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Single Case

An Intra-Abdominal Desmoid Tumor with Edematous Loose Collagen Fibers

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Keywords

Aggressive fibromatosis · Central necrosis · Desmoid tumor · Edema · Loose collagen fiber

Abstract

Introduction: Diagnostic physicians tend to judge a low-dense area on computed tomography (CT) as central necrosis when it has no contrast enhancement and locates in the center of large tumors. ***Case Presentation:*** An 80-year-old woman was referred to our hospital due to the detection of an abdominal mass on ultrasound (US). CT showed a well-demarcated oval mass, 11 cm in size, with a central low-density area. US showed high internal echoes and enhanced posterior echoes. Magnetic resonance imaging (MRI) showed the low-density area on CT to be hypo-intense on T1-weighted images and hyper-intense on T2-weighted images. MRI further showed the central part of the tumor to be hyper-intense both on diffusion-weighted images and apparent diffusion coefficient images. Under the tentative diagnosis of a gastrointestinal stromal tumor with central necrosis, the patient underwent tumor resection, revealing the tumor to be a jejunal submucosal tumor. Pathological study showed collagen fibers with heterogeneous density and sparse proliferation of spindle cells. The center of the tumor had marked edema in addition to sparse collagen fibers. Immunostaining showed that the atypical cells were diffusely positive for β catenin and negative for S100 protein, desmin, and DOG1, leading to the diagnosis of desmoid tumor (DT). ***Conclusions:*** Physicians should note that intra-abdominal DT can have edematous loose collagen fibers and may show central necrosis-like findings on CT.

Introduction

Desmoid tumor (DT), also called aggressive fibromatosis, highly infiltrates the surrounding tissue but never develops distant metastasis. It, therefore, is classified as an intermediate tumor between benign and malignant tumors by the World Health Organization [1]. DT is extremely rare with an incidence of 2–4 per million people and occurs twice as often in women as in men [2].

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DTs are classified into two subtypes according to the tumor site: extra-abdominal and intra-abdominal DTs. DTs sometimes regress spontaneously [3] and therefore need careful differential diagnosis with other disorders such as gastrointestinal stromal tumors (GISTs) [4] and malignant lymphomas.

We herein report a case of DT which had edematous loose collagen fibers and was preoperatively misjudged as a GIST with central necrosis [5]. The CARE checklist has been completed and attached as an online supplementary file (for all online suppl. material, see https://doi.org/10.1159/000543498).

Case Presentation

An 80-year-old woman was referred to our hospital because her attending physician detected a mass in her left upper abdomen with ultrasound (US). Computed tomography (CT) showed a well-demarcated oval mass, 11 cm in size, with a central low-density area (Fig. 1). US showed that the tumor had high internal echoes in its central area and enhanced posterior echoes (Fig. 2). Magnetic resonance imaging (MRI) showed that the low-density area on CT was expressed as hypo-intense on T1-weighted images and hyper-intense on T2-weighted images (Fig. 3a, b). MRI further showed that the central area of the tumor was hyper-intense both on diffusion-weighted images (DWI) and apparent diffusion coefficient (ADC) images and the major parts of the tumor's peripheral areas were hyper-intense on DWI and hypo-intense on ADC images (Fig. 3c, d). Both aspirin use and possible dissemination of malignant cells to the abdominal cavity led us not to perform endoscopic US-guided biopsy. Therefore, under the tentative diagnosis of a GIST with central necrosis, the patient underwent surgical resection of the target tumor. The operation revealed the tumor to be a jejunal submucosal tumor and consisted of tumor resection with sufficient safety margins followed by jejuno-jejunostomy. Postoperative pathological study showed that the tumor had sparse proliferation of spindle cells in the background collagen fibers (Fig. 4a) and central edematous areas with sparse collagen fibers. The low-density area on CT well corresponded to the area of the edematous sparse collagen fibers. Immunostaining showed that the atypical cells were strongly positive for β catenin [6] (Fig. 4c), were negative for S100 protein, desmin, and DOG1, and had a Ki-67 labeling index of 2.7%, leading to the diagnosis of an indolent DT. The patient recovered without any events and was discharged from the hospital on the 7th postoperative day.

Discussion

Internal echoes of the central part of the tumor were high in this case. Ischemia-induced liquefied tissue with uniform acoustic impedance, if had been present, would have generated less US backscattering and have made the internal echoes low. Conversely, high internal echoes suggested the presence of heterogenous components with different acoustic impedances [7]. Therefore, co-presence of spindle cells and edematous loose collagen fibers had presumably resulted in the high internal echo formation in this case.

CT showed a clearly demarcated mass mainly composed of two areas with different CT Hounsfield unit (HU) values: the low-dense central areas and the highly dense peripheral areas. Small differences in CT HU values can occur in various pathological conditions. We cannot speculate about the pathological components in detail based on CT findings alone. It,

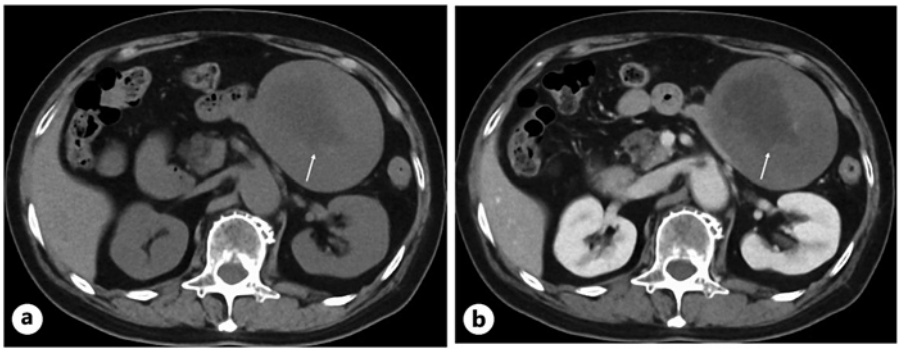

Fig. 1. Computed tomography (CT) findings. **a** Plain CT showed an oval mass with a somewhat hypo-dense area (arrow) in the center of the tumor. **b** Contrast CT showed no contrast effect in the central area (arrow) and made the hypodensity more prominent.

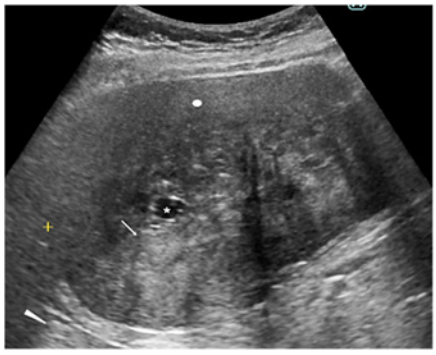

Fig. 2. US findings. US showed low echoic areas in the peripheral part of the tumor (closed circle), high echoic areas (arrow) with a small cyst (asterisk) in the center of the tumor, and posterior echo enhancement (arrowhead).

therefore, was too hasty for us to judge that the tumor was a GIST with central necrosis based only on the tumor size and the slightly low central HU value on CT.

Peripheral areas of the tumor showed a hypo-intense pattern on both T1- and T2-weighted images, presumably having reflected the abundance of collagen fibers. In addition, judging from the ADC image findings, the hyper intensity on DWI in the central areas of the tumor did not reflect the diffusion impairment by tumor cells but was presumably caused by T2 shine-through effect, i.e., very edematous [8].

Considering all of the above pathological and image findings, it can be concluded that the presence of edematous loose collagen fibers generated the central necrosis-like findings on CT. Ganeshan et al. [9] had also pointed out the possible presence of myxoid stroma in intra-abdominal DTs. This case clearly demonstrates that an abdominal mass without a preoperative pathological diagnosis can be accurately predicted of its pathological components based on the thorough understandings of basic principles of each image modality.

Aggressive DTs have denser spindle cells (Fig. 4b) and often infiltrate surrounding organs. Aggressive DTs, however, generally have low internal echoes because homogenous pathological components of them generate less US wave backscattering. High internal echoes, therefore, imply the indolent biology of the tumor. High intensity on DWI by T2 shine-through further supports this tumor biology.

In this case, regardless of whether the preoperative diagnosis was DT or GIST, surgery remained the mainstay of treatment. However, if the tumor had been in contact with surrounding vital organs such as the pancreas or bile duct, possible invasion could have a significant impact on resectability and the surgical method. In other words, if the hyperechoic

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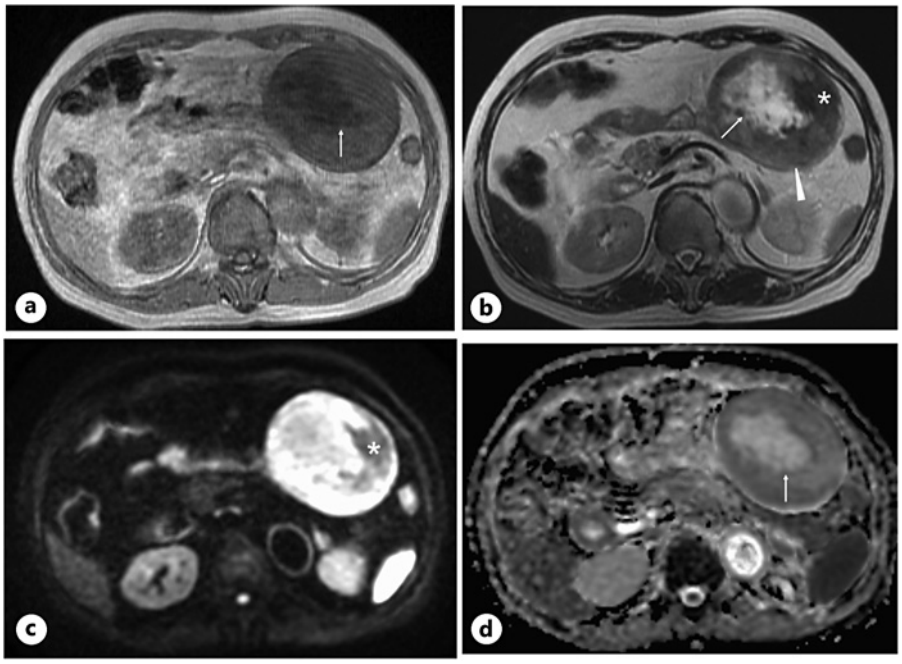

Fig. 3. Magnetic resonance image (MRI) findings. **a** T1-weighted images showed a hypo-intense oval mass with more hypo-intense areas (arrow) in the center of the tumor. **b** T2-weighted images showed hyper-intense areas (arrow), hypo-intense areas (arrowhead), and more hypo-intense areas (asterisk) in the tumor. **c** DWIs showed a hyper-intense pattern except for some areas of the tumor borders (asterisk). **d** ADC images showed the center of the tumor to be somewhat hyper-intense (arrow).

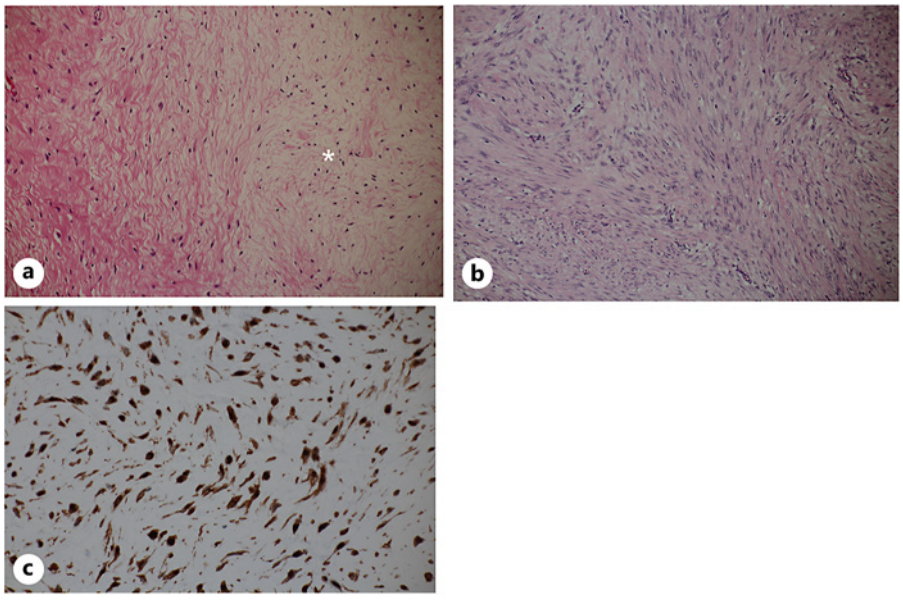

Fig. 4. Pathologic findings. **a** The tumor had sparse and uniform spindle cells with background collagen fibers throughout the tumor and the central areas had sparser collagen fibers with marked edema (asterisk). **b** Extra-abdominal desmoid fibromatosis as a reference pathology showed much more dense spindle cells and collagen fibers than those observed in this case. **c** Immunostaining showed the spindle cells to be strongly positive for β-catenin.

areas with T2 shine-through effect on DWI had been present in the areas adjacent to surrounding organs at risk of infiltration, these image findings would strongly suggested that dissection between the tumor and adjacent organs was possible.

Conclusion

Physicians should note that intra-abdominal DT can have edematous loose collagen fibers and show somewhat different image findings than extra-abdominal DTs. Furthermore, surgeons should understand that if hyperechoic areas, such as those seen in this case, are observed in areas adjacent to surrounding organs suspected of infiltration, echo findings highly suggest possible dissection between the tumor and the surrounding organs.

Statement of Ethics

This article does not contain any studies involving human participants or animals performed by any of the authors. Ethical approval was not required for this case study in accordance with local or national guidelines. Written informed consent was obtained from the patient for the publication of this case report and accompanying images.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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Author Contributions

M.I. contributed to the design of the report. S.O. drafted the manuscript. All authors have read and approved the final version of the manuscript.

Data Availability Statement

All data generated or analyzed during this study are included in this article and its online supplementary material files. Further inquiries can be directed to the corresponding author.

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