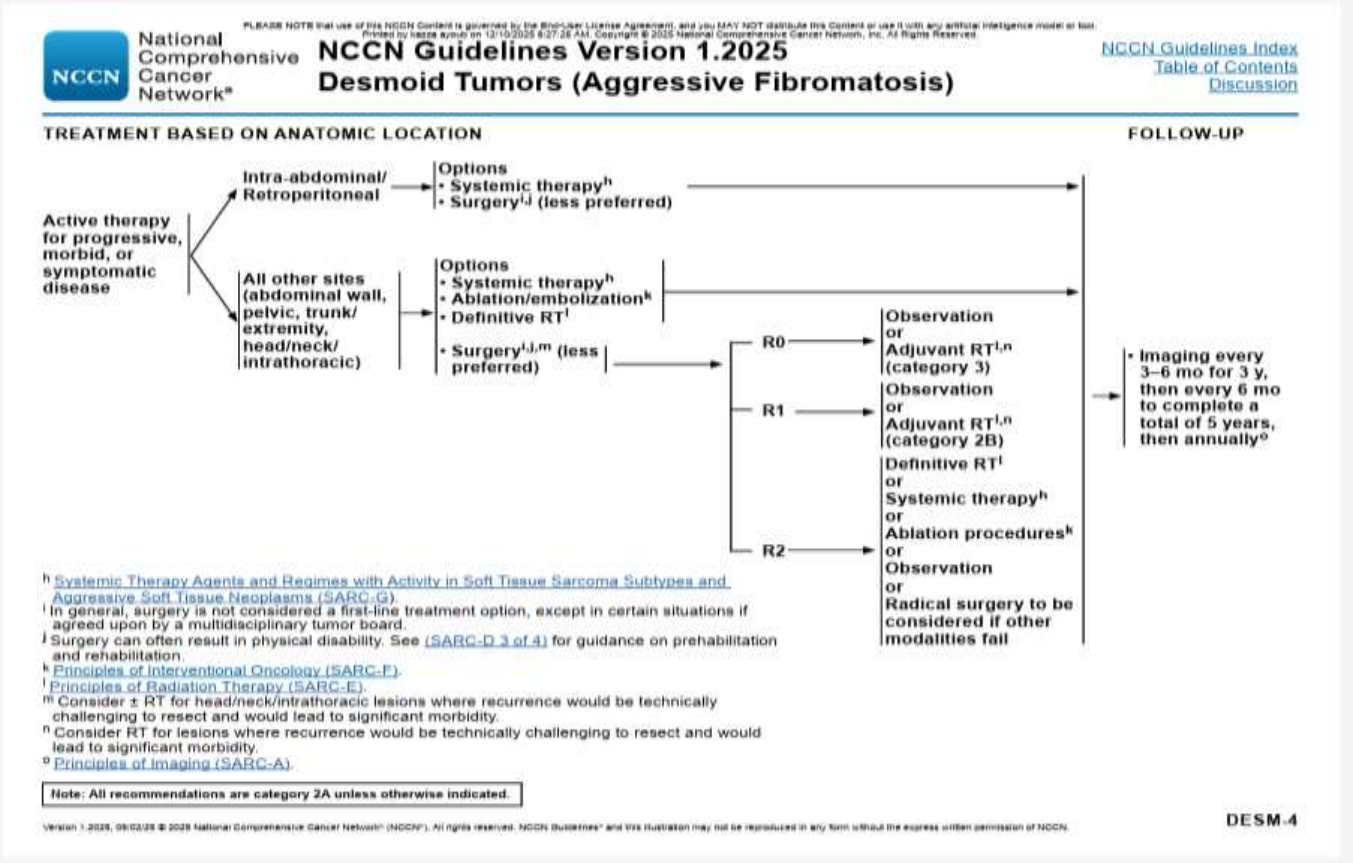


**Figure 3: NCCN Guidelines for desmoid tumors**

In the case of surgery, no consensus has been established regarding the surgical technique for closing abdominal defects. The surgeon will have the option to use synthetic parietal prostheses or free muscle-skin flaps and aponeurotic approximation procedures [11].

In our clinical case, surgery was indicated due to the tumor's increasing size, which was associated with a parietal defect, and the surgical intervention aimed to prevent a complication of ventral hernia and provide curative treatment for the patient.

## CONCLUSION

Desmoid tumors are rare with slow and unpredictable progression; they are never metastatic, but their invasive potential and recurrence rate after surgery make them feared tumors, this evolution conditions their management, which has become more codified with multiple therapeutic options.

**Conflicts of Interest:** The authors declare no conflict of interest.

### Authors Contributions

All authors contributed to the conception and drafting of this manuscript. They also state that they have read and approved the final version of the manuscript.

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