Mesenteric Fibromatosis in a Postpartum Patient: A Case Report

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Abstract

Background: Desmoid tumors are benign and rare fibroblastic neoplasms which behave in a locally-aggressive manner to infiltrate into the adjacent tissue and have a high incidence of local recurrence after surgical excision. Desmoid tumors may develop during or shortly after pregnancy.

Case presentation: We report here a case of abdominal desmoid tumor involving the ileum and its mesentery in a 22-year-old woman after pregnancy. Magnetic resonance imaging (MRI) revealed a well-bordered, well-encapsulated mass with 7x8 cm dimensions on the right side of the abdomen neighboring intestinal loops and the mesentery. The tumor was excised radically along with a part of the ileum with 2 cm tumor-free margins. Histological examination revealed desmoid-type fibromatosis with no immunoreactivity for estrogen and progesterone receptors. The patient had an uneventful postoperative course and was discharged on the 10th postoperative day.

Conclusion: We conclude that, intra-abdominal masses other than fetus should raise suspicion of desmoid tumor which requires timely-intervention during obstetric follow-up.

Keywords: Desmoid Tumor, Mesenteric Fibromatosis, Pregnancy

Introduction

Intra-abdominal desmoid tumors, also known as mesenteric fibromatosis, often are associated with familial adenomatous polyposis (FAP) mutation in a syndrome known as Gardner’s Syndrome. Although sporadic cases of desmoid tumors do occur, unlike Gardner’s syndrome, they predominantly occur extra-abdominally. Although the exact etiology is unknown, several factors are associated with increased risk, the most significant being the FAP mutation. Other risk factors include preceding trauma or surgery, and excess estrogen exposure [1].

Case Presentation

We report here a case of abdominal desmoid tumor involving the ileum and its mesentery in a 22-year-old woman after pregnancy. The patient was referred to our department two months after delivery for investigation of a fist-sized, firm tumor in the ileum and its mesentery which was first noticed six weeks after delivery. A firm, solid, intraabdominal mass with smooth contours adjacent to anterior abdominal wall was detected with ultrasonography. Computed tomography (CT) of the abdomen revealed a solid, encapsulated, well-bordered mass with 7x7 cm size and heterogeneous contrast pattern between inferior renal pole and vena cava inferior (VCI) which compressed the VCI and pushed it medially. Magnetic resonance imaging (MRI) revealed a well-bordered, well-encapsulated mass with 7x8 cm dimensions on the right side of the abdomen neighboring intestinal loops and the mesentery. The mass had muscle intensity on T1A-weighted series and was more hyperintense than paravertebral muscles on T2A-weighted series; signal intensity was greater on its exterior than that on its interior (Figure 1). Gastroduodenoscopy was normal. No other pathology, including polyps, was detected with total colonoscopy. The tumor was excised radically along with a part of the ileum with 2 cm tumor-free margins (Figure 2). Size of the resected specimen was 10×11.5×11.5 cm. Histological examination revealed desmoid-type fibromatosis with no immunoreactivity for estrogen and progesterone receptors (Figure 3). The patient had an uneventful postoperative course and was discharged on the 10th postoperative day. The patient was symptom-free with regard to this tumor during pregnancy.

Figure 1: Abdominal magnetic resonance imaging (MRI) appearance of the well-bordered and well-encapsulated intra-abdominal mass with 7x8 cm dimensions
Discussion

Desmoid tumors are benign and rare fibroblastic neoplasms which behave in a locally-aggressive manner to infiltrate into the adjacent tissue and have a high incidence of local recurrence after surgical excision [2,3]. Desmoid tumors can occur in almost every part of the body and are generally classified as abdominal and extra-abdominal. Abdominal desmoids are the predominant form in FAP-associated cases but most of the sporadic desmoids are seen in extra-abdominal region or on the abdominal wall. Patients with FAP have a 1000-fold increased risk of developing desmoid tumors compared to the general population, and 2% of all cases of desmoid tumors are associated with FAP [5]. Approximately 10% to 15% of the sporadic desmoid tumors are located in the abdominal cavity. Intra-abdominal desmoid tumors, also known as mesenteric fibromatosis, are a rare form of soft tissue tumor. Other risk factors include preceding trauma or surgery, and excess estrogen exposure [1,6,7].

Desmoid tumors may develop during or shortly after pregnancy. The predominance of cases affecting young women during or after pregnancy is particularly apparent. These findings raise two questions: is it the trauma of pregnancy that induces desmoid growth and does the hormonal influence itself ameliorate disease expression [4,8,9,10,11]? Clark and Phillips reported an increase in the risk for desmoid tumors with the current use of oral contraceptives, as well as a reduction of risk after menopause, pinpointing the role of sexual hormones in the pathogenesis of desmoid tumors [10].

Estrogen treatment may induce, both in animals and in humans, the formation of desmoid tumors that regress after cessation of drug administration or after addition of progesterone[12].

Tamoxifen, a drug with anti-estrogen activity, has been shown to be partially responsible for anti-desmoid effect [4,13].

Common presentations for abdominal desmoids include abdominal pain, nausea, vomiting, diarrhea, hematochezia, small bowel obstruction, and hydronephrosis [1]. On gross examination, desmoid tumors are firm, poorly circumscribed masses which often infiltrate adjacent structures. Histologically, they are comprised of bundles of spindle cells in a collagenous stroma. The lesion is generally of low cellularity, and fibroblasts generally concentrate on the lesion’s periphery. Histologically, mature fibroblasts of uniform size and shape are observed, with mitosis being unusual [4].

Treatment of advanced aggressive fibromatosis remains to be challenging. Although benign and rarely metastatic, desmoid tumors carry a mortality rate of up to 10% due to their aggressive, locally-invasive nature. Surgical intervention for intra-abdominal desmoids is generally considered to be the treatment of choice and is curative in many cases. One possible exception to this rule is the subset of cases involving the superior mesenteric vessels. In such cases, the complex nature of the surgery carries increased surgical risks, leading some authors to advocate the use of non-surgical alternatives, including radiation as well as various pharmacotherapies such as high-dose tamoxifen and sulindac [1]. Surgical resection of these tumors has been performed successfully both during pregnancy and soon after delivery, and the role of postpartum radiotherapy, chemotherapy and other medical intervention remains controversial [5,7]. However, in some cases, an aggressive combination of chemotherapy is needed to obtain a fast response [10].

Jones and Harvey proposed that radical surgery should not be performed in patients with FAP and mesenteric desmoid tumors except for symptomatic tumors [8,14]. However, despite the risks, there remains a role for surgery in the management of desmoid tumors. In case reports by other authors, surgery was beneficial for patients with desmoid tumors who required life-saving surgery as a result of failure of nonsurgical treatments or the development of complications [1,9].

In conclusion, mesenteric fibromatosis is a rare disease which may occur in a higher rate in pregnancy. Therefore, intra-abdominal masses other than fetus should raise suspicion of desmoid tumor which requires timely-intervention during obstetric follow-up.
References


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