A Rapidly Growing Abdominal Mass: Desmoid Tumor in Pregnancy

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Abstract

Background Desmoid tumors are benign soft tissue tumors that locally invade adjacent tissue. There is a paucity of reports describing the rapid growth of these tumors during pregnancy.

Case A giant desmoid tumor arising from the left abdominal wall of a young female patient with rapid growth during pregnancy is described. Preoperative evaluation included ultrasonography and magnetic resonance imaging. Decision made by a multidisciplinary team was not to intervene before birth, and abdominal delivery at term was accomplished.

Conclusion Desmoid tumors should be part of the differential diagnosis in an abdominal wall tumor of rapid growth during pregnancy. Future studies are needed for better understanding of the pathogenesis, diagnosis, and treatment of desmoid tumors in pregnant women.

Keywords
- desmoid tumor
- pregnancy
- rapid growth
- abdominal wall

Desmoid tumors (or desmoid fibromatosis) are rare, soft tissue monoclonal neoplasms that arise from mesenchymal stem cells. No association with metastatic disease has been reported, however, they often show a strong tendency to infiltrate local adjacent tissue causing significant morbidity and mortality.[1] Sporadic desmoid tumors (those not associated with the familial adenomatous polyposis coli [FAP] syndrome) have an estimated incidence of two to five cases per million per year.[2,3] Pregnancy-related desmoid tumors are even more infrequent, with limited published literature available.[2] The etiology of this condition is currently unknown, however it has been shown that the majority of spontaneous desmoid tumors show activating mutations of the beta catenin gene,[1,4,5] as well as somatic mutations of the adenomatous polyposis coli gene.[6] They have also been associated with hyperestrogenic states and trauma, but the evidence is largely based on retrospective and anecdotal case reports.[2] It seems that the hormonal and immunological changes occurring in pregnancy may play a role in the severity and course of the disease, however, their true association with the pathogenesis of the disease remains unclear.

The relevance of the present case lies in the rapid growth of a small tumor turning into a colossal mass toward the end of pregnancy.

Case

A 24-year-old woman presented to our department for prenatal care at 14 weeks of gestational age (GA). Her obstetrical history included a first trimester spontaneous abortion and two full term pregnancies resulting in spontaneous vaginal deliveries. She had a 2-year history of a hyperdense lesion documented by computed tomography scan that had undergone minimal interval growth within the anterior left abdominal wall (►**Fig. 1**). No other relevant surgical or medical history was noted. Physical examination at 23 weeks of GA revealed a mass protruding from the left abdominal

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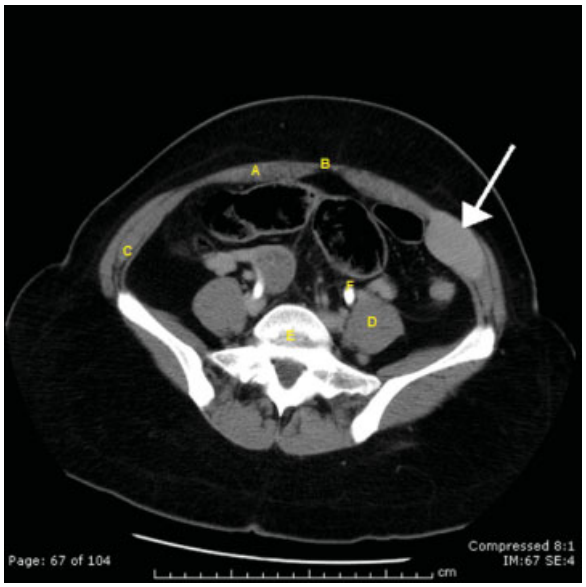

Fig. 1 (A) Rectus abdominis muscle. (B) Linea alba. (C) Right internal abdominal oblique/transversus abdominis muscle. (D) Left psoas muscle. (E) Spine. (F) Left ureter. Arrow: left abdominal wall mass.

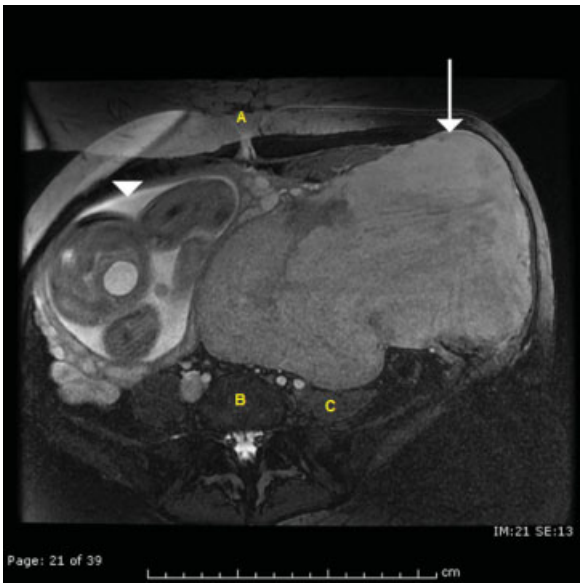

Fig. 3 (A) Linea alba. (B) Spine. (C) Left psoas muscle. Arrow: Left abdominal wall mass. Tip of arrow: Gravid uterus at 28 weeks of gestational age.

wall. Ultrasonography revealed a single intrauterine pregnancy, as well as a large 11.7 × 12.3 × 17 cm left lower quadrant mass located above the bladder and below the gravid uterus. Considering the characteristics of the mass, the first diagnostic presumption was that of a uterine fibroid. A follow-up ultrasound at 28 weeks demonstrated interval growth of the mass to 16.5 × 11.8 × 18.5 cm (►**Fig. 2**). Magnetic resonance imaging (MRI) for better characterization showed a 19.5 × 10.8 cm transverse × 13.3 cm craniocaudal mass with heterogeneously T2 hyperintense and T1 iso- to slightly hyperintense mass centered within the left abdominal wall musculature (►**Fig. 3**). The mass displaced the gravid uterus and compressed the left gonadal vessels, left ureter, and left psoas muscle without a clear fat plane between the mass and these structures. It also abutted several loops of small bowel in the left abdomen and extended into the peritoneal cavity. Mild left hydronephrosis and hydroureter were also found proximal to the ureteral compression. No enlarged lymph nodes were found. Given patient's age, gender, and tumor location, a large abdominal wall desmoid tumor was suspected. A multidisciplinary evaluation including general surgery resolved not to intervene until after delivery. This decision was driven by the vast vascularization of the tumor and the need for further testing. At 38 weeks and 6 days of GA a slightly tender mass was palpated on physical examination (►**Fig. 4**). Follow-up ultrasonography reported a mass of 18 × 13 × 18.9 cm and a fetus in complete breech presentation.

A primary low transverse cesarean section via right paramedian vertical incision was performed. Intraoperative findings included a large, white-gray, hypervascular mass located in the left lower quadrant measuring approximately 25 cm in

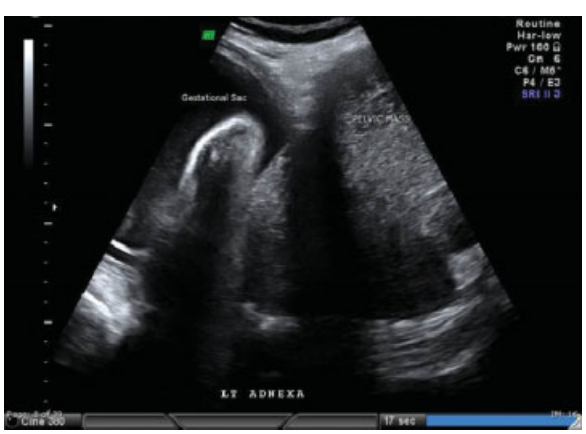

Fig. 2 Ultrasound image at 28 weeks of gestational age showing the gestational sac and the pelvic mass.

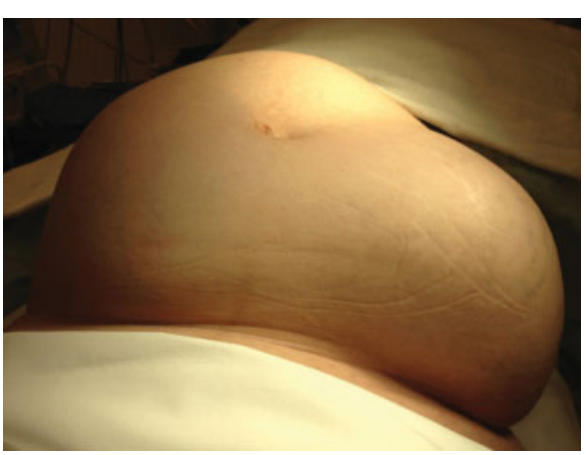

Fig. 4 Abdomen at 38 weeks and 6 days of gestational age.

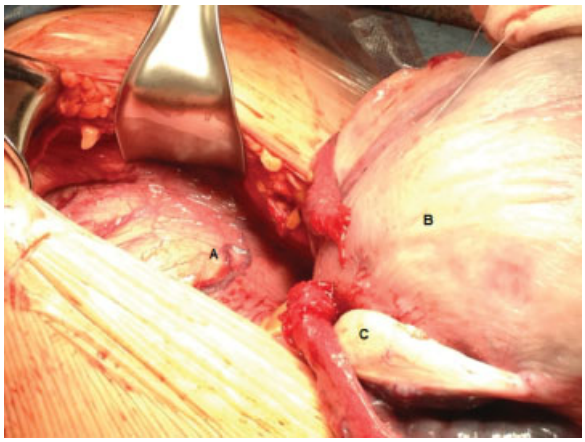

Fig. 5 Intraoperative findings: Hypervascular mass compressing adjacent organs. (A) Hypervascular mass. (B) Uterus. (C) Right ovary.

its largest diameter, and arising from the anterior abdominal wall (►**Fig. 5**). The uterus was displaced to the right side. The left uterine sidewall, adnexa, and vasculature were all located in the midline of the abdomen. Following the delivery of a live male newborn (birth weight: 3,350 g; frank breech position; Apgar score 9 at 5 minutes) intraoperative evaluation by general surgery was performed and decision was made not to intervene. Patient tolerated the surgical procedure well, and the postoperative course was uneventful. At the monthly follow-up, the patient presented with a new palpable non-mobile mass of 3 × 4 cm at the superior border of the incision that was tender to palpation. This was not present at the time of surgery and subcutaneous hematoma or seroma were ruled out. Unfortunately, the patient's lack of adherence to medical recommendation was a serious obstacle for the definite treatment, and she has yet to undergo surgery for complete resection of the tumor.

Comment

The aggressive growth of a desmoid tumor during pregnancy is a rare occurrence.[2] Overall, they constitute 3% of soft tissue tumors and 0.3% of all neoplasms.[7] Even though considered rare, this condition has been substantially investigated with variable results. No significant gender difference has been recorded in the FAP population; however, there is evidence that estrogen may play a role in the pathogenesis and clinical course of sporadic tumors. Indeed, an incidence of estrogen receptor positivity up to 33%, with equal sex distribution has been reported.[3] Spontaneous regression in menopause or after bilateral oophorectomy has been previously described, as well as in vitro studies that reveal that hormone-related proliferation of fibroblasts increases during pregnancy and progressively decreases thereafter.[2] This evidence supports the effect of estrogen on desmoid pathogenesis; however, larger studies are needed to assess this association.

There is good evidence that β-catenin mutations and dysregulation may in fact be associated with the development of sporadic desmoid tumors. Animal model testing has granted more weight to this hypothesis by recognizing increased proliferation, motility, and invasiveness in transgenic mice with the targeted β-catenin mutation when compared with a control group.[1] Recent data suggest that genomic factors such as identifying the exact mutation in β catenin (CTNNB1) within a given desmoid, helps stratifying patient risk and prognosis.[4] A limitation of our report is that the tumor was not assessed for the presence of estrogen receptors and no studies aiming β-catenin mutations were held. Anti-estrogen binding sites and estrogen receptor studies are important in this particular case since an accelerated growth of the mass was evidenced during the entire course of pregnancy. Other risk factors such as trauma (laparoscopic port sites and previous cesarean delivery scars) and cytogenetic abnormalities have also been associated with the development of desmoid tumors; however, their true effect is questionable.

Ultrasonography remains the most frequently used imaging modality for the initial evaluation of an intra-abdominal mass suggestive of a desmoid tumor.[6] In the presence of similar findings, it is important to incorporate these uncommon lesions in the differential diagnosis given the variability of sonographic characteristics. In fact, they can easily be misinterpreted as uterine leiomyomata like in this case. When evaluating pregnant women, MRI is the best next step to lay out the borders of the tumor and its relationship to adjacent organs and structures. Moreover, if resection of the mass is considered, MRI is helpful for surgical mapping. Some authors have proposed MRI to be used not only as a diagnostic tool, but also for prognostic purposes. They believe MRI scanning using T2-weighted imaging may correlate with the behavior of the tumor, with a bright signal indicating high water content, collagen, and fibrous tissue that associates with rapid growth.[1,2] The final diagnosis is made by core or open biopsy of the mass, since clinical characteristics and imaging are not enough to truly differentiate desmoid from other soft tissue tumors.

Surgical treatment remains the gold standard; however, there is convincing evidence that local recurrence may be as much as 25% in people with sporadic desmoid fibromatosis,[8] and much more for families with FAP. When counseling the patient for recurrence, it is important to bring attention to the fact that a subsequent pregnancy is not necessarily a risk factor for recurrence of the disease.[8]

Different clinical treatments have been widely used including radiation therapy, nonsteroidal anti-inflammatory drugs (sulindac), hormonal therapy (tamoxifen and toremifene), and cytotoxic chemotherapy all with variable outcomes.[1] For asymptomatic desmoids tumors, close observation might be an acceptable strategy,[4,5] but that is not the case when there is evidence of adjacent organ damage like in our patient. Case reports with successful surgical treatment during and after pregnancy have both been described in medical literature,[9–14] but controversy is still present. The size and rapid growth of these tumors make them prone to complicate the outcomes of pregnancy, hence the importance of reports and reviews on the topic.

In conclusion, desmoid tumors during pregnancy represent a true challenge from the point of view of both, diagnosis

and treatment. Future studies are needed to reveal the true pathogenesis of the disease and its natural clinical course. Treatment should be individualized and appropriate patient counseling should be undertaken. The timing of surgical treatment has not been clearly defined and further research is required to answer this clinical question. Our case report highlights the importance of considering desmoid tumors in the differential diagnosis when evaluating abdominal wall soft tissue tumors of rapid growth during pregnancy.

Conflict of Interest
None.

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