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Abdominal Fibromyxoid Sarcoma in Pregnancy: An Unusual Cause of Preterm Labor and Sepsis

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Abstract

Cancer in pregnancy is rare, with incidence less than 1%, and the most common cancers being melanoma, breast, and cervical cancers. Fibromyxoid sarcoma is a soft tissue tumor involving deep soft tissues of the extremities and trunk, rarely located in the abdomen. A low-grade fibromyxoid sarcoma (LGFMS) falls in the family of fibrosarcoma. Only two cases of LGFMS in pregnancy have been reported. We report a case of abdominal LGFMS in pregnancy leading to preterm labor, sepsis, and an acute abdomen requiring surgery in the third trimester. A 19-year-old woman, gravida 1 at 32 weeks and 5 days presented to an outside hospital with preterm contractions and cervical effacement. She had a known abdominal mass, suspected to be accessory liver lobe, measuring 9.0 × 6.4 × 7.7 cm in the right upper quadrant. At 33 weeks of gestation, she developed fever and hypotension. Magnetic resonance imaging confirmed the presence of the mass, which was now on the left side of the abdomen and associated with a suspected abscess. She underwent cesarean delivery, and complete surgical resection of the mass along with a small bowel resection. Final pathology of the mass revealed a LGFMS. This case also highlights the need for a multidisciplinary approach to manage a rare presentation of sepsis and preterm labor in pregnancy.

Keywords
- sarcoma
- pregnancy
- sepsis
- preterm labor

Cancer during pregnancy is rare, with an estimated incidence of 0.07 to 0.1%.[1–3] An increasing incidence has been reported in several studies, likely related to advanced maternal age.[1,4]

The most common cancer types during pregnancy are malignant melanoma, breast, and cervical cancer.[1] Bone and soft tissue sarcomas have been reported in pregnancy with osteosarcoma, liposarcoma, and Ewing's sarcoma being the most likely histological types, comprising over 50% of the cases.[2] Fibromyxoid sarcoma in pregnancy has been reported in two studies: one in the left atrium and ventricle and the other in the gluteus.[5,6]

A fibromyxoid sarcoma is a soft tissue tumor involving deep soft tissues of the extremities and trunk, rarely located in the abdomen, and often presenting as a painless mass. A low-grade fibromyxoid sarcoma (LGFMS) falls in the family of fibrosarcoma. First described in 1987, it is histologically benign appearing but with a known high metastatic potential.[7,8] They are commonly found in young or middle-aged men.[8–11] The incidence is estimated at 0.18 per million, accounting for 0.6% of all soft tissue sarcomas.[12] However, the true incidence is unknown, as the World Health Organization characterizes it as an "uncommon tumor."[12]

We report a case of a symptomatic abdominal fibromyxoid sarcoma in the third trimester of pregnancy, presenting initially with signs and symptoms of preterm labor (PTL), and then evolving into an acute abdomen and sepsis.

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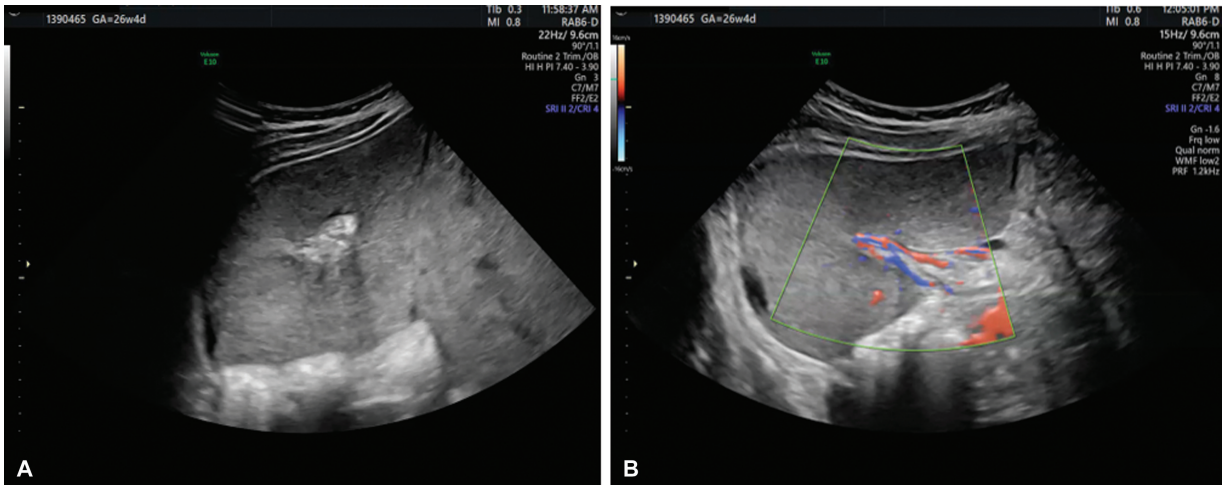

Fig. 1 Outpatient ultrasound at 26 weeks' gestation showing a solid mass, (**A**) approximately 10 cm, suspected to be in the right adnexa and (**B**) with a central vascular hyperechoic area seen with vessels tracing into mesentery.

Case

A 19 year-old woman, gravida 1 at 32 weeks' and 5 days' gestation presented to an outside hospital with regular contractions and cervical effacement. For concerns for PTL, she was given betamethasone for fetal lung maturity, nifedipine for tocolysis, and intravenous fluids. She was then transferred to our tertiary care hospital for further care due to fetal prematurity. Upon arrival, her cervical exam was unchanged and her contractions had partially improved.

Her past medical history was remarkable for a known right adnexal mass measuring $9.0 \times 6.4 \times 7.7$ cm, documented on her 26-week ultrasound (►**Fig. 1**). At that time, the etiology was unknown. The mass was described as solid with homogenous echotexture and prominent vasculature, adjacent to the liver, suspicious for being an accessory liver lobe. A magnetic resonance imaging (MRI) was recommended to further elucidate the characteristics and location of the mass. However, imaging was not performed before her presentation for PTL.

On hospital day 3 (33 weeks' and 1-day gestation), the patient reported worsening abdominal pain, now in her left flank. A repeat ultrasound was immediately performed at bedside, considering the new location of her pain. The mass that was previously described in the patient's right adnexa was now demonstrated in the left adnexa, with stable size and characteristics. This new finding was suspicious for a torsion of the mass, most likely resulting from the increased pressure of the growing uterus (►**Fig. 2**). An MRI was performed to better characterize the mass (►**Fig. 3**). Findings were consistent with ultrasound, reporting a large, soft tissue, vascular mass now visualized in the left side of the abdomen. There was also a suspicion for an abscess collection adjacent to the mass.

Her condition worsened as she developed a new onset of fever with a maximum temperature of 103°F (39.4°C), tachycardia to the 120s, and hypotension (blood pressure in the 90s/50s). She had severe lower abdominal tenderness on the left lower abdominal quadrant with guarding. Findings were suggestive of an acute abdomen and sepsis, and the

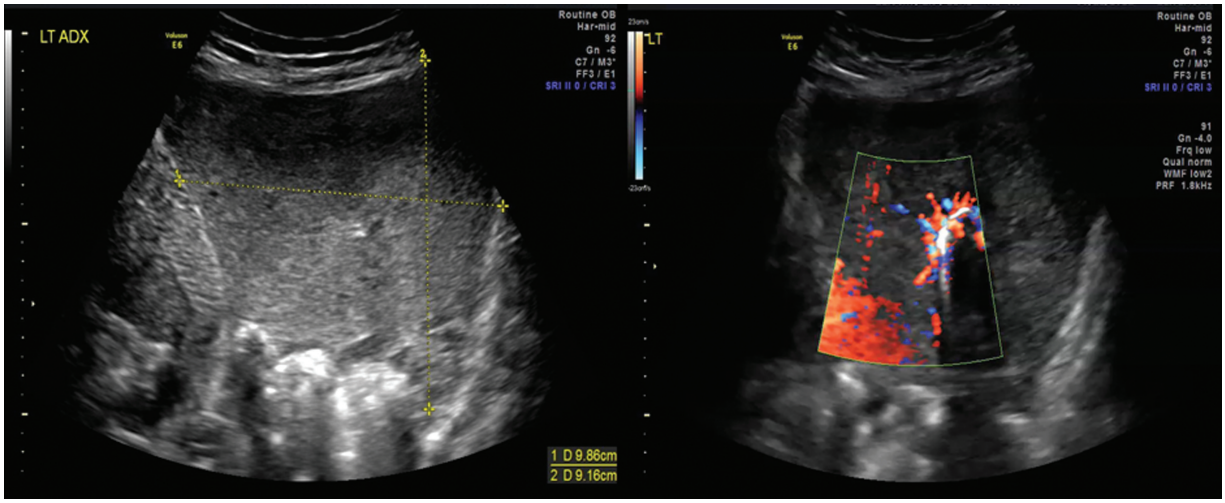

Fig. 2 Inpatient ultrasound showing a previously identified right adnexal mass now on the left, with stable size and characteristics, including prominent vascularity.

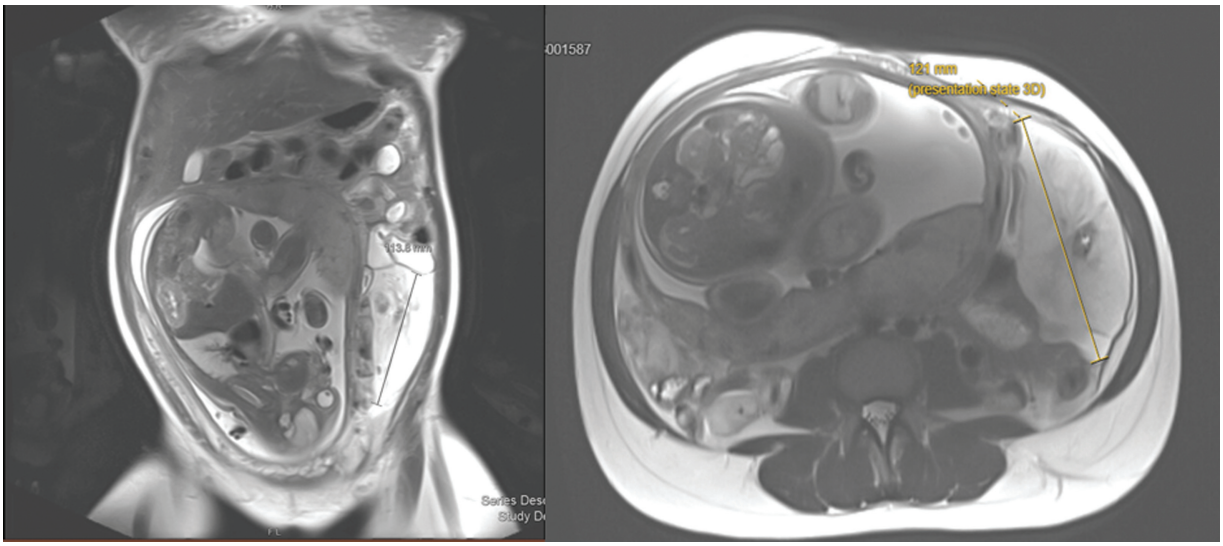

Fig. 3 Inpatient magnetic resonance imaging (MRI) without contrast showing left-sided 12-cm abdominal mass.

patient was taken to the operating room along with general surgery and started on broad-spectrum antibiotics due to concerns of intraabdominal infection with an unknown source.

A cesarean was followed by an exploratory laparotomy, via a low midline vertical skin incision and low transverse hysterotomy. During the cesarean the patient had significant uterine atony requiring several uterotonics, followed by blood transfusion of 2 units. Purulent peritoneal fluid from the left lower quadrant was sent for cytology. The mass was continuous with the peritoneum and resected along with a portion of the small bowel due to adhesions (►**Figs. 4** and **5**). A small bowel side-to-side anastomosis was completed.

The patient was discharged on postoperative day 7—afebrile, tolerating regular diet, and with adequate pain control. The intraoperative fluid sample grew pan-sensitive *Klebsiella pneumoniae* for which she completed the antibiotics course. The infant did well and was discharged home on day 11 of life.

Final pathology of the mass revealed a low-grade fibromyxoid spindle cell neoplasm. Pathology reported the tumor to be composed of spindle cells arranged in short fascicles in a fibromyxoid matrix. The cells had pale eosinophilic cytoplasm and oval nuclei with coarse speckled chromatin (►**Fig. 6**). The mitotic rate was less than 1 mitosis per 10 high-power fields. Proximal, distal, and radial margins appeared clear of neoplasm, but the tumor extended to the visceral peritoneum.

Patient was referred to the oncology service and is currently under surveillance every 6 months with imaging, to assess for risk of recurrence. To date, patient has been stable with no reported recurrence.

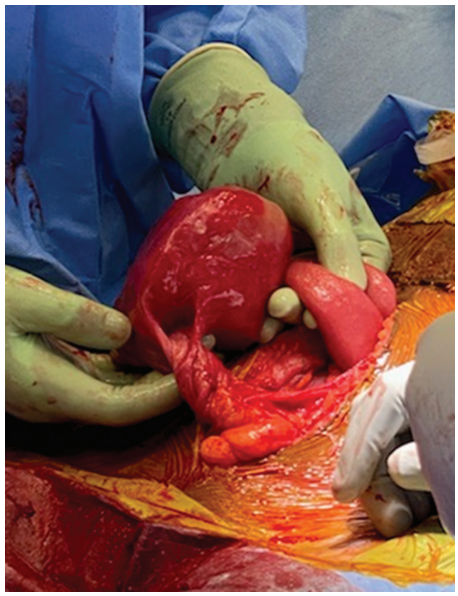

Fig. 4 Intraoperative assessment of the mass, seen adhered to the small bowel.

Discussion

Fibromyxoid sarcomas are soft tissue tumors involving deep soft tissues rarely found in the abdomen as described in the nonpregnant population.[13] They affect patients of all ages, with peak incidence in young adults, with an equal male-to-female ratio.[13,14] In a large series of pregnancies complicated by bone and soft tissue tumors, the most common affected locations were the pelvis, abdomen, and extremities.[2] Very few case reports have been published about intra-abdominal fibromyxoid sarcoma.[9,10,15]

LGFMS has a histologically benign appearance; however, it is a malignant and late metastasizing tumor.[13] Rates of recurrence are difficult to assess due to low incidence of these tumors and difficult follow-up with long intervals from primary diagnosis to recurrence.[7] In a series of 33 LGFMS cases, patients with negative margins at diagnosis had better survival.[7] Smaller tumors (<3.5 cm) were associated with decreased recurrence. Local recurrence was demonstrated in 63% of the cases and metastasis was reported in 46% of cases with site of metastasis in lung, pleura, and chest wall.

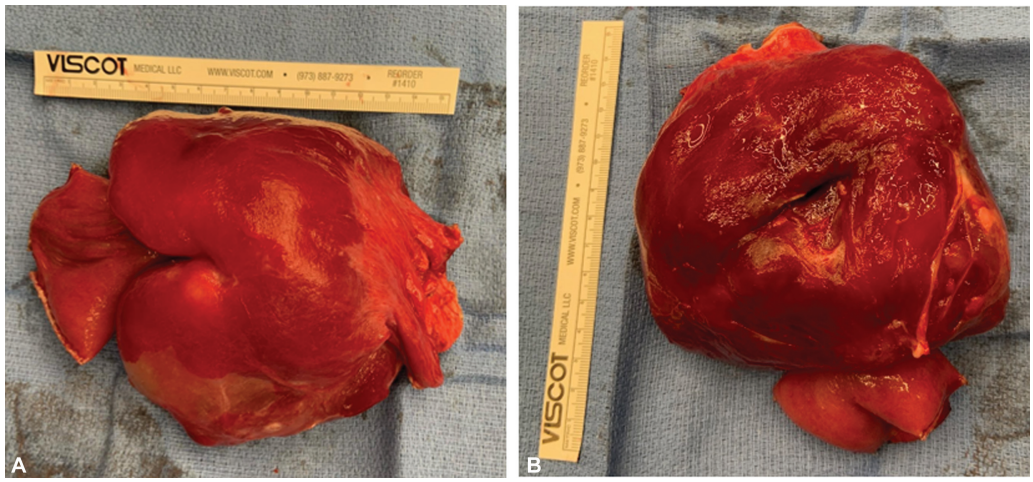

Fig. 5 The intraoperative mass was resected with a portion of small bowel attached (**A** anterior and **B** posterior, showing orifice draining pus).

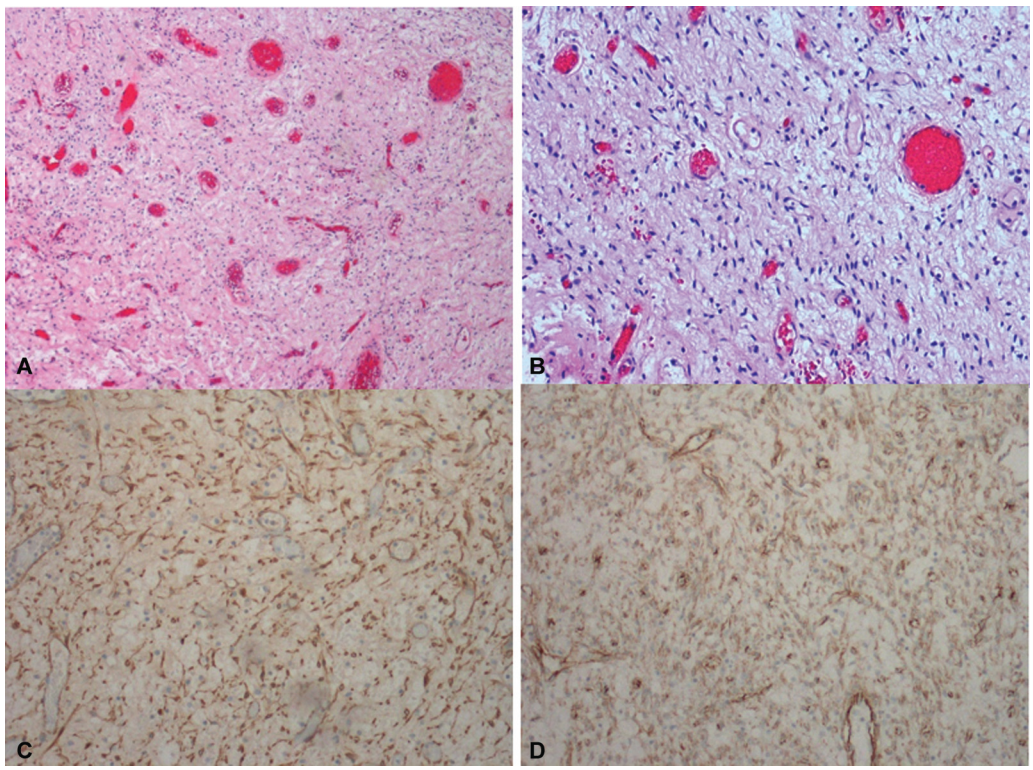

Fig. 6 Hematoxylin and eosin stained at 40× (**A**) and 100× (**B**), smooth muscle actin immunostain (**C**), and β catenin immunostain (**D**) showing the characteristics of the low-grade fibromyxoid spindle cell neoplasm.

The management of pregnancies complicated by cancer remains challenging due to late diagnosis in most of the cases resulting from overlap between pregnancy physiological changes and cancer symptoms as well as delayed treatment due to potential maternal and fetal risks.[1,2]

The best treatment approach of sarcomas in pregnancy remains debatable. Overall, pregnant women are candidates for standard surgical management in pregnancy.[2] The decision for chemotherapy or radiation should be evaluated on an individual basis.[2] Multidisciplinary approach between obstetricians, oncologists, and neonatologists is essential to improve outcomes.[2] Due to its low nuclear grade and infrequent mitotic activity, LGFMSs are not sensitive to chemotherapy or radiation.[13] Surgical excision with clear resection margins remains the first-line treatment option.[13,14]

In the largest systematic review of the literature currently available on 40 pregnancies complicated by sarcomas (including uterine, vaginal, vulvar, and retroperitoneal), the mean age of patients was 27.8 ± 7.0, onset of symptoms was described in the third trimester in 50% of the cases, 42% did not have initial suspicion for malignancy, and 82% had live-born infants with premature deliveries in almost 50% of the cases.[3] In all cases, the primary tumor was treated with excision, five cases used neoadjuvant radiation, and in one case, chemotherapy was used.[7] Our case is the first one in

the literature to report an abdominal fibromyxoid sarcoma in pregnancy. As evidenced by intraoperative finding, the sarcoma was thought to have torsed to include a part of the small bowel causing an intra-abdominal infection leading to sepsis and PTL. Additionally, a common gut flora, *Klebsiella pneumonia*, was cultured from the intraabdominal fluid collection, which is indicative of a possible bowel perforation.[16] This case not only demonstrates that our patient's characteristics and outcomes are similar to what has been previously reported[3] but also highlights the importance of keeping a broad differential when evaluating PTL, including causes of acute abdomen and sepsis as well as understanding how the anatomical changes in pregnancy need to be taken into consideration when making diagnosis of a surgical abdomen such as in cases of appendicitis or this specific case. This case also highlights the need for a multidisciplinary approach to safely evaluate and manage a rare presentation of sepsis and PTL in pregnancy.

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Conflict of Interest
None declared.

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