Management of giant desmoid type fibromatosis of abdominal wall

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Arch Oncol 2026;32(1):30-34

https://doi.org/10.2298/AOO250413012N

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Received: **2025-04-13**
Received in revised form: **2025-07-27**
Received in its last revision form: **2025-09-26**
Accepted: **2025-09-26**
Published online: **2025-11-07**

SUMMARY

Introduction: *Desmoid type fibromatosis (DT) represents a rare benign tumor formation characterized by local aggressive behavior but an absence of metastatic potential. Histologically, they are often misdiagnosed, leading to unnecessary radical surgery. Even with proper diagnosis, there are different approaches and unclear protocols for the treatment of DT.*

Case Presentation: *The study presents the case of a 33-year-old male who presented to the emergency department with a large swelling of the abdominal wall that had been present for two years. An ultrasound was performed, and the abdominal swelling was described by the radiologist as a ventral hernia. After further radiological studies (CT scan and magnetic resonance imaging), a diagnosis of a tumor of the abdominal wall was established. A partial biopsy led to the diagnosis of sarcoma. Surgery was planned as the tumor was growing rapidly, reaching around 30 cm and causing necrosis of the abdominal wall skin. After the resection of 8.5 kg of tumor with clinically clear margins, the defect was reconstructed by placing a polypropylene mesh and using a darn nylon suture with a local rotational skin flap. In the postoperative days, an infection of the surgical wound developed. The infection resolved in two weeks without further complications. The definitive histopathological diagnosis was desmoid type fibromatosis.*

Conclusion: *Active surveillance is now regarded as the most acceptable strategy for the treatment of DT. A better understanding of the genetic alterations underlying DT and the identification of prognostic factors that favor aggressive behavior will lead to the development of appropriate individual strategies and tailored therapy for each patient.*

Keywords: *aggressive, fibromatosis, desmoid tumor, abdominal wall, reconstruction*

INTRODUCTION

Desmoid-type fibromatosis (DT) is a rare, benign tumor known for its locally aggressive behavior and multifocal presentation without metastatic potential. The incidence is approximately 2-5 cases per million per year in Europe, predominantly affecting women aged 30-40 years (1,2). Although DTs can grow rapidly and resemble malignant tumors, they remain benign, primarily causing local tissue invasion and recurrence without jeopardizing the patient's life. DTs commonly occur extra-abdominally in the abdominal wall or extremities but can also appear intra-abdominally. Current treatment favors active surveillance over radical surgery due to DT's potential for spontaneous regression. However, surgery becomes necessary for aggressive, disabling forms, as illustrated in this case report.

CASE PRESENTATION

A 33-year-old male presented to the emergency department with a large, rapidly growing abdominal wall swelling that had been present for two years. Initially thought to be a ventral hernia based on an ultrasound, further radiological studies (CT and MRI) revealed a tumor in the abdominal wall. The MRI described a large, lobulated, well-demarcated soft tissue mass measuring 28x18x25 cm (Figure 1).

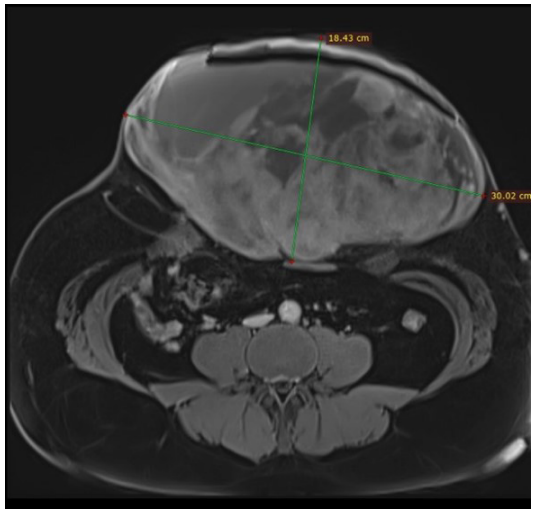

Figure 1. Magnetic resonance of abdominal wall tumor.

Alt contained both solid components and necrotic areas, indicating a myxoid component. The tumor infiltrated the left rectus abdominis muscle, extended towards the subcutis and the greater omentum, and was in close contact with the transverse colon. No intra-abdominal abnormalities or lymph node involvement were noted. The radiologist suggested the mass was likely a sarcoma, possibly pleomorphic. Before surgery, the patient developed fever and skin necrosis over the tumor. Despite these symptoms, he had been asymptomatic aside from mild discomfort and pain. There was no family history of colorectal cancer, nor

did he have any prior medical conditions.
He was admitted to the Department of Plastic and Reconstructive surgery, antibiotic therapy was initiated, and the skin wound was dressed daily until the infection was solved and the patient was prepared for surgery (Figure 2).

An incisional partial biopsy was performed, and preliminary histopathological analyses suggested sarcoma of unknown origin, possibly liposarcoma. Immunohistochemical profile: Vimentin+, CD10+, β-catenin+, a1-antitripsine+, SMA-, CD68-, CD34-, S100- (Figure 3).

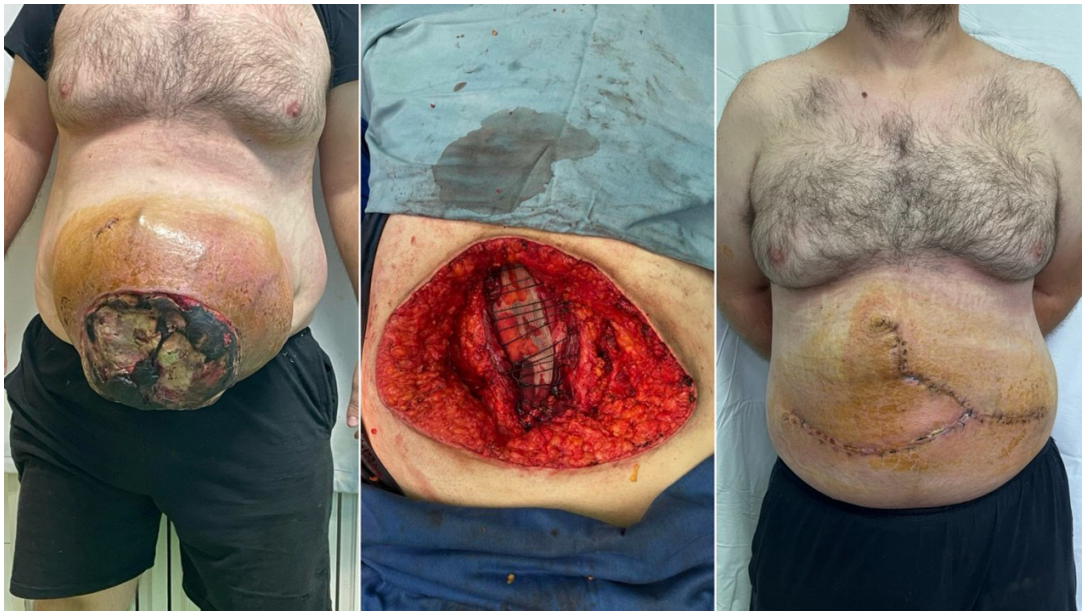

Figure 2. Patient with large abdominal wall tumor – before, during and after surgery.

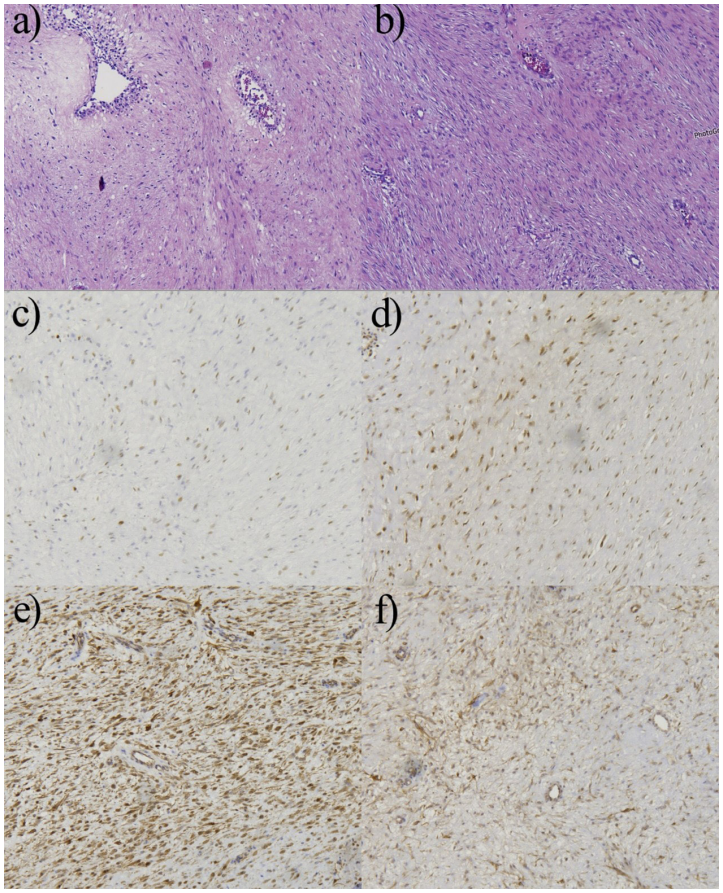

Figure 2. Histopathological presentation of the tumor (objective magnification ×10). a,b) HE; c) LEF1+; d) AR+; e) β-catenin+; f) CD10+.

The patient underwent surgery under general anaesthesia with the collaboration of both abdominal and plastic surgeons. Complete resection of the tumor was performed including partial resection of both rectus muscles, as the tumor had infiltrated them. The tumor was in contact with the peritoneum, and the abdominal cavity was opened during resection. No involvement of the intestines or any other intra-abdominal organs was observed. After achieving macroscopically clear margins, a 25 cm defect of the abdominal wall remained, including a defect of the medial half of both rectus muscles. Various scenarios for reconstruction of the expected abdominal wall defect had been considered preoperatively in order to make the best decision depending on the amount of muscle and peritoneum that might need to be resected. The risk of using a polypropylene mesh in potentially contaminated tissue was considered, but in the end, the abdominal surgeon opted for mesh placement and a "tension-free" darn repair as a double reinforcement for defect closure. The peritoneum was closed directly, the polypropylene mesh was placed over the fascial defect, and a Nylon darn repair was performed above the mesh, connecting the free margins of the remaining rectus muscles (Figure 2.). A large proximally based rotational flap was created on the right side of the abdominal wall, and the surrounding tissue was undermined to facilitate defect closure. The tumor was located in the subcutaneous tissue and associated with a large area of overlying skin necrosis. It measured 40x40x24 cm and included surrounding adipose tissue covered by ulcerated skin measuring 26x23.5 cm. The total weight was 8.5 kg. It appeared relatively well-demarcated and encapsulated. The base of the tumor contained grayish-white, friable deposits. On cross-section, the tumor displayed a solid, whitish structure with a glossy appearance, interspersed with areas of haemorrhage and necrosis. The tumor tissue also contained multiple small cystic spaces filled with mucinous, yellowish material. On histological sections, tumor tissue is made up of two cell types: spindle and stellate cells, with elongated nuclei and eosinophilic cytoplasm arranged in bundles and the other ones round to epithelioid cells. Blood vessels are elongated and branched. There are smaller foci of necrosis. The stroma consists of dense collagen fibers and extensive myxoid areas. The tumor tissue initially infiltrates to striated muscle fibers. The immunohistochemical profile of the tumor is: AR+, LEF1+, BCL2-, CD31-, CD99-, Caldesmon -, ER-, PR-, MDM2-, myogenin-, myoD1- (Figure 3). The histopathological examination differed from previous analyses of the biopsy sample and a diagnosis of desmoid type fibromatosis was established.

In the early postoperative period, there was seropurulent secretion in the central part of the wound with clinical signs of wound infection. *Klebsiella* and *Enterococcus faecalis* were isolated in the wound. Antibiotics were prescribed according to the bacteriological analysis and the wound dressings were changed twice a day. In two weeks, the infection was resolved without compromising the skin flaps and polypropylene mesh underneath. The patient was discharged and returned to usual activities. No additional therapy was advised by the oncologist.

After two months, the first postoperative MR was performed, and no tumor was detected. Further close follow-up is planned.

DISCUSSION

Two forms of DT are described in clinical practice - the sporadic one (85-90%) and the Familial adenomatous polyposis type (FAP)-associated form. Our patient had no knowledge of FAP in his family, nor had he undergone any previous abdominal surgeries, so his form is considered sporadic. Nevertheless, we advised him to undergo colonoscopy in order to exclude undiagnosed FAP.

The main challenge in managing DT lies in identifying the most effective treatment strategy, as there is currently no clear consensus. Two major approaches to DT treatment exist: one emphasizes surgery as first-line therapy, while the more recent, conservative concept of active surveillance delays or avoids surgical intervention, based on the observation that DTs may spontaneously regress or stabilize over time. *Fiore* et al. conducted a study involving 142 patients with DT in order to evaluate the "wait-and-see" approach and concluded that this conservative strategy was justified, as progression-free survival (PFS) was comparable between the active treatment group and the observation group (3). In contrast, Gour et al. examined the outcomes of surgery as a first-line treatment for two DT subtypes: intra-abdominal fibromatosis (IAF) and abdominal wall fibromatosis (AWF) (4). Their findings emphasized the importance of surgical intervention in preventing disease progression, particularly in cases where the "wait-and-see" approach was not suitable (4). Furthermore, a recent study by Penel et al. compared surgical and non-surgical approaches by assessing event-free survival (EFS) (5). They found that the anatomic location of the tumor significantly influenced the outcomes (5). In favorable locations such as the abdominal wall, breasts, lower extremities, intra-abdominal cavity, and digestive viscera, EFS was similar in the two strategies. However, in unfavorable sites, including the chest wall, head and neck, and upper extremities, the EFS was significantly better in the patients managed with the active surveillance approach. In our case, surgical treatment was indicated based on the initial histopathological diagnosis suggestive of sarcoma, which necessitated timely and aggressive management. However, even if the initial diagnosis had indicated a desmoid tumor, the decision for surgical intervention would have remained the same, given the exceptionally aggressive clinical behavior of the lesion. The rapid local progression, size, and infiltrative nature of the tumor posed a significant risk to surrounding structures, justifying a proactive surgical approach regardless of histological classification. A whole spectrum of doubts and options would have opened if the tumor was smaller, asymptomatic, and initially diagnosed as DT.

Desmoid tumors are frequently (30-40%) misdiagnosed histologically as soft tissue sarcomas due to a significant morphologic overlap (6,7). Penel et al in a large survey published in 2016 analysed 861 cases confirmed by expert pathologists. In one large referral center study, 29% of DTs were initially misclassified, with some erroneously diagnosed as low-grade fibrosarcomas or unclassified spindle cell sarcomas (8). Both entities may exhibit spindle cell morphology, infiltrative growth, and a collagenous stroma, especially on limited or superficial biopsies, which can hinder accurate interpretation by general pathologists (1). Although immunohistochemical staining for nuclear β-catenin is a key diagnostic tool for DT, it is not entirely specific. Nuclear β-catenin positivity is observed in up to 80–90% of desmoid tumors, but similar staining can also be seen in up to 30% of other soft tissue neoplasms, including certain low-grade sarcomas and benign fibrous lesions (9). This limits its diagnostic utility when used in isolation. The diagnostic accuracy can be improved by combining β-catenin with LEF1 immunostaining, which increases specificity to 96% (10).

Another challenge was determining how to close the abdominal wall defect following tumor resection, as the patient developed a skin infection adjacent to an area of necrosis caused by the tumor's progressive growth just days before surgery. When planning the surgical strategy, we carefully considered all the relevant factors: the potential exposure of abdominal viscera, the need for the partial resection of the rectus muscles, the possibility of the contamination of the tumor-involved area, the feasibility of a component separation technique, and the availability of local tissue flaps with consideration for donor site morbidity. A key question was whether open mesh repair in a potentially contaminated field would be safe, or whether the component separation technique should be favored.

In addition to the potential contamination associated with the tumor itself, we ruled out other common risk factors for infection, such as smoking, obesity, diabetes, and enterotomy. Based on this risk assessment, the abdominal surgeon determined that the polypropylene mesh would be an acceptable option in this case. Birolini et al. conducted an important clinical study on the use of a synthetic mesh in contaminated abdominal wall repairs and demonstrated that complication rates were similar to those seen in clean ventral hernia reconstructions (11). Furthermore, several authors have published guidelines on abdominal wall reconstruction, as this remains a complex issue often dependent on the surgeon's experience and expertise (12-16).

Regarding skin closure, the plastic surgeon had several reconstructive options, depending on the extent of skin that would need to be excised. In the event that local flaps were insufficient to cover the defect, an anterolateral thigh (ALT) flap was considered the next best option. Ultimately, following complete tumor resection, the abdominal surgeon closed the fascial defect using a polypropylene mesh reinforced with a Nylon darn suture, applied in a tension-free manner above the mesh. The skin defect was successfully reconstructed using a proximally based rotational flap raised on the right side of the defect, with mobilization of the lower abdominal skin. A postoperative wound infection was managed effectively with antibiotics and regular wound dressings, without further complications or the need to remove the mesh.

Besides surgery and the "wait-and-see" approach there are case reports in literature describing various medical treatments used to address DT such as different hormonal agents, chemotherapy, NSAIDs, interferons and others (17). Testa et al. did a retrospective analysis of 262 patients with DT and concluded that active surveillance was a treatment of choice with the best EFS in all patients, in small and minimally symptomatic DT it was the best choice while in bigger, more aggressive tumors similar results were seen in patients treated with tyrosine kinase inhibitors, local ablation or surgery (18). Scientists also analysed the response of DT to radiotherapy or radiotherapy and surgery as a combo treatment (19-21). Baumert et al. published data confirming that postoperative radiotherapy significantly improved EFS compared to just surgery treatment especially in non-radical surgery (20). Goy suggested that radiotherapy has an important role in positive margins after a wide resection of the recurrent tumor but a questionable role in clear margins (21). The consensus group, keeping in mind all previous studies, said that this combo therapy approach can be applicable for recurrent DM where further surgery in case of recurrence would be difficult to perform (6). The level of evidence for radiotherapy as an adjuvant therapy is considered low. All these different approaches in the treatment of DT suggest a lack of consensus in the management of this tumor. Penel et al. suggested in a recent study (2021) that the stepwise approach should be considered with aggressive forms of DM, while active surveillance with periodic MRs for other milder forms (22). He underlined the quality of life of these patients and that the side effects of every new medical treatment, including morbidity and recurrence rates involved with surgery must be discussed with the patient.

The Desmoid Tumor Working Group and European Consensus Initiative emphasize that active treatment should be considered only in cases of persistent progression and even then, if localisation is considered favourable and there are no significant symptoms, surgery should be postponed (6,23). As our patient developed a huge tumor that invaded the skin and produced massive necrosis in a short time leading to infection and hight fever, we decided to go for a surgery and leave active surveillance for the follow-up and possible recurrence treatment strategy, considering that in such aggressive forms recurrences are frequent, voluminous and unresectable (24).

CONCLUSION

A better understanding of the molecular and genetic alterations underlying DT and determining the prognostic factors that influence the progression of tumor and favour aggressive behaviour, will lead to the creation of an appropriate individual strategy and tailored therapy for every patient. Active surveillance is considered today the dominant strategy in the treatment of DT. Furthermore,

organizing referral centres with expert pathologists will reduce the misdiagnoses of DT and help avoid unnecessary surgeries. This will facilitate the decision-making process for physicians and support an improved quality of life for patients with DT.

Acknowledgments: None
Authors declare that there is no conflict of interest.
The patient provided informed consent to participate in this scientific study.

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