



Case report

Giant Abdominal Wall Desmoid Tumor with Mass Effect: A Case Report and Review of the Literature

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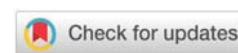
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Abstract

Desmoid tumors, or aggressive fibromatoses, are rare fibroblastic proliferations of soft tissues. Although benign from a metastatic standpoint, they possess a high potential for local invasion. Giant forms, inducing a compressive mass effect, pose complex therapeutic challenges. This case report highlights the critical decision-making process in giant abdominal wall desmoid tumors where immediate salvage surgery takes precedence over conservative active surveillance due to severe symptoms and tissue compression. We report the case of a 46-year-old female patient presenting with a large mass on the abdominal wall evolving over 6 months, causing chronic pain, functional impairment, and a significant mass effect on adjacent pelvic structures. Magnetic resonance imaging (MRI) revealed a large infiltrating tissue lesion. Biopsy and immunohistochemistry confirmed the diagnosis of aggressive fibromatosis. Given the significant mass effect, wide surgical excision with tumor-free margins was performed. The postoperative course was uneventful, with no local recurrence at the 6-month follow-up.

Conclusion: The management of giant desmoid tumors with mass effect requires a personalized, multidisciplinary approach, oscillating between monitoring, medical alternatives, and salvage surgery.

Introduction

Desmoid tumors, or deep fibromatoses, are rare entities representing less than 3% of all soft tissue tumors and approximately 0.03% of all neoplasms. Characterized by a monoclonal proliferation of fibroblasts and myofibroblasts, they are distinguished by the absence of metastatic potential, but by marked local aggressiveness and a high recurrence rate [1-3].

While recent international guidelines advocate for initial active surveillance (“wait-and-see”) in asymptomatic or indolent desmoid tumors, management strategies must rapidly pivot when faced with “giant” presentations (typically >10 to 15 cm). These large masses frequently generate a severe compressive mass effect, jeopardizing adjacent anatomical

structures and visceral function. The primary scientific contribution of this report lies in illustrating the precise clinical and radiological tipping points—specifically scar-tissue history, rapid expansion, and severe symptomatic compression—that necessitate abandoning conservative observation in favor of immediate, margin-clear surgical resection to achieve disease control and functional restoration [3].

Patient and observation

Clinical presentation

A 46-year-old female patient with no notable medical history, but with a surgical history of four prior cesarean sections and one myomectomy, consulted for a progressively enlarging, painful abdominal mass over the past 6 months.

Physical examination revealed a firm, fixed, poorly mobile mass located in the lower abdominal wall and pelvic region, measuring approximately 11 cm in its largest diameter. The mass exerted a marked compressive mass effect, resulting in severe local pain, a persistent feeling of heaviness, daily functional impairment, and significant aesthetic deformity.

Diagnostic and methodological evaluation

To establish an accurate anatomical map and rule out visceral invasion, an abdominal ultrasound combined with contrast-enhanced pelvic Magnetic Resonance Imaging (MRI) was performed. MRI remains the gold-standard imaging modality for defining tumor boundaries, fascial involvement, and proximity to major neurovascular bundles.

- **MRI Findings:** High-resolution T1-weighted, T2-weighted, and contrast-enhanced T1-fat-suppressed sequences demonstrated a large, well-circumscribed yet locally infiltrating mass measuring 11 × 8 × 7 cm originating within the rectus abdominis muscle and anterior pelvic wall fascia. The lesion exhibited heterogeneous signal intensity on T2-weighted images with pronounced enhancement following gadolinium administration, reflecting high cellularity and active fibroblastic proliferation. Significant compressive mass effect was noted on the anterior uterine wall and adjacent bladder dome, without direct intravascular extension or intraperitoneal organ erosion (Figure 1).

Histopathology and immunohistochemistry

To obtain diagnostic certainty before surgery, an image-guided core needle biopsy was performed.

Histological Analysis: Microscopic examination revealed a uniform, noncapsulated proliferation of elongated spindle cells (fibroblasts and myofibroblasts) arranged in long, sweeping fascicles within an abundant collagenous stroma. The cells displayed pale amphophilic cytoplasm with regular, oval nuclei containing fine chromatin and small nucleoli. There was a notable absence of marked nuclear atypia, pleomorphism, hyperchromasia, or atypical mitotic figures, ruling out high-grade soft tissue sarcomas (such as fibrosarcoma).



Figure 1: IRM abdomino-pelvienne.

High-resolution pelvic MRI (Axial T2-weighted and T1 contrast-enhanced views) illustrating a giant tissue mass (11cm) located in the anterior abdominal wall, exerting a distinct compressive mass effect on adjacent pelvic viscera.

Immunohistochemical (IHC) Profile: Immunohistochemical staining demonstrated intense, diffuse nuclear positivity for β -catenin, which is pathognomonic for desmoid-type fibromatosis resulting from CTNNB1 or APC gene pathway alterations. The tumor cells were negative for CD117 (c-Kit), DOG-1, CD34, and S100, definitively excluding gastrointestinal stromal tumors (GIST), solitary fibrous tumors, and neural sheath tumors.

Therapeutic management and follow-up

Following discussion in a Multidisciplinary Oncology Tumor Board (MTB), immediate surgical excision was deemed mandatory due to the progressive nature, size, severe symptoms, and mechanical pressure on pelvic organs.

An *en bloc* wide surgical resection of the parietal mass was performed, maintaining a 2 cm healthy surgical margin around the pseudocapsule. Reconstruction of the abdominal wall defect was achieved using a synthetic mesh to ensure anatomical integrity. The immediate postoperative course was uneventful. At the 6-month follow-up, clinical examination and control MRI demonstrated complete restoration of abdominal wall function and no signs of local recurrence.

Discussion

Desmoid tumors most often occur in young adults (between 30 and 40 years of age), with a marked female predominance. Their etiology includes sporadic forms (most often linked to mutations in the β -catenin gene or resulting from trauma/scarring) and syndromic forms associated with Gardner syndrome [4].

Clinically, the presentation is closely dependent on the location. In giant abdominal wall tumors, the dominant feature is the mass effect, which can cause visceral, vascular, or nerve compression, or severe functional impairment [5].

L'MRI remains the imaging examination of choice for mapping tumor extension, assessing relationships with vascular structures, and planning treatment strategy. However, diagnostic certainty relies exclusively on histology combined with immunohistochemistry (positive catenin).

Therapeutic management has undergone a major paradigm shift over the past decade. While extensive surgery was the norm, the concept has evolved towards active monitoring ("wait and see") initially for asymptomatic or small tumors, given the high risk of postoperative recurrence (which can reach 30 to 50%) and the fact that surgery itself can stimulate tumor regrowth.

Gustave roussy

However, as our observation demonstrates, the appearance of a compressive mass effect or a major functional impact reinstates the surgical indication, ideally combined or preceded by medical alternatives (non-steroidal anti-inflammatory drugs, hormone therapy, tyrosine kinase inhibitors, or low-dose chemotherapy) if prior tumor reduction is deemed necessary.

Conclusion

Giant desmoid tumors of the abdominal wall are a rare condition, and their therapeutic management remains challenging. The compressive mass effect is a major deciding factor, leading to targeted surgical intervention. A coordinated approach within specialized oncology and soft tissue pathology networks is essential to optimize local control and minimize the risk of recurrence.

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