Case Report

Desmoid Tumor in the Rectus Abdominis: A Case Report

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ABSTRACT

Desmoid tumors are relatively rare, locally invasive mesenchymal tumors classified as benign or malignant. Patients with desmoid tumors in the abdominal wall that progress during active surveillance may be considered for surgical resection with adequate margins to prevent recurrence. Following radical resection of abdominal wall desmoids, the resulting defect leads to abdominal wall closure difficulties or increased risk of herniation. Herein, we report a case of a desmoid tumor within the rectus abdominis muscle that was treated radically by resection and subsequent reconstruction of the abdominal wall with a synthetic mesh.

Key words : abdominal wall, desmoid tumor, reconstruction, rectus abdominis, tumor resection

Introduction

Desmoid tumors are relatively rare; although they do not metastasize distantly, they are locally invasive, have a high recurrence rate, and can be challenging to treat. For this reason, they are classified as “intermediate, locally aggressive” neoplasms according to the World Health Organization classification of soft tissue tumors¹⁾. Desmoids account for 0.03% of all neoplasms and 3% of all soft tissue tumors²⁾. Estrogen stimulation and pregnancy, previous abdominal surgery, and trauma are associated with this disease in approximately 20% to 30% of cases³⁾. Approximately 30% of patients develop the disease in association with familial adenomatous polyposis (FAP). The anterior abdominal wall is the most common site of tumor development⁴⁾, and aggressive surgery with extensive resection of the tumor and adjacent tissue is the first-line treatment for resectable tumors⁵⁾. Herein, we report a case of an abdominal wall desmoid treated by resection of the rectus abdominis muscle and anterior sheath en-masse, followed by reconstruction of the abdominal wall with a synthetic mesh.

Materials and methods

Patient

A 33-year-old woman was diagnosed with a painful palpable

mass on the left side of her lower abdomen 2 years prior. A desmoid tumor was suspected based on computerized tomography (CT) images taken at a local doctor's office; the patient was followed up with CT and medical treatment with nonsteroidal anti-inflammatory drugs. However, her symptoms did not improve, and the mass increased in size; thus, she was referred to our hospital. On physical examination, the mass was firm and fixed to the left rectus abdominis muscle. She had no relevant family history of abdominal surgery, trauma, alcohol consumption, smoking, or medications. There were no abnormal findings on preoperative blood tests. Preoperative magnetic resonance imaging (MRI) showed a well-demarcated mass that was T1 hypointense, with dimensions of 26 × 45 × 33 mm within the left rectus abdominis muscle. It demonstrated iso- to partly high-signal intensity with muscle and banded low-signal areas on T2 and homogeneous enhancement on contrast-enhanced MRI, suggestive of abdominal wall desmoid, neurogenic tumor, or granular cell tumor (Fig. 1). Written informed consent was obtained from the patient prior to surgery, including for surgery and publication of the findings.

Surgical procedure

Surgery was performed under general anesthesia. A gentle S-shaped incision was made just above the tumor along the Langer's line for a cosmetically appealing scar; the tumor was found under the anterior sheath of the rectus abdominis muscle (Fig. 2a, b). The anterior sheath was incised in the cephalocaudal direction and a prominent white lesion was identified within

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Fig. 1. Preoperative MRI image.

A well-defined mass originating from the left rectus abdominis muscle with attenuation comparable to that of the muscle is seen.

(a) T1-weighted image.

(b) T2-weighted contrast image. Uniform contrast effect and internal band of low signal area is seen. The tumor is indicated by a yellow arrow.

Fig. 2. Intraoperative findings.

(a) Preoperative incision design.

(b) A tumor under the anterior sheath is seen (yellow dotted line).

(c) Resected specimen.

(d) Post-resection wound. The posterior sheath of the rectus abdominis muscle was not infiltrated.

(e) The rectus abdominis muscle was sutured and reconstructed by reinforcing the abdominal wall with an artificial mesh.

(f) Image after closing the wound.

the rectus abdominis muscle. The tumor was partially adherent to the anterior sheath and not the posterior sheath. Resection of the muscle and anterior sheath containing the mass with a tumor-free margin of at least 2 cm was performed (Fig. 2c, d). After resection, the rectus abdominis muscle defect was sutured

with 3-0 polygractin suture. After subcutaneous dissection, the anterior sheath was sutured with 2-0 nylon suture as far as possible, and Bard soft mesh® was placed over the incision and fixed with 2-0 nuroton and 3-0 vicryl to repair the abdominal wall (Fig. 2e, f). A suction drain was placed under the anterior

sheath, and the superficial fascia, dermis, and skin were sutured in layers. The patient was discharged without complications on postoperative day 7.

Results

Pathological examination showed a desmoid tumor with at least a 2-cm negative margin. Histological examination showed dense bundles of fibroblasts and myofibroblast-like spindle-shaped cells in the collagen stroma (Fig. 3). The patient wore an abdominal binder for 3 months postoperatively to prevent hernia. The patient had an uncomplicated course during the 1-year follow-up and was satisfied with the excellent aesthetic result.

Discussion

Desmoids are rare tumors accounting for 0.03% of all neoplasms and are proliferative lesions of clonal fibroblasts. Desmoids are classified as “intermediate or locally aggressive” neoplasms because of their local invasiveness and high recurrences. The anterior abdominal wall is the most common site for desmoids (50%). Desmoids of the abdominal wall originate specifically from the rectus abdominis and internal oblique muscles and generally do not cross the midline. Externally, they do not cause skin swelling or contracture and may be detected incidentally on imaging studies⁶.

Desmoids occur primarily in young women during their reproductive years due to hormonal influences, especially during or after pregnancy. These tumors have also been associated with abdominal surgery, estrogen therapy, trauma, and FAP³; however, our patient had no history of FAP or Gardner syndrome.

Preoperative diagnosis is important for surgical planning and reconstruction. In particular, imaging can help determine the extent of the tumor, the organs involved, and the resection plan. On MRI, desmoids often have low signal intensity, compared to muscle, on T1-weighted images and high signal intensity on T2-weighted images. Desmoid tumors can be differentiated from malignant tumors such as liposarcoma and myxofibrosarcoma based on the uniformity of the contrast effect and the banded low-signal area on T2-weighted images characteristic of desmoid tumors⁷.

A definitive diagnosis should be made by histopathological analysis of the diffuse cellular infiltration of adjacent tissue structures. The immunohistochemical response for actin may be partially positive, and immunohistochemical myocyte markers may distinguish desmoid tumors from fibrosarcoma⁸. We strongly suspected a desmoid tumor based on the preoperative MRI; following consultation with the patient, we performed a one-stage resection and abdominal wall reconstruction. However, if preoperative differentiation of the

Fig. 3. Histopathological image: Fibroblasts and myofibroblast-like spindle cells in the collagen interstitium were observed in dense bundles.
bar=100µm.

tumor from other histologies (fibrosarcoma, lymphoma, rhabdomyosarcoma, liposarcoma, or leiomyosarcoma) is difficult, CT-guided biopsy should be performed.

For the treatment of desmoid tumors⁹, a surveillance period of 1–2 years is recommended to avoid overtreatment of tumors that may regress spontaneously (20%) or to assess if symptomatic treatment alone is sufficient. In cases of symptomatic progression, the treatment strategy depends on the anatomical location. Surgery is the first option in cases with abdominal wall tumors. In the case of intra-abdominal/retroperitoneal/pelvic/extremity/chest wall tumors, systemic therapy with drugs should be considered as the first treatment option⁹.

In this case, surgery with wide resection margin was chosen since the patient's tumor showed an increase in size during the 2-year follow-up period, the pain was persistent, and the tumor site was the abdominal wall.

Surgically, the mass should be resected with adequate margins and the defect reconstructed. Currently, there is no consensus on resection margins, and it depends on the patient's background and the surgeon's preference^{10–12}. However, if the resection is incomplete, the recurrence rate has been reported to be 25%–50%¹³. Therefore, adequate resection margins should be planned prior to surgery, and reconstructive options should be considered for any abdominal wall defects. Although there is no definition of surgical margins, previous studies have shown that a positive or close (<1 mm) margin significantly increases recurrence rates¹⁴; for tumors smaller than 5 cm, excisional marginal tumor-negative resection markedly reduces local recurrence rates¹³. However, unnecessarily large defects result in significant functional losses. Considering that a 2-cm margin achieved cure in a previous report¹⁵, we performed a resection of the left rectus abdominis muscle containing the tumor with a tumor-free margin of 2 cm. Recent studies have reported that minimally invasive resection preserving the

fascia leads to no difference in recurrence rate compared with that obtained with extended resection¹⁶⁾. In this case, the posterior sheath was preserved as it was uninvolved, and only the anterior sheath was resected since the tumor was adherent to it. The resulting abdominal wall defect was 6 × 5 cm, and we determined that mesh reconstruction was necessary to restore the physiological function of the abdominal wall and prevent scar site herniation. Synthetic materials are susceptible to bacterial infections and other complications; however, there were no symptoms of hematoma, hernia, infection, or bleeding in our patient. In addition to synthetic materials, there are other methods of reconstructing the abdominal wall using fascial lata patches and myocutaneous flaps. However, considering the reports of good outcomes with synthetic meshes¹⁷⁾, we opted to use these and obtained good results.

Recurrence rates vary depending on the location, extent, and completeness of the initial resection. However, the recurrence rate for abdominal wall desmoids is significantly lower at 20%. Recurrences are usually noticed within 6 months after resection or subsequent pregnancy; yet, there is no consensus on the follow-up period¹⁾. Our patient has been recurrence-free for 1 year postoperatively but needs to be followed up further.

The role of nonsurgical treatment modalities is not clear. Radiation¹⁸⁾, chemotherapy¹⁹⁾, and hormonal therapy²⁰⁾ have been reported as alternative treatments. These therapies should be considered in treating unresectable and recurrent desmoid tumors.

Conclusion

In the treatment of resectable desmoid tumors, surgery should be considered based on the course and location of the tumor, and aggressive resection with adequate margins is a major determinant of outcome. Reconstruction of the abdominal wall defect with a synthetic mesh provides a functional and satisfactory result. Further case studies are required to establish the best surgical strategy.

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None declared.

Conflicts of interest

None.

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