

Desmoid-Type Fibromatosis of the Rectus Abdominis: An Uncommon Anatomical Manifestation

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Abstract

A female patient presented with irregular menstrual cycles and a progressively enlarging lower abdominal swelling two years after undergoing a lower-segment cesarean section. Clinical examination revealed a large, firm abdominal wall mass extending from the right iliac fossa into the umbilical region with restricted mobility and attachment to the abdominal wall. Magnetic resonance imaging demonstrated a well-defined solid mass arising from the right rectus sheath, compressing adjacent pelvic structures including the urinary bladder and uterus while preserving fat planes. Trucut biopsy with immunohistochemistry confirmed desmoid-type fibromatosis, showing strong nuclear β -catenin positivity along with smooth muscle actin expression and low proliferative activity.

The patient underwent neoadjuvant chemotherapy followed by wide local excision of the tumor. Intraoperatively, the lesion was found adherent to surrounding structures including the bladder, uterus, and pubic symphysis, necessitating extensive resection involving the rectus muscles and peritoneum. The resulting large abdominal wall defect required complex reconstruction using bilateral transversus abdominis release, polypropylene mesh reinforcement, fascia lata grafting, and rotational flap coverage to restore structural integrity.

This case highlights the rare presentation of desmoid-type fibromatosis arising from the rectus abdominis following cesarean section and emphasizes the importance of multidisciplinary management. Accurate diagnosis through imaging and immunohistochemistry, combined with meticulous surgical planning and advanced reconstructive techniques, is essential for achieving optimal functional and oncological outcomes in extensive abdominal wall desmoid tumors.

BACKGROUND

Desmoid tumors, also referred to as deep fibromatosis or aggressive fibromatosis, are rare mesenchymal neoplasms characterized by a dense, fibrotic, band-like consistency(1). Although histologically benign, such tumors exhibit locally invasive behavior, often infiltrating surrounding tissues, yet they lack metastatic potential. The principal contributors to morbidity and, less commonly, mortality in affected individuals include local recurrence and involvement of adjacent organs.

The estimated incidence of desmoid tumors in the general population is between 2 to 4 cases per million persons each year, with a slight female predominance. The highest occurrence occurs during the 3rd and

4th decades of life. In younger patients, these tumors often exhibit more aggressive clinical activity; some studies have shown recurrence rates as high as 87% (3). In individuals with familial adenomatous polyposis (FAP) who have had a colectomy, desmoid tumors represent a significant source of morbidity as well as mortality (4).

Radiologically and grossly, desmoid tumors often demonstrate spiculated, infiltrative margins and a tendency to adhere to surrounding anatomical structures. Histologically, they are made up of fibroblasts and uniformly produced spindle-shaped cell fascicles that are poorly defined and embedded in a thick collagenous stroma. Necrosis is uncommon, and mitotic figures are frequently rare (2,5).

Desmoid tumors most commonly arise in proximal extremities, abdominal wall, and mesentery—particularly in individuals with FAP (6). In sporadic cases, they may develop at sites of previous trauma, surgical scars, or areas exposed to radiation.

CASE PRESENTATION

A female patient presented with a history of irregular menstrual cycles and a gradually enlarging, non-tender mass in the lower abdomen. 2 years earlier, she had had a lower-segment cesarean section (LSCS). On clinical examination, a visible abdominal mass was noted in the right iliac fossa region, approximately 15 × 20 cm in size. The mass was irregular in shape, with a conical upper border extending just above the umbilicus and an ill-defined lower border. The overlying LSCS scar was intact, the umbilicus appeared normal, and there were no dilated veins or visible pulsations. No pedal edema was noted in the right lower limb, and the left flank was free.

Palpation revealed a large firm mass measuring approximately 21 × 17 cm, extending from the right iliac fossa and suprapubic area into the umbilical region. The mass had a smooth surface, with restricted side-to-side mobility and was fixed in the craniocaudal plane. Leg-raising and head-raising tests confirmed The abdominal wall was attached to the abdominal wall. The upper and lateral borders were well-defined, while the lower border merged with the mons pubis and right inguinal ligament; the left inguinal ligament was easily palpable. The remainder of the abdomen was soft. Femoral and dorsalis pedis pulses were palpable on the left side, while the right dorsalis pedis was not palpable.

Imaging Findings

MRI of the abdomen and pelvis revealed a large, well-defined solid pelvic-abdominal mass originating from the right rectus sheath. The lesion was compressing the fat planes of anterior abdominal wall musculature, with splaying of right rectus muscle. Posteromedially, the mass abutted the anterior myometrium of the uterus with preserved fat planes. Inferiorly, it caused a mass effect on the dome of urinary bladder, again with preserved fat planes. Laterally, it compressed right iliopsoas muscle and the right common iliac vessels.

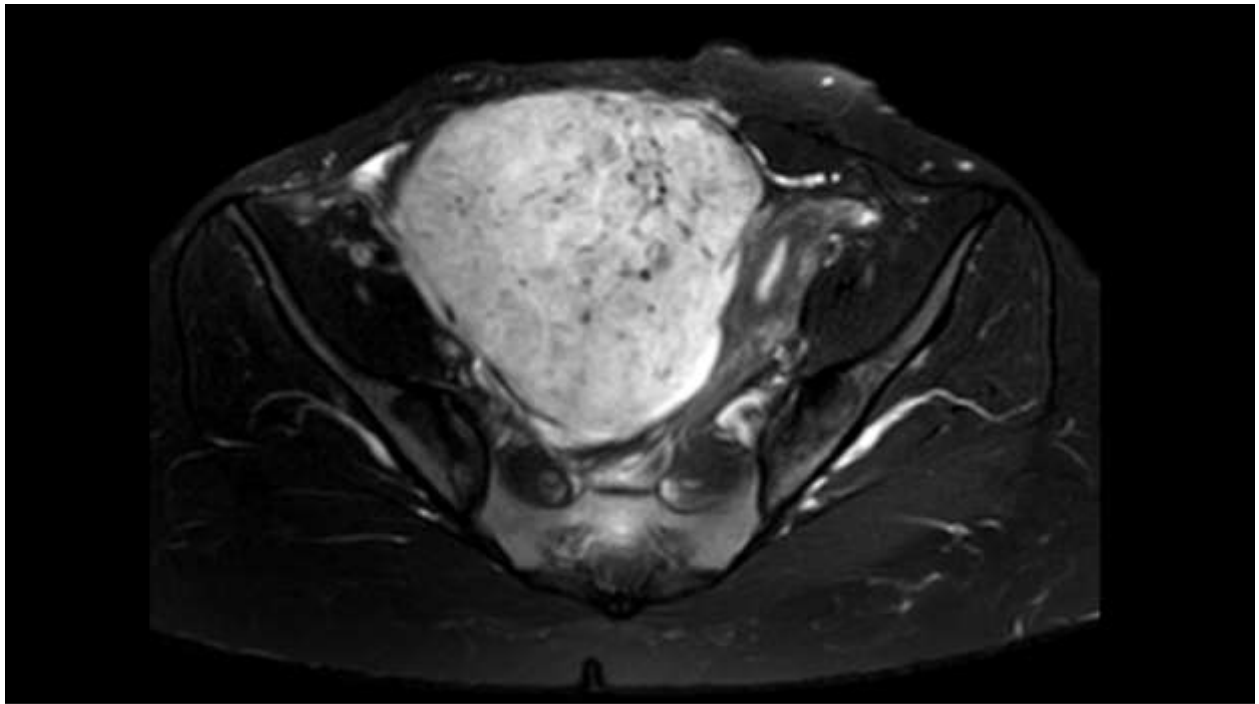


Figure 1: T2W flair image showing tumor abutting the uterus

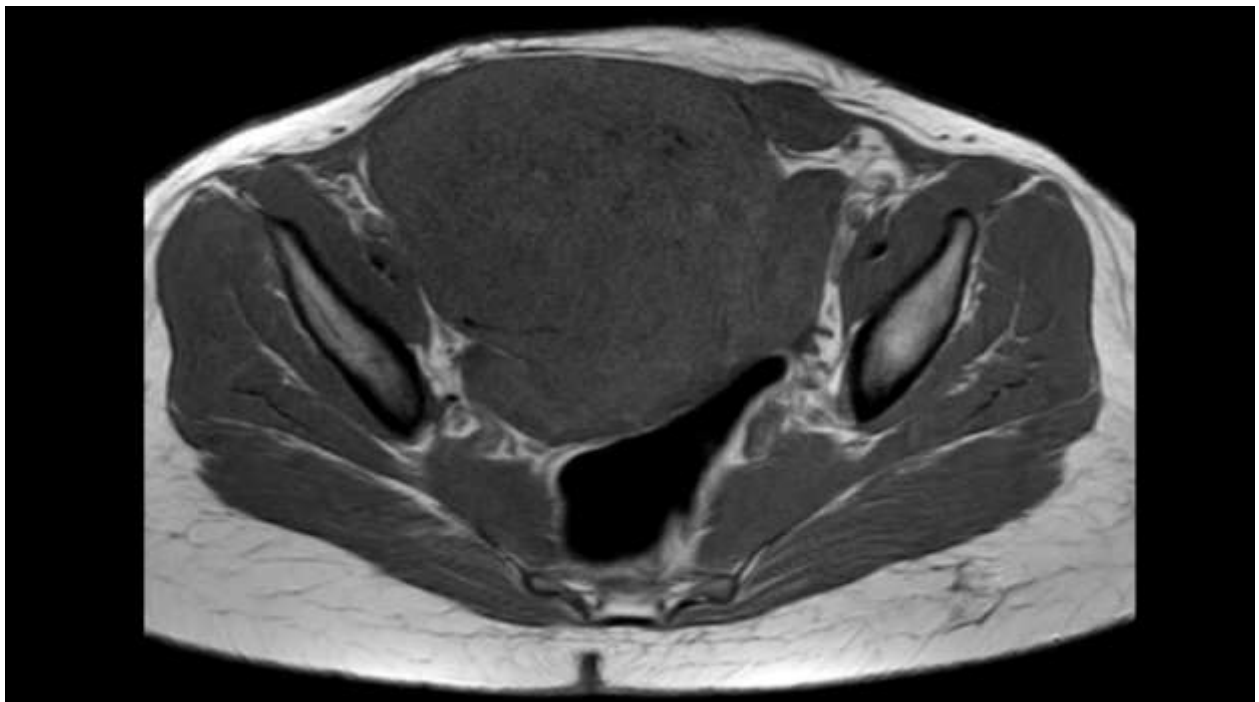


Figure 2: T1W image showing tumor abutting the urinary bladder

Histopathology and Immunohistochemistry

A trucut biopsy obtained via the anterior abdominal wall demonstrated histological features consistent with fibromatosis. Immunohistochemical (IHC) analysis revealed:

- SMA: Strong cytoplasmic positivity in 70% of spindle cells
- β -catenin: Strong nuclear positivity in 80% of spindle cells
- CD34 & S100: Negative

- Ki-67: Labeling index of 3–5%

These findings confirmed the diagnosis of desmoid-type fibromatosis.

Management

Patient received three cycles of chemotherapy with Adriamycin, Vinblastine, and Tamoxifen, followed by surgical intervention for wide local excision of the tumor.

Intraoperatively, the mass had displaced the rectus muscles toward the left. Both peritoneum and rectus muscles within oncologic margins were excised. The tumor had adhesions between it and other organs such as the bladder and uterus, which were carefully removed. The inferior of the tumor was attached to the pubic symphysis and had to be shaved off the pubic bone.

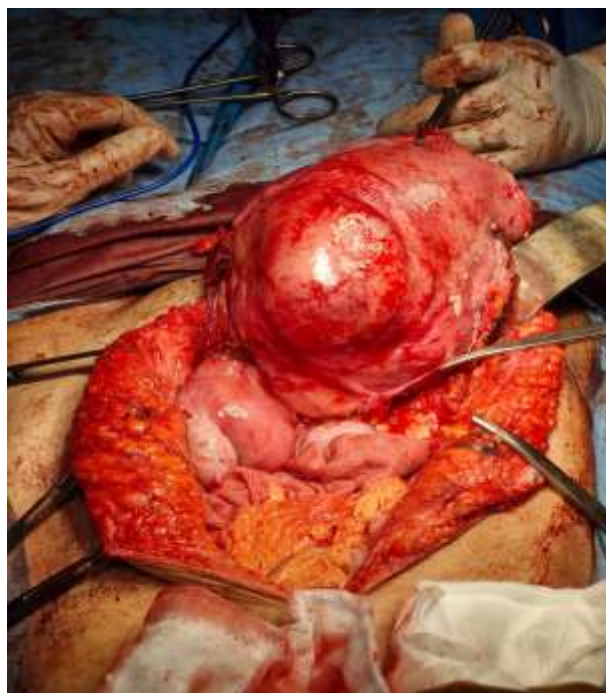


Figure 3: Shows tumor abutting the uterus



Figure 4: Shows tumor abutting the bladder

A large peritoneal defect measuring 20 × 20 cm was created following tumor excision. Bilateral transversus abdominis muscle release was performed, followed by advancement and primary closure of the peritoneum. A 30 × 30 cm polypropylene mesh, tailored to 22 cm longitudinal and 25 cm horizontal dimensions, was secured to reinforce the abdominal wall. Romovac drains were placed over the mesh. To further strengthen the abdominal wall, a left fascia lata graft and a right fascia lata rotational flap were harvested and sutured over the repair site.

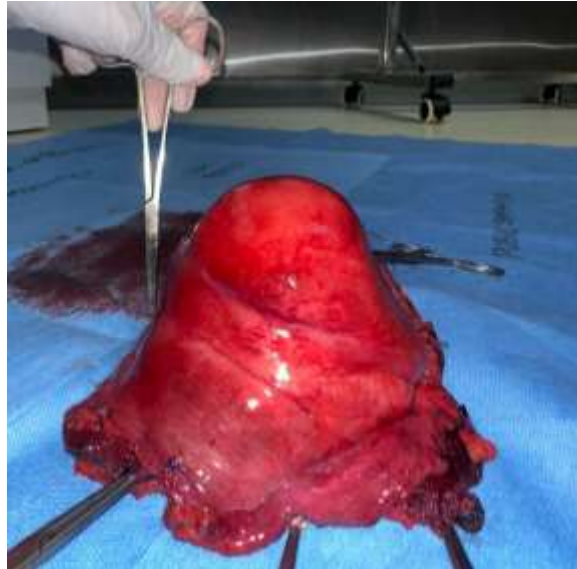


Figure 5: Excised Desmoid Tumor



Figure 5: Polypropylene mesh used to strengthen the anterior abdominal wall

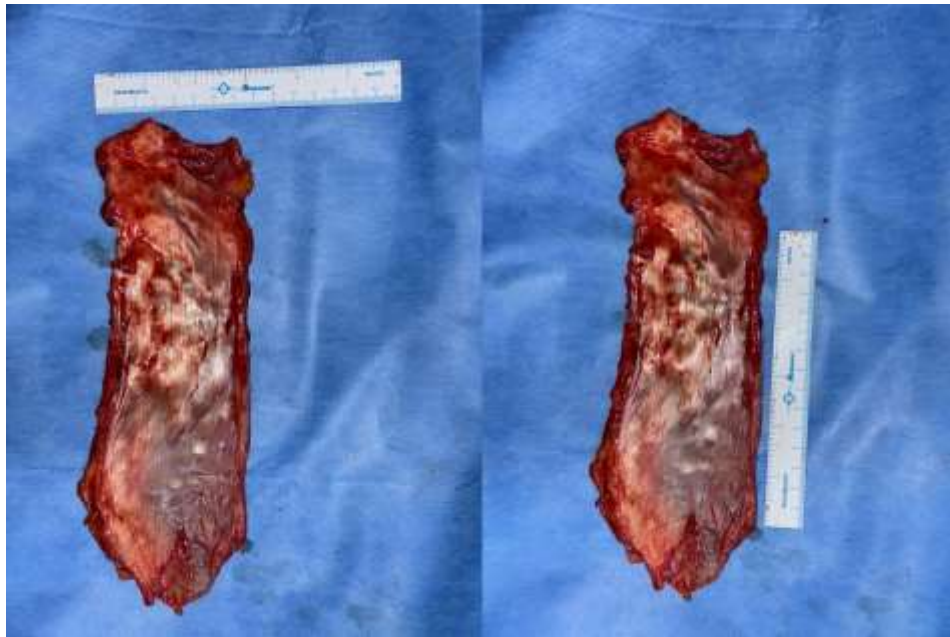


Figure 6: Tensor Fascia Latae graft taken from left thigh



Figure 7: Vascularised Tensor Fascia Latae flap harvested from right thigh



Figure 8: Reconstruction of the anterior abdominal wall.

DISCUSSION

Desmoid tumors (DT), also known as deep fibromatoses (DF), are benign but locally aggressive fibroblastic neoplasms with an infiltrative growth pattern that commonly develops from the skeletal muscle's fascia or aponeuroses (7). These tumors are more frequently observed in females and have well-established associations with familial adenomatous polyposis (FAP)(8), as well as with prior surgical trauma or intervention(9).

It is essential for the histopathological diagnosis to recognize a DF/DT from other fibroblastic or myofibroblastic proliferations and from smooth muscle neoplasms. To distinguish desmoid tumors from histologic mimics, immunohistochemical analysis—particularly, nuclear β -catenin staining—is essential. The interpretation of β -catenin staining is crucial because it is only the nuclear localization that is diagnostically important (10).

But it is important to understand that the absence of nuclear β -catenin staining does not exclude a desmoid tumor diagnosis. Nuclear staining can be focal or patchy and cannot be observed in small or superficial biopsy specimens, thus requiring careful interpretation of limited tissue samples (11).

The main aim of surgically managing desmoid tumors is to attain radical resection with histologically clear margins. Nonetheless, depending on the location of the tumor, extensive resections can cause significant soft tissue defects, especially on the abdominal wall (12,13). Although smaller full-thickness abdominal wall defects may often be repaired with direct suture repair methods (14), larger or more complicated defects often require reconstruction with synthetic mesh to provide structural integrity (15). The use of distant or free muscle flaps has become supported by literature in recent years as a method of reconstruction in cases where local tissue flaps are inaccessible or inadequate. Such advanced reconstruction methods are especially helpful in the case of large abdominal wall deficiencies in which long-term coverage and functional restoration are critical. (13,14).

ABBREVIATIONS

1. computed tomography scan (CT)
2. familial adenomatous polyposis (FAP)
3. magnet-resonance-tomography (MRT),

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