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Case Report

Mesenteric desmoid tumor: De novo occurrence or recurrence following appendectomy? ☆

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ARTICLE INFO

Article history:
Received 25 August 2021
Revised 10 October 2021
Accepted 11 October 2021

Keywords:
Desmoid tumor
Desmoid fibromatosis
Recurrent desmoid

ABSTRACT

Desmoid type fibromatosis (DF) is a rare, locally aggressive but benign proliferation of fibrous tissue which produces a fibroblastic mass that can cause a wide range of symptoms secondary to mass effect. When resected, these masses most commonly recur in the first 2 years. We present a case of a 33-year-old male with a history of an appendectomy 2 years prior, though his pathology report did not identify inflammation in the appendix, who presented with gradual onset of abdominal pain, and radiographs that demonstrated a large mass in the right lower abdomen. Given his symptoms the mass was resected and pathologic evaluation revealed a desmoid tumor. This case presents a unique possibility of a recurrent desmoid tumor in which the patient's surgical history and radiographic findings can contribute to the overall management strategy of the patient given the evolving options for treatment of desmoid fibromatosis.

Introduction

Desmoid type fibromatosis represents a spectrum of benign proliferation of fibroblastic tumors that may be classified by location, intra or extra abdominal, and superficial vs deep. When present in the abdomen, they most frequently are associated with the mesentery though may be retroperitoneal. Exact etiology remains unknown, although fibromatoses are frequently associated with previous trauma or surgical incision as with our case. The molecular pathogenesis associated with desmoid tumors arises from mutations in the Wnt signaling pathway via β-catenin and APC genes for wild type and/or sporadic forms and familial adenomatous polyposis (FAP), respectively [2,3]. Though most occur sporadically, these tumors have long been associated with familial polyposis, and Gardner Syndrome. Additionally, despite their benign nature, these neoplasms do have a propensity for multiplicity, and recurrence [4].

DF frequently appears as a well circumscribed soft tissue mass though they may have infiltrative margins, which can be seen as radiating spicules in mesenteric fibromatosis [5,6]. The appearance may also vary from a homogenous mass with attenuation similar to muscle to a heterogeneous mass with varying enhancement depending on the degree of collagen and myxoid elements [7]. In the case we present, the patient exhibited a slow-growing intra-abdominal mass that was initially diagnosed on an abdominal radiograph, and was later determined to be a mesenteric desmoid fibromatosis following surgical resection.

☆ Competing Interests: None.

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https://doi.org/10.1016/j.radcr.2021.10.028

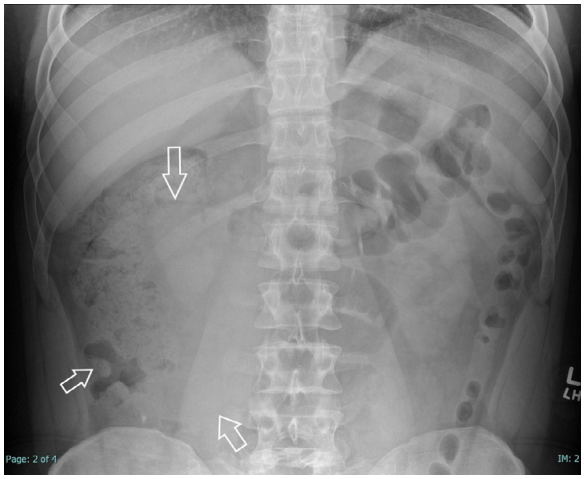

Fig. 1 – Frontal abdominal radiograph demonstrating a large, homogenous mass in the right abdomen projecting over the right 12th rib, psoas, and kidney with black arrows pointing to the margins of the mass.

Case presentation

A 33-year-old otherwise healthy male with a history of an appendectomy 2 years prior presented to an outside facility with several days of right lower quadrant abdominal pain and nausea. Orthogonal abdominal radiographs were obtained which revealed a faint, homogenous, round 13 cm soft tissue mass with displacement of bowel in the right hemi abdomen (Figs. 1 and 2).

A CT scan with oral and IV contrast was then obtained following transfer to our facility which confirmed a large intra-abdominal mass in close proximity to the cecum and terminal ileum in the right lower quadrant mesentery with punctate locule of gas which raised the possibility of bowel origin. The mass was well circumscribed though there were small linear areas of spiculated increased attenuation at its margins into the mesentery. These findings were felt to be concerning for a GIST, leiomyoma, desmoid tumor, lymphoma or adenocarcinoma (Figs. 3 and 4).

Based on the imaging findings and symptoms, the patient was taken to the operating room for exploratory laparotomy where the mass was found to be emanating from the mesentery adjacent to the cecum. An oncologic right colectomy was performed with a photo of the gross specimen below (Fig. 5). Pathologic evaluation of the mass revealed mesenteric fibromatosis and/or desmoid tumor.

Discussion

Intra-abdominal desmoid type fibromatoses are rare tumors (0.03% of all neoplasms) and most commonly occur in the small bowel mesentery. These tumors are most frequently encountered between 20-40 years of age but are found in all age groups with female predominance. Unique and pertinent to

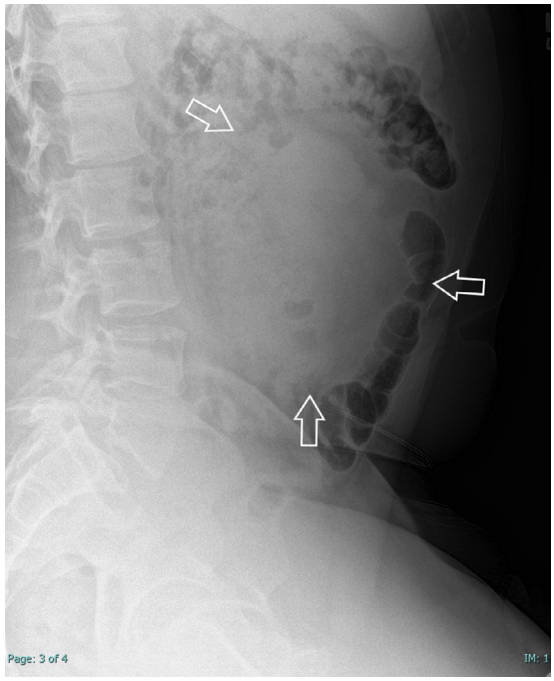

Fig. 2 – Lateral radiograph demonstrating the abdominal mass which displaces bowel anteriorly with white arrows pointing to the margins.

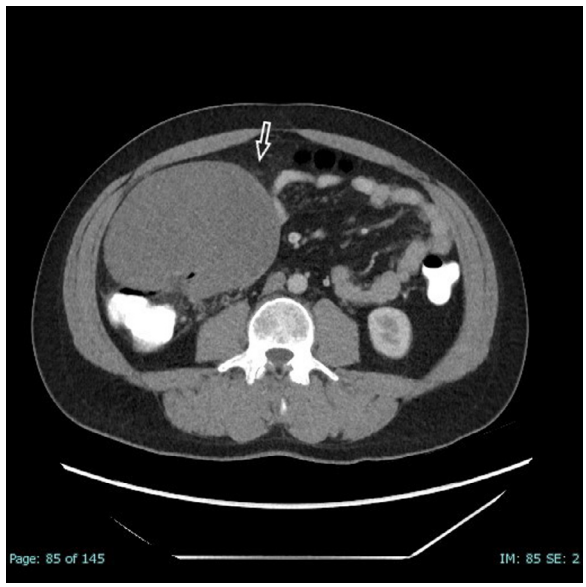

Fig. 3 – Axial contrast enhanced CT demonstrating a large round homogenous mass in the right lower quadrant mesentery with small focus of air posteriorly along with small spiculated margins in the mesentery (white arrow). The mass is isodense relative to abdominal wall musculature, typical of desmoid-type fibromatosis.

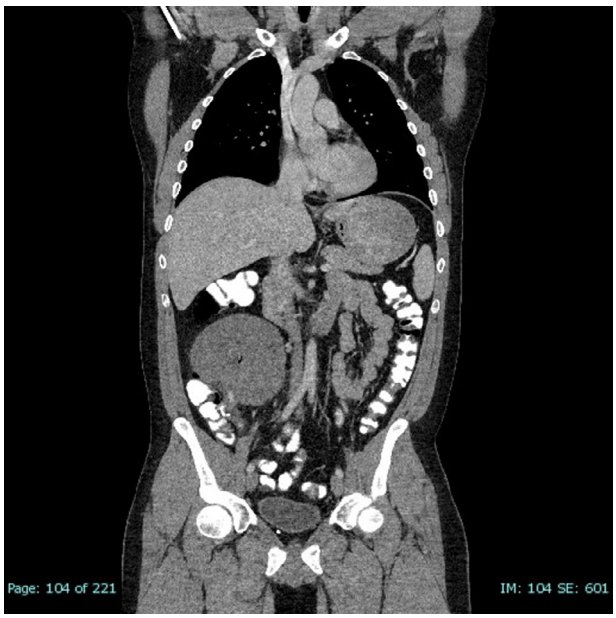

Fig. 4 – Coronal contrast enhanced CT showing a large desmoid manifesting as a homogenous well circumscribed soft tissue mass with a single air locule.

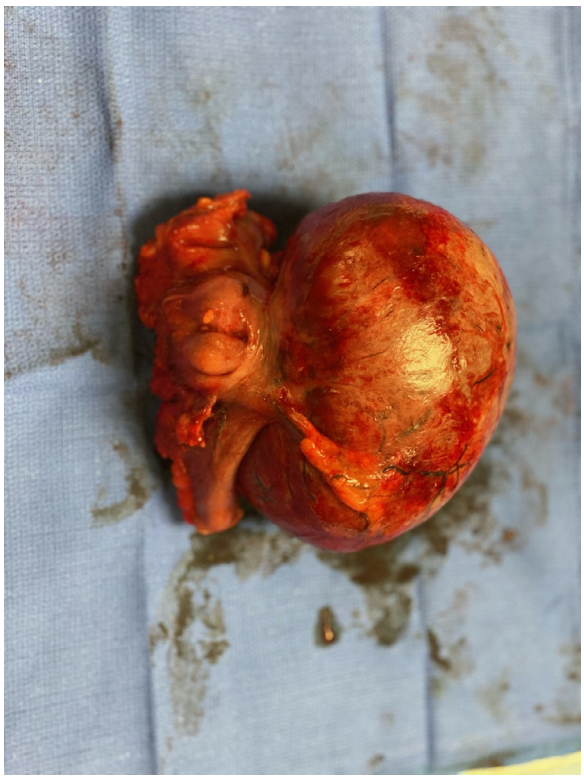

Fig. 5 – Gross specimen.

the case, desmoid tumors that have been resected most often re-occur within 2 years, which is a similar time course from the prior appendectomy in this case [1]. There are reported cases of cecal and appendicular desmoid tumors raising the possibility that his initial presentation of appendicitis was the beginning of desmoid proliferation, especially given the location of recurrence [7,8]. The gross pathology report from an outside hospital stated the appendix did not contain a fecalith, was not grossly inflamed, and contained a lumen with "scant amount pink-red soft material." Differential diagnoses for desmoids include soft tissue sarcomas, lymphoma, soft tissue metastasis, carcinoid tumor, and sclerosing mesenteritis.

Treatment

Classically treatment has consisted of early surgery due to the locally invasive risks of DF and/or desmoid tumors. Yet recent evidence shows that a multidisciplinary approach with more conservative options may have better outcomes, to include watchful waiting [9]. Other treatment options include systemic therapy in the form of antiestrogen therapy and NSAIDs, Tamoxifen, and Sulindac. Chemotherapy and radiation may also be considered for more aggressive tumors that have invaded critical structures, though are used less commonly due to their side effect profiles [1]. Once a patient is symptomatic due to mass effect, surgical resection will be the treatment of choice if resectable, as was the case for this patient. Without a pathologic diagnosis, the aim is a complete resection without microscopic involvement at the margins as to reduce the risk of recurrence in addition to treatment of symptoms secondary to mass effect with preservation of critical structures [6].

Treatment strategies often rely on important information that can be gained from imaging findings. Though symptoms dictate management, it is important for a radiologist to be able to provide a differential that may guide operative planning to help reduce the risk of recurrence. Desmoid tumors can have a varying appearance on CT, ranging from a homogenous mass with attenuation similar to skeletal muscle to a heterogenous lesion depending on the internal cellularity. Additionally, these can be well circumscribed though may also have infiltrative, spiculated margins. When the clinical situation allows, MR can provide more information on the gross histologic characteristics of the mass, and is therefore the preferred modality for evaluation [4]. When mostly comprised of fibrous, collagen bands desmoids will have low intensity on T1 and T2 sequences, however, if they contain more cellular stroma or myxoid elements there will be increased signal on T2 weighted sequences [4,8].

All genetic forms of desmoid show an equivocal risk of recurrence, thus, after resection or definitive therapy follow up imaging should be conducted with CT or MRI every 3-6 months for 2-3 years, then every 6-12 months thereafter. Ultrasound may also be considered for select anatomic locations such as abdominal wall tumors for long term imaging surveillance [10,11]. It is also important to note that there is an increased risk for recurrence if there are positive microscopic

margins which reiterates the importance of providing an accurate differential for operative planning [12]. Ancillary testing for APC gene mutations or colonoscopy may also be considered, especially in a patient who presents with mesenteric DF. Results from genetic testing in this case are pending which will allow for further risk stratification in addition to a future colonoscopy following recovery from surgery.

Lastly, if this patient were to have a local recurrence of the same area, the operating surgeon should be aware that intraoperative frozen section analysis will be unreliable due to the inability to differentiate desmoid fibromatosis from scar on frozen analysis. [1]

Patient consent

Written consent was not obtained prior to submission due to use of anonymized, non–identifiable radiographs, single images from a CT scan, and a single gross pathology image.

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