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CASE REPORT

Pregnancy-Associated Giant Abdominal Desmoid Tumor: A Case Report of Active Surveillance and Surgical Management

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Objective: There is not much data on pregnancy-related desmoplastic fibroma (DF), and the effectiveness of active surveillance as a primary treatment strategy remains uncertain. Thus, our case illustrates that DF tumors can persist into late pregnancy. Termination of pregnancy in conjunction with surgical removal of the tumor may reduce trauma and complications, ultimately resulting in a healthy infant.

Case Presentation: This case describes a pregnant woman with a suspected desmoid fibromatosis tumor who refused termination of pregnancy, opting for close monitoring until late gestation and successfully delivering a healthy baby.

Conclusion: This case reveals the feasibility and potential safety of individualized, multidisciplinary management approaches - such as active surveillance followed by timely surgical intervention - for DF in pregnancy. It highlights the importance of balancing maternal and fetal risks and presents insights that may assist the clinical-decision making process in similarly complex scenarios involving pregnancy-related DF.

Keywords: desmoid tumors, aggressive fibromatosis, targeted therapy, active surveillance, pregnancy

Introduction

Desmoid-type fibromatosis (DF) is a mesenchymal tumor recognized for its local invasiveness but lacking metastasis. Despite the absence of metastasis, these tumors exhibit significant local destruction, often necessitating surgical excision. It is estimated that 2–4 new cases of DF occur annually per million people. Multiple studies suggest that estrogen may play a crucial role in the development of desmoid fibromatosis.[1] The expression levels of estrogen receptor beta have been shown to influence DF proliferation.[2] Stretching trauma to the abdominal wall is closely associated with DF development,[3] and this may explain why desmoid tumors often occur during pregnancy, especially in the later stages. Pregnancy-associated DF is a rare but distinctive condition. The uniqueness of this case lies in its occurrence and treatment during pregnancy, introducing novel perspectives on multimodal therapy to safeguard fetal health.[4,5] Similar cases have been reported cases in the literature, with one case report primarily focusing on the pathologic clinical complexity of a large pelvic desmoid tumor during pregnancy.[4] DF frequently occurs during pregnancy/postpartum, necessitating careful planning for subsequent pregnancies.[2] Other studies have emphasized the risk of DF progression during pregnancy and its potential impact on the health of women.[6] Given the intricacies of DF, comprehensive treatment of cases requires multidisciplinary collaborative efforts from experts.[5] It is an aggressive soft tissue tumor typically originating from fascia, muscles, and deep connective tissues.[7] The etiology of DF remains elusive. DF can occur in any part of the body. Radical surgical resection remains the primary treatment for DF. However, the local recurrence rate within five years post-resection is 20–65%. Therefore, continuous follow-up and appropriate interventions are essential.[8,9]

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Currently, there is scarce literature on pregnancy complicated by DF. Moreover, there is no established clinical guideline particularly addressing the management of DF during pregnancy. Management approaches are typically extrapolated from general DF protocols or based on expert opinion and multidisciplinary consensus. As such, each case needs individualized decision-making that carefully evaluates maternal and fetal risks. Herein, we present a case report of concurrent pregnancy and DF, aiming to make a contribution to the limited literature and present insight into potential management approaches.

Case Presentation

A 35-year-old female patient underwent a cesarean section at 35 weeks of gestation to terminate the pregnancy. The newborn weighed 2460 g with Apgar scores of 8 at 1 minute (deducting 1 point each for breathing and skin color), nine at 5 minutes (deducting 1 point for breathing), and nine at 10 minutes (deducting 1 point for breathing). The newborn was transferred to the intensive care unit for observation. Meanwhile, the patient underwent the removal of a massive right pelvic abdominal mass (Figures 1 and 2). Intraoperative findings revealed a mass located on the right pelvic abdominal wall, measuring approximately 18×15×12 cm. It extended into the pelvic cavity, tightly adhering to the uterus. The upper edge reached approximately 3 cm above the umbilicus, and the lower edge extended to the iliacus muscle. The

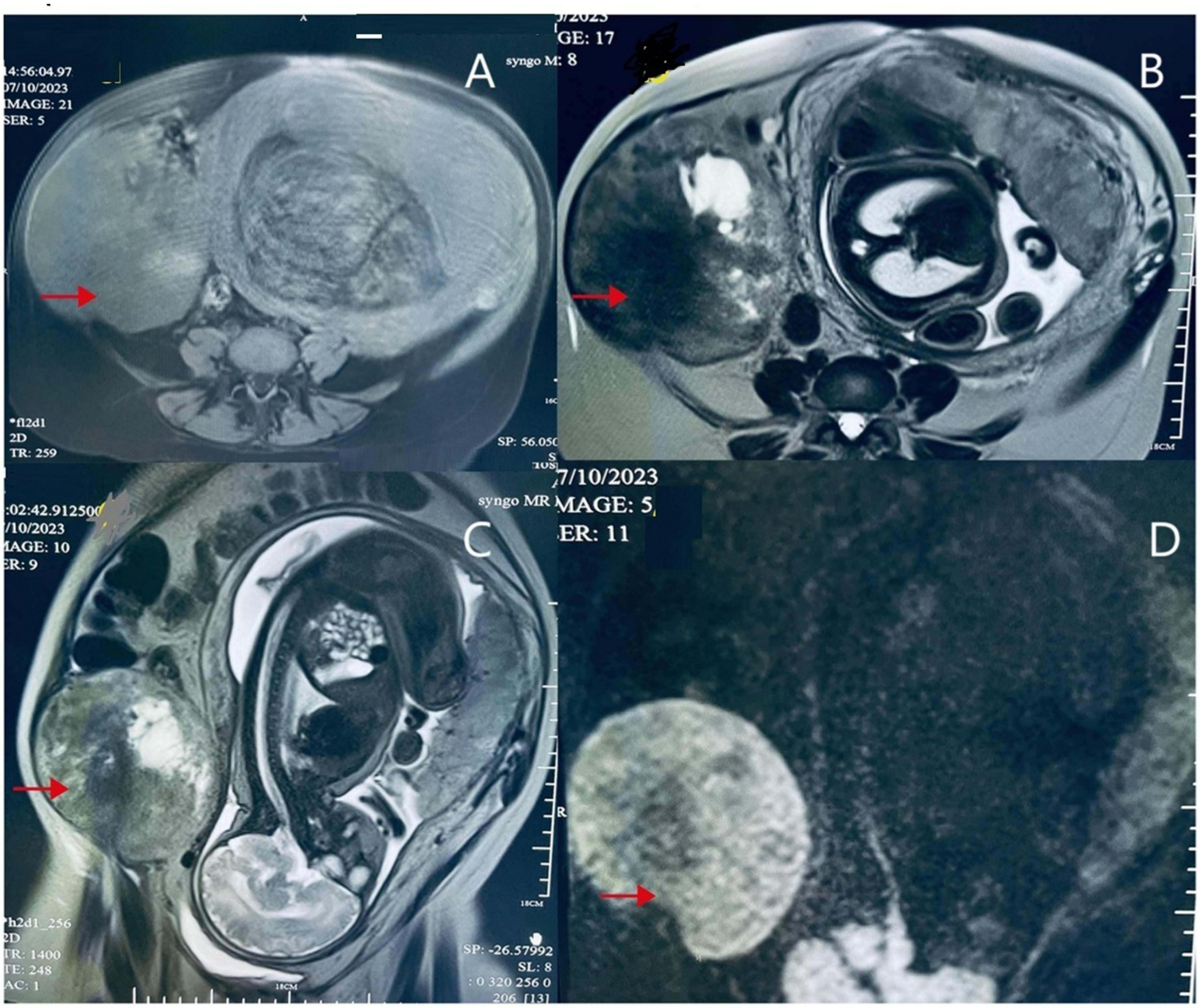

Figure 1 Horizontal magnetic resonance image containing (**A**–**D**). An abnormal-signal mass (red arrows) is seen adjacent to the right lower abdominal wall and extending into the abdominopelvic cavity with well-defined borders, peritoneal pushing inward, and mild compression and displacement of the uterus and right ovary.

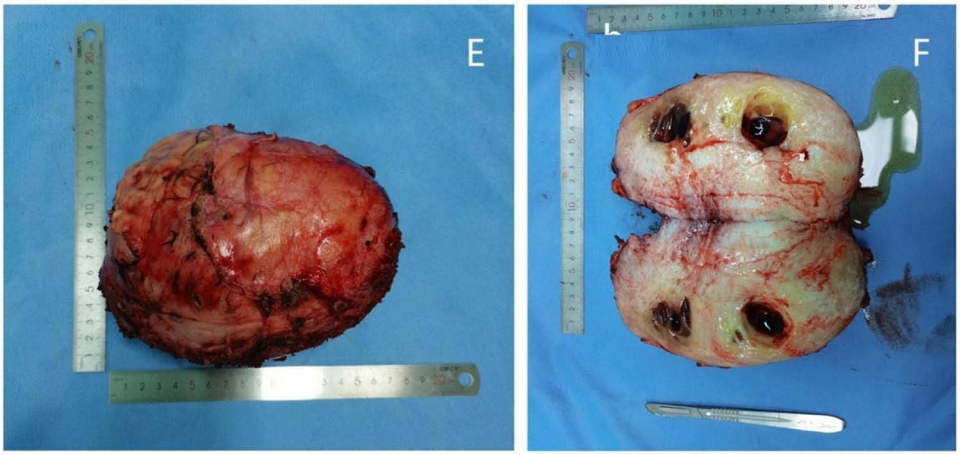

Figure 2 Images E and F show the actual size of tumors removed by surgery.

mass was closely connected to the right iliac bone, displaying a smooth surface with numerous small blood vessels and restricted mobility. The mass was surgically removed, confirming its origin from the right iliac muscle. A pregnant woman in her second month reported worsening right lower abdominal pain and a palpable mass. Pelvic ultrasound showed an intrauterine pregnancy alongside a solid mass in the right lower abdomen, measuring approximately 6.4×5.2×6.4 cm, characterized by unclear borders and close association with muscle layers. The mass showed limited movement with respiration, and color Doppler flow imaging indicated internal blood flow. Magnetic resonance imaging confirmed a tumor near the right rectus abdominis, connected to surrounding tissues. The biopsy revealed a spindle cell tumor, suggesting DTF. The tumor progressively grew, leading to increased pain exacerbated by movement and changes in position. Despite a prior history of proper tubal removal due to an ectopic pregnancy and a cesarean section three years ago, the patient opted to carry the current pregnancy. Due to the rapid growth of the estrogen-sensitive tumor during pregnancy, recommendations included terminating the pregnancy, performing tumor removal, and exploring potential systemic treatment options. However, the patient chose to continue the pregnancy and underwent regular follow-up and imaging assessments. At 30 weeks of pregnancy, the tumor had grown significantly, and postoperative pathology confirmed an invasive fibromatosis with ischemic areas and cystic margins. The tumor measured approximately 17*13*9.5 cm with positive β-catenin and Ki67 (<5%).

Postoperative pathology revealed a spindle cell tumor in the abdominal wall (Figure 3). By combining morphology and immunohistochemistry, the diagnosis aligned with invasive fibromatosis (DTF) with focal ischemic infarction and cystic borders. The tumor dimensions were approximately 17*13*9.5 cm. Immunohistochemistry showed positive β-catenin and Ki67 (<5%), while estrogen receptor, progesterone receptor, desmin, S-100, CD34, and Signal transducer and activator of transcription-6 (STAT-6) were negative. The patient was discharged one-week post-surgery and is under continuous follow-up.

Discussion

The present study reports a case of a multiparous woman with a history of cesarean section and laparoscopic surgery. During an 8-month pregnancy, she underwent a cesarean section, delivering a healthy baby. The tumor was completely removed during the procedure. The etiology of DF remains vague, possibly related to surgery, trauma, or hormonal factors. Approximately 8–18% are associated with pregnancy,[10] potentially attributed to stretching abdominal muscles and fascia during gestation. However, this causative connection fails to elucidate the occurrence of fibromatosis in different locations during and after pregnancy. Hormonal changes and immune variations during pregnancy may also play significant roles.[5,11]

DF accounts for approximately 3% of all adult soft tissue tumors, with an annual incidence rate of 2–5 individuals per million.[12] In female patients, common tumor types such as leiomyomas, desmoid tumors, ovarian cysts, and ovarian

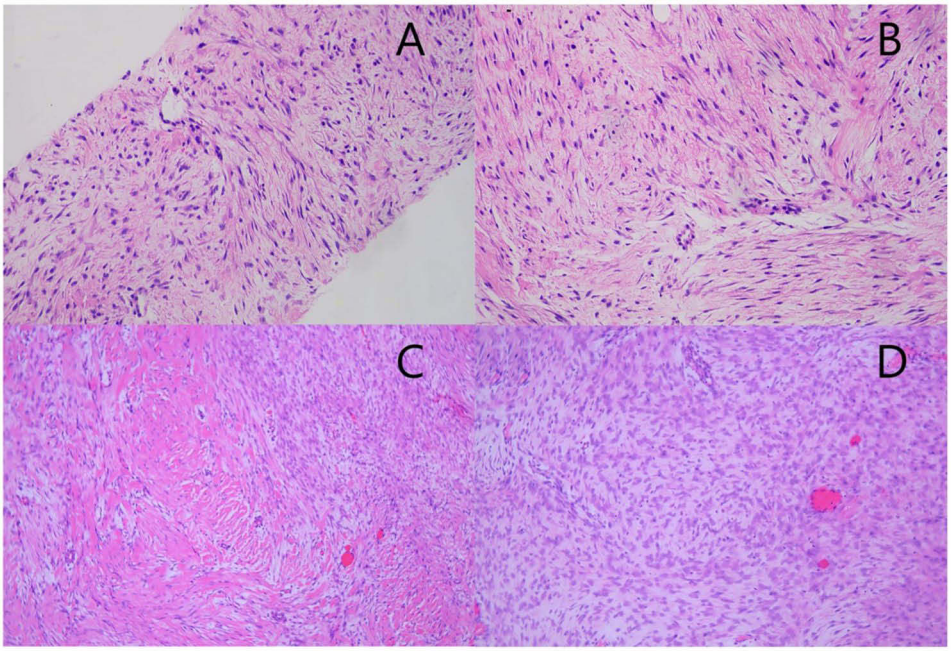

Figure 3 Part (**A** and **B**) show that the cells are spindle-shaped in morphology and sparsely arranged. Part (**C** and **D**) show that the nuclei are short spindle-shaped or ovoid, without nuclear division, and the cytoplasm is light eosinophilic red.

malignancies pose a diagnostic challenge as ultrasound, computed tomography, or magnetic resonance imaging lacks specificity in DF presentation, undoubtedly increasing clinical misconception and misdiagnosis rates.[13]

Currently, there are no established recommendations for the timing and methods of treating DF during pregnancy. Since 2005, a conservative active monitoring approach has been the primary strategy for managing DF during pregnancy.[14] A multicenter retrospective analysis demonstrated the safety of regular monitoring, with most patients not requiring treatment.[5] In situations necessitating tumor removal during pregnancy, a multidisciplinary approach involving obstetricians, neonatologists, and surgical teams is crucial. Surgical strategies should carefully consider the extent of lesion removal and its potential impact on the fetus, ensuring optimal outcomes for both the mother and the newborn.

While open resection has been opted for in our case owing to the tumor's size, location, and pregnancy-associated anatomical variations, it is critical to understand the emerging role of laparoscopic surgery in dealing with abdominal wall DF. Recently, Buzzaccarini et al have revealed that laparoscopic resection can be a safe and effective choice, indicating benefits including intraoperative blood loss, shorter hospital stays, quicker recovery, and reduced post-operative pain.[15] Nonetheless, limitations include technical challenges when tumors are large, deeply infiltrative, or closely linked with crucial structures-as was the case here. Post-operative concerns of laparoscopic DF resection, though infrequent, may include port-site hernias, seromas, or injury to surrounding tissues. While laparoscopy may not always be useful during pregnancy due to uterine size and different anatomy, it is a valuable strategy for selected postpartum cases or early-stage tumors. Additional studies are necessary to determine patient selection criteria and long-term findings following laparoscopic intervention in DF.

There is a need to balance fetal maturity and the need for tumor removal before considering the termination of a 35-week pregnancy to minimize the risks for both the mother and the child. Abdominal wall DF surgery is associated with low complications and recurrence rates, even in cases with positive margins (R1), making surgery a viable option.[9] The woman felt that the pressure and pain in the abdomen disappeared after the operation and did not feel uncomfortable after changing positions or walking. The case of this woman demonstrates that tumors during pregnancy exhibit faster growth and greater size due to hormone levels. In addition, the non-malignant tumor tended to infiltrate growth and local recurrence. This case underscores the significance of personalized treatment tailored to the specific needs of pregnant

women. Adequate assessment of the pregnant woman and the fetus is carried out to develop an individualized surgical plan, looking at the size and location of the tumor, the relationship of the tumor to the surrounding tissues during pregnancy, the relationship of the tumor to the pregnant uterus, and whether the tumor is affecting the fetus' growth and development. It is crucial to balance the possibility of tumor recurrence with the health of the mother and the fetus. Despite successful surgery, DF has a high recurrence rate and, therefore, requires long-term follow-up. Long-term surveillance following pregnancy-related DF is crucial because of the high occurrence rate of the tumor, even after complete surgical excision. Current post-treatment approaches mainly include periodic clinical assessments and imaging-such as MRI or ultrasound-every 3 to 6 months for the first two years, then annually if no recurrence is noted. Nonetheless, standardized surveillance protocols are still undefined, especially in postpartum patients. In addition, gaps exist in our knowledge of DF behavior affected by hormonal shifts in pregnancy and the postpartum period. This case highlights the requirement for prospective studies and consensus-driven guidelines to inform optimal timing of intervention, safe monitoring threshold, and long-term follow-up practices tailored to reproductive-age women. Ongoing surveillance and timely intervention can improve the overall prognosis.

Conclusions

Pregnancy-associated DF is a non-malignant tumor of muscle and connective tissues that occurs during pregnancy. It is often associated with hormonal changes and uterine expansion. Symptoms of DF include abdominal pain, palpable masses, compressive symptoms (such as increased urination and constipation), and restricted mobility. The treatment approach is contingent on the severity of the condition, the pregnant patient's overall health, and the fetus's safety. Observation and monitoring are frequently employed as treatment methods. However, in cases of severe symptoms or when there is a threat to the fetus, considerations may involve surgical intervention, radiation therapy, or other interventional measures.

Ethics Approval and Consent to Participate

The reported women signed informed consent forms to participate this study. In addition, the ethical number of our study was 2023A105, which was simultaneously discussed and approved by the Ethics Committee of Obstetrics and Gynecology of Hangzhou Women's Hospital, Hangzhou, China. This study was performed according to the principles of the Declaration of Helsinki.

Consent to Publish

The reported women signed informed consent forms to publish this study. The Ethics Committee of Obstetrics and Gynecology of Hangzhou Women's Hospital, Hangzhou, China discussed and approved this study (Approval No. 2023A105).

Author Contributions

All authors made a significant contribution to the work reported, whether that is in the conception, study design, execution, acquisition of data, analysis and interpretation, or in all these areas; took part in drafting, revising or critically reviewing the article; gave final approval of the version to be published; have agreed on the journal to which the article has been submitted; and agree to be accountable for all aspects of the work.

Disclosure

The authors report no conflicts of interest in this work.

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