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Management of Giant Solitary Desmoid-type Fibromatosis: A Rare Presentation of Abdominal Pain

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1. Introduction

Desmoid-type fibromatosis are rare, locally aggressive, benign tumors of soft tissue origin [1]. They compose 0.03% of all neoplasms and less than 3% of all soft tissue tumors. Desmoid tumors often appear on the extremities or external abdominal wall [2]. Desmoid tumors commonly affect people between the ages of 15 and 60 and have a slightly higher incidence in women [1]. These tumors have a high local recurrence rate and have a high potential to infiltrate surrounding organs, although they lack malignant potential [3]. Between 5% and 15% of desmoid tumors have been associated with cases of familial adenomatous polyposis, a genetic condition involving mutations in the APC gene [4].

Desmoid tumors have also been linked to pregnancy, either related to the trauma of the pregnancy or the elevated hormone levels. Similarly, up to 30% of patients with desmoid tumors report a history of prior trauma, elucidating a possible connection between the pathogenesis of desmoid tumors and the process of wound healing [1].

2. Case Report

A 50-year-old male with a significant past surgical history of a left hepatectomy performed for living liver donation three years prior to a first degree relative presented to the Northwest Texas Hospital for abdominal pain, fullness and a rapidly growing abdominal mass. The mass began growing about 1 year prior to presentation but had increased significantly in size over the past two months. On physical examination, the patient was hemodynamically stable and had a large palpable abdominal mass present in the periumbilical area extending radially outwards (FIG. 1).

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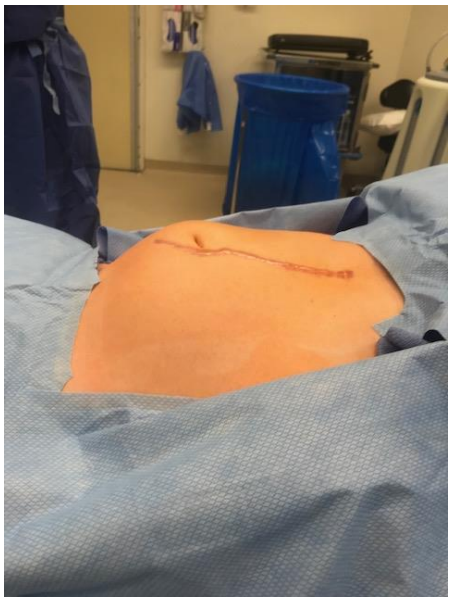

FIG. 1. **Abdominal Mass with surgical scar.**

Laboratory examination was unremarkable. CT Abdomen/Pelvis showed a large, well-defined lobular intra-abdominal mass measuring 21 cm × 10 cm × 16 cm centered in the right mid abdomen extending across the midline (FIG. 2). The patient then had a CT-guided biopsy which showed a low-grade spindle cell neoplasm suggestive of fibromatosis. The patient reported pain that was worsened with eating and sneezing, and increasing shortness of breath. The patient underwent exploratory laparotomy and excision of the abdominal mass. The midline wound was infiltrated with Kenalog. The skin overlying the mass exhibited a 15 cm in length healed tan-grey scar.

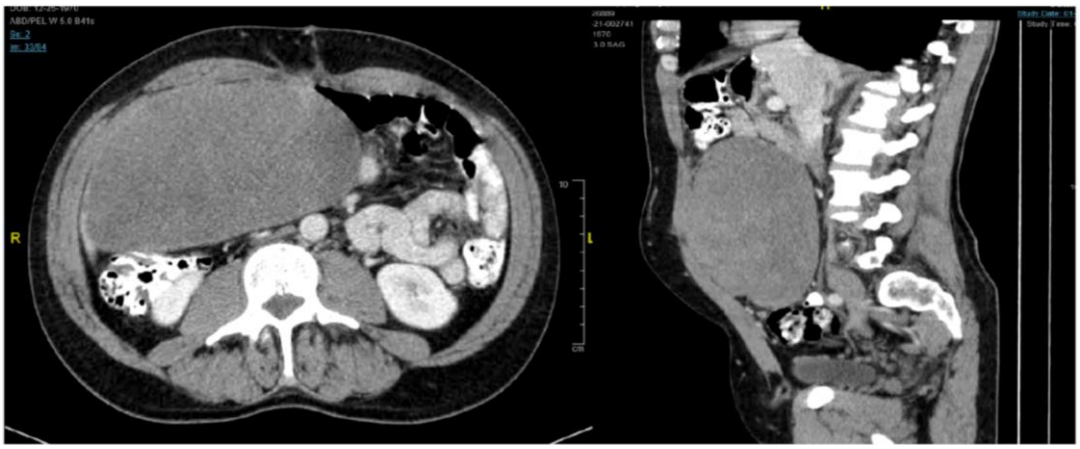

FIG. 2. **CT Scan of Abdomen/Pelvis.**

Pathology reports done post-operatively described a 2379 g, 22 cm × 14 cm × 8.5 cm encapsulated fibrous abdominal mass (FIG. 3). The capsule was described as purple-gray, shaggy and smooth with soft tissue adhesions and hemorrhagic changes. A beta catenin immunostain was performed with appropriate controls showing both positive nuclear and cytoplasmic staining of variable intensity. Patchy and weak staining for cyclic D1 was also observed. These findings were consistent with desmoid type-fibromatosis. Patient was discharged on POD #2 and was seen for follow up without any recurrence.

FIG. 3. **Extracted Desmoid-type fibromatosis.**

3. Discussion

Desmoid tumors are exceedingly rare fibroblastic neoplasms, ranging from 2-4 new cases per million individuals [1]. Despite their benign nature, they are capable of infiltrating other organs and have a very high recurrence rate (29%-30%). Mortality rates from desmoid tumors are elevated in patients with intra-abdominal tumors compared to tumors at extra-abdominal sites and can lead to bowel obstruction and perforation [2]. Desmoid tumors have a tendency to develop at sites of surgical scarring, thus supporting our patients tumor development post liver resection. The management of these tumors is based on the symptoms, location and number. Multiple small skin lesions may not be amenable to a single stage surgical resection. However, a giant abdominal tumor would require wide local excision with removal of overlying muscle and soft tissue. Laparoscopic or robotic surgery may be contraindicated if the tumor requires a large extraction site.

4. Conclusion

Many other differentials including abscess, textiloma, hematoma, lymphoma, endometriosis, other tubo-ovarian pathology and gastrointestinal cancer could lead to an abdominal mass. In this case, we present a patient with a very rare diagnoses of an abdominal mass. The aim of this case report is to enhance recognition of desmoid tumors, particularly in patients who have undergone abdominal surgery. Earlier identification of intraabdominal desmoid tumors can prevent infiltrative processes and improve long-term patient outcomes.

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