Management of A Desmoid Tumor in Pregnancy: A Case Report

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Introduction

The desmoid tumor, also known as aggressive fibromatosis or musculoaponeurotic fibromatosis, is a rare, non-metastasizing, locally aggressive, monoclonal proliferative disease of fibroblastic origin with a high recurrence rate. The clinical course of this neoplasm is unpredictable, making management challenging [1,2]. The etiology of desmoid tumors has yet to be fully clarified. However, certain factors have already been linked to their development: an injury or muscle strain with a disproportional inflammatory response, an estrogen-related endocrine factor, or even genetic factors [3]. The vast majority of these tumors develop sporadically; however, in 5-15% of cases they are associated with familial adenomatous polyposis (FAP) [4,5]. Desmoid tumors in cases of FAP occur in individuals with deletion of the alleles of the adenomatous polyposis coli (APC) tumor suppression gene (5q21-
Desmoid tumors can be found on the abdominal wall, in the abdominal cavity, trunk and limbs [7]. Sporadic desmoid tumors most commonly originate in the region of the abdominal wall and are found principally in the rectus abdominis muscle in young women of reproductive age, those who have suffered trauma, particularly postpartum, or in pregnant women [3]. Conversely, tumors associated with FAP are often situated intra-abdominally [7].

The histology of biopsy specimens shows highly differentiated fibroblasts with no mitotic activity. Immunohistochemistry shows positive staining for beta-catenin, actin and vimentin and negative staining for cytokeratin and S-100 [1].

Complete resection with a wide safety margin used to be considered the first-line treatment. However, this approach has been questioned due to the high rate of local recurrence, particularly when excision is performed exclusively, and in view of the unpredictable clinical course of the disease, with spontaneous regression occurring in up to 29% of cases [2,8].

In advanced tumors, the use of radiotherapy, estrogen-receptor antagonists, non-steroidal anti-inflammatory drugs (NSAIDs) and systemic chemotherapy has been evaluated [9]. The detection of estrogen receptors, in association with pregnancy and the use of oral contraceptives, supports the use of anti-estrogens such as tamoxifen for the treatment of aggressive fibromatosis; however, response is slow [1].

The objective of this paper was to describe a case of a large desmoid tumor on the abdominal wall that developed in the 10th week of pregnancy in a woman with no history of FAP. Conservative management was adopted and the patient was monitored postpartum, with no intervention.

The internal review board of the Santa Casa de Misericórdia Hospital in Vitória, Espírito Santo, Brazil approved the publication of this paper under reference CAAE 39043320.0.0000.5065. The patient signed an informed consent form giving her permission to publish this case report.

**Case report**

A 29-year old pregnant white woman (G3P2A1), who had delivered by Cesarean section 18 months previously, arrived at the maternity hospital at 40 weeks and 6 days of pregnancy complaining of pelvic pain that resembled contractions. Prenatal ultrasound images showed a tumor in the anterior portion of the uterus, which had been identified as a uterine fibroid, measuring 5.22 cm in diameter in the first trimester of pregnancy (Figure 1) and 13.5 cm in the third trimester (Figure 2). At hospital admission, the patient was found to be in good general health, alert and well-oriented, with fundal height of 41 cm, fetal heartbeat of 140, regular uterine contractions, and 3 cm of dilatation. Due to her history of short intervals between pregnancies and considering her wish regarding mode of delivery, the decision was made to immediately proceed with a Cesarean section.

![Figure 1: First-trimester ultrasound: a nodular image of 5.22 cm, identified as a uterine fibroid.](image1)

![Figure 2: Third-trimester ultrasound: a tumor anterior to the fetus, measuring 13.5 cm x 6.71 cm.](image2)
moved to one side (Figure 3). No uterine fibroids were found.

**Figure 3:** Tumor in the rectus abdominis muscle pushed aside to enable access to the abdominal cavity.

At surgery, it was decided to proceed with conservative management; therefore, the mass was not removed and the layers were closed.

The patient progressed well following surgery and was discharged from hospital on the second day following delivery. Ultrasound-guided core biopsy was performed four and a half months after delivery, with histology showing the presence of fibrous proliferation with areas of sparse spindle cells resembling fibroblasts immersed in a matrix that consisted predominantly of collagen. The conclusion was reached that this was a benign spindle cell neoplasm, with possible musculoaponeurotic fibromatosis (Figure 4). Immunohistochemistry confirmed diagnosis of a desmoid tumor (Table 1).

**Figure 4:** Fibrous proliferation with areas of sparse spindle cells resembling fibroblasts immersed in a collagen matrix.

| Table 1: Immunohistochemistry of the specimen. |
|-----------------------------------------------|
| **Target antigens** | **Antigen expression** |
|---------------------|------------------------|
| Protein S           | Negative               |
| Beta-catenin        | Positive               |
| Smooth muscle actin | Positive in the vessels|
| Calretinin          | Negative               |
| Desmin              | Negative               |
| CD34                | Positive in the vessels|
| MUC-4               | Negative               |
| Estrogen receptor   | Negative               |
| Progesterone receptor| Negative              |

Very importantly, the patient had no family history or personal history of FAP. Colonoscopy was performed and results confirmed that there was no such association, with the procedure failing to detect FAP-associated polyposis. At a follow-up visit six months after delivery, the patient was asymptomatic. At physical examination, her abdomen was flaccid, superficial and deep palpation was painless, and no masses or visceromegaly were found. At follow-up ultrasound performed at that same time, the tumor was found to have reduced by 29.5% in length and by 40.5% in thickness. Nine months following delivery the patient continues to be monitored under expectant management, with no complaints or symptoms.

**Discussion**

Desmoid tumors of the abdominal wall are commonly associated with pregnancy and appear to be related to trauma and strained rectus abdominis muscle. In addition, hormonal changes such as increased estrogen and progesterone levels and an increase in circulating growth factors may be involved [10,11].

Although the progression rate of desmoid tumors in pregnancy is high, prognosis is generally good [10]. In the case reported here, ultrasound images revealed an increase of around 158% in the size of the lesion over a period of approximately four months during pregnancy. Speranzini et al. [3] studied 14 patients and found only one case in which pregnancy was not involved. In 30-59% of cases, the desmoid tumor appeared during or following a second pregnancy, increasing in size particularly in the final trimester of pregnancy, as occurred in the patient reported here [12]. In this reported case, the findings suggest that female sexual hormones, particularly estrogens, did not play an important role in the growth of this type of fibromatosis, since immunohistochemistry was negative for hormone receptors. Nevertheless, the distension of the abdominal wall and aponeurosis that developed as pregnancy progressed, as well as the trauma caused by the previous Cesarean section, could have played a role in the appearance and growth of the tumor.

Ultrasonography continues to be the imaging system most commonly used in the initial evaluation of an intra-abdominal mass suggestive of a desmoid tumor [13]. Nevertheless, since this is a rare disease, interpretation can be confusing, as in the case reported here in which the first hypothesis was of a uterine fibroid, with the site on the abdominal wall only being identified at surgery.
Different clinical treatments have been widely used, including radiotherapy, non-steroidal anti-inflammatory drugs, hormone therapy and cytotoxic chemotherapy, with varying outcomes in all cases [2]. For a long time, the standard treatment was radical excision of the tumor, with a wide surgical margin. However, the current trend is to opt for conservative management in the initial approach to the disease [14]. The wait-and-see policy has been the conduct of choice for asymptomatic patients with desmoid tumors that are not invading or compressing structures or for patients with only minimal symptoms. Some authors have argued that the growth of the desmoid tumor is self-limiting and that simply observing the lesion is sufficient, with surgery contraindicated in most cases [15].

Other studies have found that up to 50% of tumors progress unremarkably and in those patients in whom the tumor remains stable for more than a year there would be no need for an active management approach [14]. Major en bloc surgery is no longer considered crucial, since recurrence rates following surgery have exceeded 60% in large series, while spontaneous regression had been documented in around 25% of cases [16]. Therefore, the current trend is towards more conservative treatment, as evaluated in different prospective studies [16]. As also seen with the patient described here, conservative management with no intervention proved effective. The patient remained asymptomatic and the lesion decreased significantly following childbirth.

**Conclusion**

A desmoid tumor is a rare disease; however, its prevalence is high in women of reproductive age and during pregnancy. Attending physicians need to be alert to the possibility of such cases to ensure that appropriate treatment is implemented and to minimize the risk of the patient being exposed to radical and even iatrogenic treatment approaches. Evaluating the possibility of conservative management can represent a major challenge to surgeons who see surgical intervention as a means of resolving the case. However, knowledge on the history of the disease and how it behaves, particularly following childbirth, is of the utmost importance in avoiding unnecessary interventions and morbidity. Therefore, it is reasonable to conclude that conservative management with outpatient follow-up, as adopted in the case reported here, can result in benefit to the patient and a satisfactory outcome insofar as quality of life is concerned, since visceral complications and the cosmetic and functional sequelae of extensive surgery would be avoided.

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