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CASE REPORT

GIANT DESMOID TUMOR LOCALIZED ON THE ABDOMINAL WALL: A CASE REPORT

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ABSTRACT

Desmoid tumors develop from musculoaponeurotic tissues. They are locally aggressive, but they cannot metastasize. In this case report, we evaluated a patient with a desmoid tumor and the approach to the tumor. A 28-year-old female patient presented with an abdominal mass. She underwent a partial nephrectomy operation on the same side as the mass. Computed tomography revealed a 65x70 mm solid mass on the left abdominal wall. The desmoid tumor was diagnosed by a tru-cut biopsy, the patient was operated on, and the mass was removed. No complications were observed.

Desmoid tumors can be classified as extra-abdominal, abdominal, and intra-abdominal according to their location and may cause different symptoms according to their localization. Treatment options should be determined according to the increase in the size of the mass and the symptoms in the patient.

Keywords: Abdominal wall, desmoid, fibromatosis, rectus abdominis

INTRODUCTION

Desmoid tumors are benign but locally aggressive mesenchymal tumors that develop from musculoaponeurotic tissues, usually arising from the abdominal wall muscles but also frequently seen in the extremities and abdominal mesentery (1, 2). They are also known as aggressive fibromatosis, deep fibromatosis, musculoaponeurotic fibromatosis, and desmoid fibromatosis. These tumors have a solid, smooth, and mobile structure. They are usually adherent to the surrounding tissues and do not affect the skin on them (3, 4).

Those located intra-abdominally may typically remain asymptomatic until they cause signs of visceral organ compression (2). Such tumors are rarely seen in the literature, and the treatment approach for giant desmoid tumors exceeding 15 cm is controversial. Although their etiology is not exactly known, they have been associated with connective tissue development disorders, familial adenomatous polyposis (FAP) (also known as Gardner's syndrome), abdominal surgery and trauma, pregnancy, and estrogen therapy (3, 4). Although they do not have metastatic potential, desmoid tumors have a high tendency to recur, leading to significant morbidity and mortality (3). In this study, we report a rare case of a desmoid tumor of more than 15 cm located in the abdominal wall.

CASE REPORT

A 28-year-old woman, who was diagnosed with livedoid vasculitis five years ago and had a history of left kidney partial nephrectomy operation due to kidney stones, was consulted by general surgery with the complaint of a palpable hard mass in the upper quadrant of the left abdomen that started about one month ago when she came to the dermatology service for a follow-up visit.

The physical examination of the patient, whose biochemistry analysis and blood values were normal, revealed no other pathological findings except for a painless, palpable, and hard mass on the left anterior abdominal wall. A contrast-enhanced computed tomography (CT) scan of the patient revealed a solid mass of 65x70 mm in size, distorting the fascia into the intra-

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abdominal area on the left anterior abdominal wall, lateral to the rectus muscle and subcutaneous fascia at the level of the iliac wing, caudal to the umbilicus, and increasing heterogeneity around it (Figure 1). However, no finding in favor of metastasis was detected. Four months later, a follow-up magnetic resonance imaging (MRI) showed a mass lesion with T1A hypointense and T2A hyperintense signal properties at the level of the umbilicus on the left side with a size of 115x86 mm (Figure 2). To have detailed information about the cytological properties of the mass, an ultrasonography (USG)-guided tru-cut biopsy was performed in the interventional radiology clinic, and the sample was sent to the pathology laboratory for examination. In the macroscopy sample, four off-white-colored biopsy materials measuring 0.1-1.4 cm were detected. In sample microscopy, the tissues observed in the examined sections were completely tumoral.

The pathology report confirmed that the tissue did not originate from a previous surgical incision scar.

Spindle cells without atypia were arranged as large fascicles or large swirls. Mitosis, atypical mitosis, and necrosis were not observed, and stromal thin collagen structuring was observed in some places. The immunohistochemical markers applied detected a Ki-67 index of 1-2%, suggesting a slow tumor progression. Additionally, CD34 negativity ruled out inflammatory myofibroblastic abdominal tumors, while s100 and SOX10 negativity indicated that malignant melanoma could be excluded from the diagnosis.

Histopathologic examination of the mass, which was mostly well-circumscribed, was evaluated as desmoid fibromatosis.

Considering the findings, surgical resection of the mass was planned. In the laparotomy performed from the level of the left paramedian incision under the umbilicus, the skin and subcutaneous tissues were passed through and the mass invading the fascia from the left lower quadrant (Figure 3), measuring 19x13x11 cm, localized to the anterior abdominal wall, was completely excised and sent to the laboratory for pathological examination (Figure 4). The defect was repaired with a mesh patch.

The operation was completed by placing two Hemovac drains in the excision lodge. The patient had no postoperative complications and was discharged on the third day with a long-term follow-up plan. Histopathological examination reported

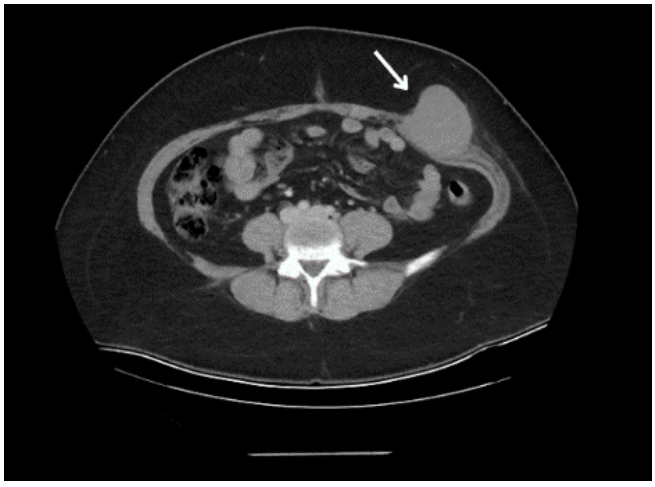

Figure 1: Contrast-enhanced computed tomography of the mass on admission (arrow).

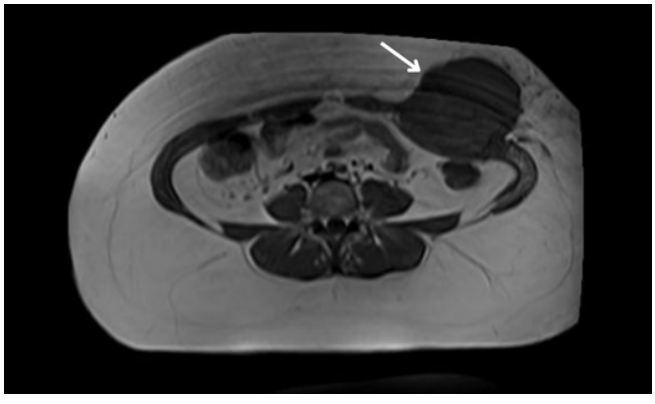

Figure 2: Follow-up magnetic resonance imaging of the mass (arrow).

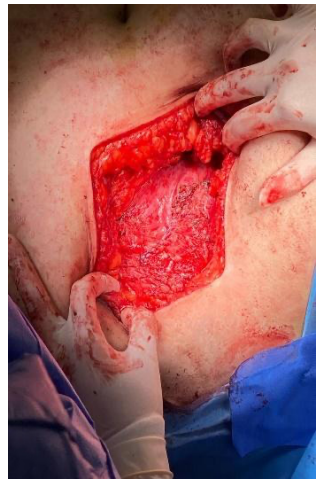

Figure 3: Laparotomy performed during the operation.

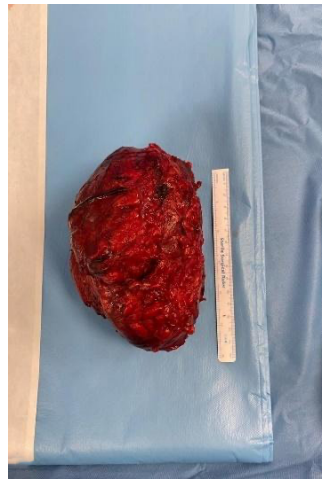

Figure 4: Excised tumoral mass measuring 19x13x11 cm.

that the mass was highly compatible with desmoid fibromatosis, and no recurrence was found in the three-month follow-up of the patient (Figure 5).

DISCUSSION

Desmoid tumors are clonal fibroblastic proliferations arising in deep soft tissues and characterized by infiltrative growth and a tendency for local recurrence but not metastasis (4). Although not malignant, they may cause morbidity and mortality by local invasion (5). It was first described by McFarlane in 1832 and named "desmoid tumor" by Mueller in 1838 (6). They constitute 3% of soft tissue tumors and 0.03% of all tumors. The annual incidence in the general population is 2-4/1000000 (7). It usually occurs in the age range of 20-44 years and is more common in women than in men (8). Our patient was a 28-year-old woman. Desmoid tumors have a high tendency to occur especially in surgical scar areas after trauma, cesarean sections, and previous surgery (3, 9). It is also closely associated with FAP and Gardner syndromes (4, 7). The incidence of desmoid tumors ranging from 1.6% to 17.2% after colectomy operation performed in the treatment of FAP and the fact that desmoid tumors constitute 12% of the causes of death related to FAP indicate the risk and importance of the development of desmoid tumors in patients with FAP (10). Estrogen has also been suggested to be one of the etiological factors in the development of desmoid tumors. Despite evidence indicating estrogen receptor expression in desmoid tumors, an elevated risk of desmoid tumor during and after pregnancy, and higher rates of desmoid tumor development in women of childbearing age and women using estrogen-containing oral contraceptives, the role of hormonal effect is not fully understood due to the limited studies in the literature (9). Our patient had a history of a surgical operation on the same side as the tumor, and since our patient had a diagnosis of livedoid vasculitis, a systemic disease, screening tests for the presence of an accompanying syndromic disease were planned.

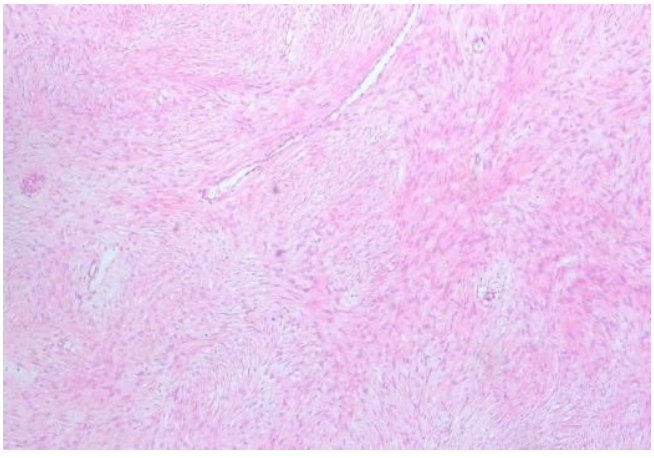

Figure 5: Tumoral lesion consisting of spindle cells without atypia (hematoxylin and eosin at x100 magnification).

Desmoid tumors can occur anywhere in the fascia but are particularly common in the muscle (11). The World Health Organization classification recognizes three different subtypes of desmoid tumors: extra-abdominal, abdominal, and intra-abdominal tumors. The first two of these occur mostly sporadically, whereas the last one has an association with FAP (8). The most common subtype is the abdominal type, with an incidence of 50% (11).

Ultrasonography, CT, and MRI play key roles in the diagnosis, follow-up, and evaluation of recurrence. On USG, it is observed as an infiltrative solid mass with variable echogenicity (12). On CT, myxoid parts of the tumor are seen as a hypodense mass compared to muscle tissue, while collagen and fibrotic parts are seen as an isodense or hyperdense mass compared to muscle tissue. On MRI, it is hypointense or isointense compared to muscles on T1-weighted images and hyperintense on T2-weighted images (3, 13). Imaging is also very crucial in the evaluation of the responses to non-surgical treatment options. Especially after systemic treatment, a decrease in CT intensity, a decrease in T2 signal intensity, and a decrease in tumor contrast enhancement are considered signs of response to treatment, even if no reduction in tumor size is observed (14). In our patient, the location of the mass and its relationship with the surrounding tissues were evaluated by CT and MRI. A tru-cut biopsy performed with USG provided information about the histopathological structure of the mass.

Surgical operations were traditionally preferred in the treatment of desmoid tumors. However, considering the general complications of surgery (such as infection, hemorrhage, and thrombosis) and the local recurrence rate, active surveillance (CT or MRI every 3-6 months), which is a more conservative approach, has started to be preferred (15). A "wait and see" approach may be preferred in newly diagnosed and asymptomatic patients. Surgical mass resection should be preferred as a treatment option in patients who refuse observation, have increased mass size, or are symptomatic, and care should be taken to perform resection with negative surgical margins in case of recurrence (1, 15).

Nowadays, cryoablation, which is a minimally invasive method in the treatment of progressive desmoid tumors, has a lower morbidity rate compared to surgery. In elderly patients with small and medium-sized tumors, the probability of controlling the disease with cryoablation, is higher. It has been reported that the risk of local recurrence is higher in patients with tumor size >5 cm and in young patients (16). We did not prefer this treatment option in our patient because the tumor size was 11.5 cm on imaging.

Radiotherapy should also be considered as a treatment option for patients who are not suitable for surgery or if recurrence is observed (15). In systemic treatment, hormonal therapy, non-steroidal anti-inflammatory drugs, chemotherapy, and targeted therapy can be used to ensure

disease stability (1, 17). In our patient, we performed surgical total mass resection and fascia repair procedures due to the rapid increase in the size of the mass on intermittent imaging and the symptomatic state of the mass in the patient. The high recurrence rate of 35% after surgery has made the management of recurrent patients an important research topic (18). The role of surgery in the treatment of recurrent desmoid tumors is controversial. In the studies analyzed, recurrence was observed in 34% after surgery for recurrent desmoid tumors (19). In non-symptomatic and non-progressive desmoid tumors, spontaneous regression and stabilization can be achieved in 50-88% with active follow-up (20). Syndromic diseases, which are etiological risk factors, should be kept in mind in cases of recurrence.

To conclude, desmoid tumors should be kept in mind in abdominal palpable masses, especially in female patients in the fertile period. Long-term follow-up of patients are recommended, considering the possibility of local recurrence.

Footnote

Ethics Committee Approval: *N/A*

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