

Pregnancy-Associated Giant Paraspinal Desmoid-Type Fibromatosis Managed with Function-Preserving Resection

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Abstract

Background: Desmoid-type fibromatosis is a rare, locally aggressive fibroblastic neoplasm that does not metastasize but may cause considerable morbidity through infiltrative growth. Pregnancy-associated giant extra-abdominal paraspinal lesions are uncommon. Main objective of the study is to share our experience of Giant Paraspinal Desmoid-Type Fibromatosis managed with Function-Preserving Resection. A 26-year-old para 1 woman noticed a painless mass over the right upper back of chest during pregnancy 30 months ago. At that time, it measured approximately 4 cm and mass enlarged rapidly after delivery. Later it triggered back discomfort which was impairing daily activities. Examination shown a firm, immobile, and non-tender posterior thoracic mass measuring about 30 × 40 cm. Magnetic resonance imaging demonstrated a giant heterogeneous soft-tissue mass involving the posterior thoracic and paraspinal musculature which was closely related to the spine. A core-needle biopsy showed a benign spindle-cell neoplasm. Function-preserving surgical excision was performed. The morphological and immunohistochemical findings supported desmoid-type fibromatosis. Giant pregnancy-associated paraspinal desmoid-type fibromatosis is uncommon and presents difficult surgical decisions. In anatomically complex disease, preservation of function may appropriately take precedence over microscopic margin clearance.

Keywords: Desmoid-type fibromatosis; aggressive fibromatosis; pregnancy; paraspinal tumor; function-preserving surgery; case report.

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INTRODUCTION

Desmoid-type fibromatosis (DTF), also known as aggressive fibromatosis or desmoid tumor, is a rare monoclonal fibroblastic or myofibroblastic neoplasm. DTF has an estimated incidence of approximately 3–5 cases per million person-years.^[1-3] Although it does not metastasize, it is characterized by infiltrative local growth and an unpredictable clinical course. Its behaviour ranges from spontaneous stabilization or regression to progressive disease associated with pain, functional impairment and involvement of adjacent structures.^[1,2] Desmoid-type fibromatosis occurs most often in adolescents and young-to-middle-aged adults, although it may occur at any age.^[3,4] A female predominance is reported, particularly during the reproductive years, supporting but not proving a role for hormonal influences in tumor biology.^[3,5] Most sporadic desmoid tumors are associated with alterations in the CTNNB1 gene, resulting in abnormal accumulation of β -catenin within tumor-cell nuclei. A smaller proportion occurs in association with germline Adenomatous polyposis coli (APC) gene alteration and Familial Adenomatous Polyposis (FAP).^[1,2] Nuclear β -catenin immunoreactivity is therefore an important finding supporting the diagnosis, although it must be interpreted together with the morphological and clinical findings.^[6] About 90% of Desmoid tumours are sporadic and only 5-10% are associated with FAP. Only 5-16% of patients with FAP will develop a desmoid tumour in their lifetime.^[3] The most common locations were the abdominal wall (40.8%),

trunk (23.5%), limbs (19%), intraabdominal (13.5%) and head & neck (3.2%).^[4] Risk factors for development of desmoid tumours include surgery, trauma and pregnancy.^[3,7] Pregnancy and the postpartum period are recognized associations with desmoid-type fibromatosis. Proposed mechanisms include hormonal influences and the effects of mechanical stretching or tissue injury. Most pregnancy-associated tumors arise in the abdominal wall, making a giant extra-abdominal posterior thoracic or paraspinal tumor an unusual presentation.^[8] Management has evolved from routine wide surgical excision to an individualized strategy incorporating active surveillance, systemic treatment. When surgery is undertaken, preservation of function is important, particularly when complete excision would require sacrifice of major muscles, nerves or other critical structures.^[1,2] Main objective of the study is to share our experience of Giant Paraspinal Desmoid-Type Fibromatosis managed with Function-Preserving Resection.

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CASE PRESENTATION



Figure 1A: right upper posterior thorax mass (posterior view).
Figure 1B: right upper posterior thorax mass (Lateral view)

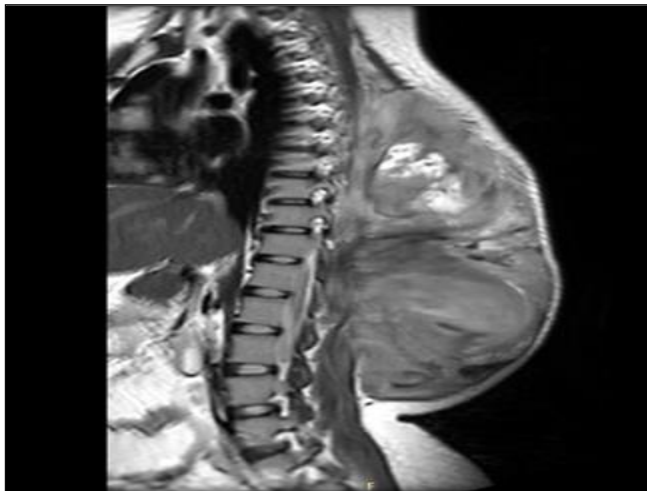


Figure 2: MRI: demonstrated a large heterogeneous posterior thoracic soft-tissue mass involving the paraspinal musculature and closely related to the thoracic spine with no obvious intraspinal extension.



Figure 3: Post-operative specimen. The tumour size measured 40 x 30cm.

A 26-year-old para 1 woman with no known medical comorbidities presented with a progressively enlarging mass over the right upper back of the chest. She had no known family history of malignancy or familial adenomatous polyposis and reported no significant smoking or alcohol history. The patient first noticed the mass 30 months ago

during pregnancy. At that stage, it measured around 4 cm and was painless and non-tender. Approximately four months after delivery, she noticed rapid enlargement of the lesion. Continued growth was associated with back discomfort and progressive impairment of her activities of daily living. On clinical examination, a very large mass measuring about 30 × 40 cm [Figure 1A & 1B] observed across the right upper posterior thorax and which was firm, immobile and non-tender. Magnetic Resonance Imaging (MRI) was performed. MRI demonstrated a complex large right thoracolumbar paraspinal mass infiltrating right erector spinae muscles which crossed the midline with penetration of contralateral left erector spinae muscles and there was no obvious extension to the adjacent vertebra and spinal cord [Figure 2]. A core-needle biopsy demonstrated a benign spindle-cell neoplasm. The differential diagnosis included desmoid-type fibromatosis, other fibroblastic or myofibroblastic neoplasms, peripheral nerve-sheath tumors, smooth-muscle neoplasms and solitary fibrous tumor. Definitive diagnosis was established following histological examination of the surgical excision of the mass. The morphological and immunohistochemical findings supported desmoid-type fibromatosis.

The extent of muscular involvement and proximity to the spine indicated that radical resection could result in substantial functional morbidity. Because of the tumor's considerable size, progressive enlargement, associated discomfort and interference with daily activities, surgical excision was performed. During surgical procedure, the tumor was found to be extended into the paraspinal musculature and towards the spine. Complete radical excision would carry a substantial risk of functional impairment. Therefore function-preserving excision was accomplished. A surgical drain was left in situ at the end of the procedure. The resected specimen weighed 5.05 kg [Figure 3] and size measured 40 × 30cm. Histopathology investigation demonstrated an unencapsulated spindle-cell neoplasm involving the dermis and arranged in long, sweeping fascicles. No significant cytological atypia or mitotic figures were identified. Immunohistochemical staining demonstrated patchy smooth-muscle actin positivity, negative muscle-specific actin staining, focal desmin positivity, negative S100 and SOX10 staining, focal CD34 positivity and patchy nuclear β-catenin positivity. The morphological and immunohistochemical findings supported the diagnosis of desmoid-type fibromatosis. The tumor was present at the superior, inferior, medial, and deep margins, but the lateral margin was tumor-free by 2 mm. The diagnosis was desmoid-type fibromatosis, incompletely excised. The postoperative course was satisfactory. The patient was discharged on the same day to the referring district hospital for continued care. She was subsequently reviewed at the surgical outpatient department. The operative wound had healed well, with no wound-related complications documented at follow-up after 3 months [Figure 4A & 4B]. The case was discussed at the institutional multidisciplinary surgical meeting after the histopathological diagnosis. Pursuing further radical surgery to obtain clear margins could lead to significant functional impairment as the tumour extended into the paraspinal muscles and proximity to the spine. Therefore, the team decided to focus on preserving function and opted for ongoing clinical and radiological surveillance.

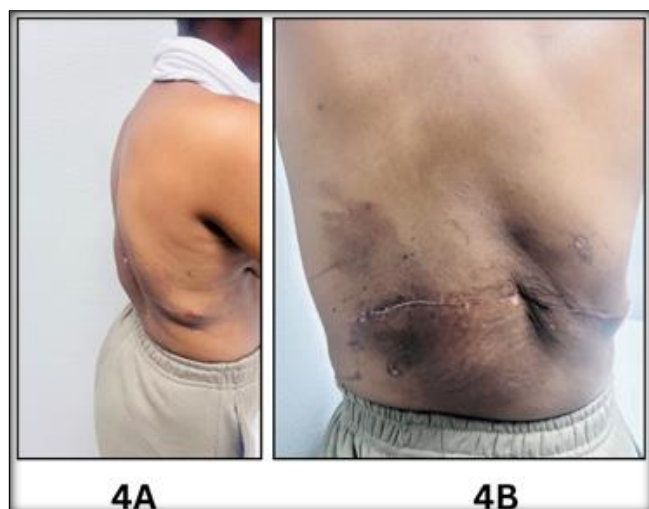


Figure 4: Postoperative scar after 3 months of operation. Lateral view (4A). Posterior view (4B)

DISCUSSION

Desmoid-type fibromatosis presents a distinctive clinical paradox. Histologically, it lacks the features of a high-grade malignant neoplasm and does not metastasize. Clinically, however, it may behave aggressively through infiltration of adjacent muscle, neurovascular structures, viscera or bone. Its natural history is unpredictable, and tumor size alone does not fully determine biological behaviour.^[1,2] Pregnancy-associated desmoid tumors often occur in the abdominal wall, where tissue stretching and injury are factors, and hormonal influences are explored, though the role of estrogen remains complex.^[5,8] Recent data suggest pregnancy may increase disease progression or relapse risk. However, many cases are manageable with active surveillance and often have a favourable course.^[5,8] The case's notable feature was the temporal relationship between pregnancy and tumor development. The patient first noticed the lesion during pregnancy, with rapid postpartum growth. In an international retrospective observational study spanning 20 years, Fiore et al. reported disease progression in 68.7% of patients diagnosed with desmoid fibromatosis during pregnancy.^[9] Among patients diagnosed during pregnancy, spontaneous regression occurred in 3.1% during pregnancy and in 28.6% after pregnancy.^[9] This case is unique, as the lesion arose in the posterior thoracic and paraspinal region, reaching 350 mm.

Various imaging modalities are essential for diagnosis, treatment and follow-up of desmoid tumours. They include ultrasound, CT and MRI. Ultrasound is preferred in pregnant women and may show an oval mass with smooth contours to an ill-defined tumour depending on disease stage. The imaging characteristic features of 'tail sign' (linear extension into fascial planes) and 'staghorn sign' (intramuscular digitiform projections) are suggestive but not exclusive for desmoid tumours.^[3] The advantages of MRI in desmoid-type fibromatosis are delineating tumor extent, muscular and fascial involvement, and relationships to critical structures. It also aids long-term surveillance.^[1,2] In this patient, MRI imaging showed extensive posterior thoracic and paraspinal

involvement near the spine, influencing surgery planning by highlighting potential morbidity from radical clearance.

Histopathological investigation is essential to confirm the diagnosis. Desmoid-type fibromatosis features bland spindle cells in fascicles within collagenous stroma, with no cytological atypia or high mitotic activity.^[1,6] These features were observed in this case. Nuclear β -catenin expression indicates activation of the Wnt/ β -catenin pathway and aids diagnosis when interpreted together with the morphology and other immunohistochemical findings.^[1,6] Lack of S100 and SOX10 expression argues against a nerve-sheath tumor. Focal CD34 positivity alone does not support a solitary fibrous tumor; the morphology and nuclear β -catenin positivity instead support desmoid-type fibromatosis.^[6] Historically, wide surgical excision with negative margins was standard. Now, management is more conservative and tailored. Active surveillance suits many new patients, especially with stable, minimally symptomatic tumors not threatening critical structures.^[1,2] Treatment is needed for progression, pain, functional issues, or threats to vital structures. Options include surgery, systemic therapy, radiotherapy, and ablative procedures, chosen based on tumor site, symptoms, progression, treatment risks, patient preference, and expertise.^[1,2] Local ablative strategies for DFT include Cryotherapy, High-intensity focused ultrasound (HIFU) and Transarterial Chemo-embolisation (TACE).^[10] Cryotherapy is indicated in patients with primary or recurrent extra-abdominal tumours and tumours close to bones. Contraindications include tumours with direct skin involvement, intra-abdominal tumours and close proximity to silicone implants or neurovascular structures. TACE is indicated for primary or recurrent extraabdominal, tumours with suitable vasculature, tumours not amenable to local ablative therapies and tumours near neurovascular structures. The absence of a dominant tumour-feeding artery or presence of collateral arteriogenesis are contraindications to TACE. HIFU is indicated for primary or recurrent intra- and extra-abdominal tumours but contraindicated in tumours close to skin, bone, bowel and neural structures. Radiotherapy is indicated for therapy-resistant tumours.^[10] In this case, the large lesion caused discomfort and impairment. Excision relieved symptoms, but paraspinal involvement risked functional loss with radical resection.

The link between microscopic margins and recurrence in desmoid-type fibromatosis isn't definitive. Some studies show higher recurrence with incomplete resection, while others emphasize tumor biology, location, age, and size as more influential. Pregnancy cohorts report good outcomes after R1 resection.^[8,11] Harati et al (2017) reported recurrence-free survival (RFS) on 90 patients with aggressive fibromatosis. The patients were followed up for a median of 7.7 years.^[12] They found the resection margin at primary tumour was superior in preventing the recurrence than tumour size, width of resection and subsequent R0 resection. No statistical difference between R0 resections of <1mm or >1mm was noticeable. The authors advocated for a more conservative surgical approach that is function and limb-preserving. In this case, wider margins would have meant more extensive removal of paraspinal muscles and possible spinal intervention. Accepting positive margins was a strategic choice, not failure. Long-term follow-up is vital since residual tumors may stay stable, regress, or progress.

Systemic therapies are optional for progressive, symptomatic, or

unresectable disease requiring active treatment.^[1,2] Tyrosine-kinase inhibitors such as sorafenib have demonstrated activity in advanced or refractory desmoid tumors.^[13] Nirogacestat, a γ -secretase inhibitor, improves progression-free survival, tumor response, symptoms, and quality of life in adults with progressing desmoid tumors.^[14] Treatment choices for a young woman must consider toxicity, reproductive goals, access, and clinical urgency. Nirogacestat can cause ovarian dysfunction, which is particularly relevant in a patient who hopes to have more children.^[15] Anti-estrogen treatments such as tamoxifen, once used because of the association between desmoid tumors and pregnancy, are no longer considered first-line systemic therapy.^[1,2] The multidisciplinary team discussed tamoxifen but decided on observation until evidence of progression appears.

CONCLUSION

Giant pregnancy-associated paraspinal desmoid-type fibromatosis is an uncommon and surgically challenging condition. Although the tumor lacks metastatic potential, its infiltrative growth may cause marked morbidity and complicate attempts at complete resection. Function-preserving resection with acceptance of microscopically positive margins may be appropriate. Careful long-term clinical and radiological surveillance is essential.

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Conflicts of interest

There are no conflicts of interest.

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