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Case Report

Contrast-enhanced ultrasonography combined with superb microvascular imaging for preoperative diagnosis of sporadic intra-abdominal desmoid-type fibromatosis: A case report☆,☆☆

Kazuhiro Komi, MD[a,b], Minoru Fujita, MD, PhD[c,], Noriaki Manabe, MD, PhD[c], Munenori Takaoka, MD, PhD[d], Katsuya Kato, MD, PhD[e], Hideyo Fujiwara, MD, PhD[f], Takako Konishi, MD[c,d], Emiko Bukeo, MD[c], Hiraku Misawa, MD[c], Jun Nakamura, MD, PhD[c], Maki Ayaki, MD, PhD[c,g], Takahisa Murao, MD, PhD[h], Mitsuhiko Suehiro, MD, PhD[i], Takashi Akiyama, MD, PhD[f], Hirofumi Kawamoto, MD, PhD[i], Tomoari Kamada, MD, PhD[h], Atsushi Urakami, MD, PhD[d], Yoshio Naomoto, MD, PhD[d], Tomoki Yamatsuji, MD, PhD[d], Takuya Moriya, MD, PhD[b], Ken Haruma, MD, PhD[i], Jiro Hata, MD, PhD[j]*

[a] *Center for Doctors' Career Development, Kawasaki Medical School Hospital, Kurashiki, Japan*
[b] *Department of Pathology, Kawasaki Medical School Hospital, Kurashiki, Japan*
[c] *Division of Endoscopy and Ultrasonography, Department of Clinical Pathology and Laboratory Medicine, Kawasaki Medical School General Medical Center, Okayama, Japan*
[d] *Department of General Surgery, Kawasaki Medical School General Medical Center, Okayama, Japan*
[e] *Department of Diagnostic and Therapeutic Radiology, Kawasaki Medical School General Medical Center, Okayama, Japan*
[f] *Department of Pathology, Kawasaki Medical School General Medical Center, Okayama, Japan*
[g] *Department of Gastroenterology and Hepatology, HITO Medical Center, Shikokuchuou, Japan*
[h] *Department of Health Care Medicine, Kawasaki Medical School General Medical Center, Okayama, Japan*
[i] *Department of General Internal Medicine 2, Kawasaki Medical School General Medical Center, Okayama, Japan*
[j] *Division of Endoscopy and Ultrasonography, Department of Clinical Pathology and Laboratory Medicine, Kawasaki Medical School Hospital, Kurashiki, Japan*

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ABSTRACT

We herein report a case of sporadic intra-abdominal desmoid-type fibromatosis in which contrast-enhanced ultrasonography (US) combined with superb microvascular imaging (SMI) was useful for preoperative diagnosis. 18-Fluorodeoxyglucose positron emission tomography performed for systematic screening for lung cancer revealed an abnormal ac-

Abbreviations: CT, computed tomography; FDG-PET, 18-fluorodeoxyglucose positron emission tomography; MRI, magnetic resonance imaging; SMI, superb microvascular imaging; US, ultrasonography.

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* Corresponding author.
E-mail address: minorufu@med.kawasaki-m.ac.jp (M. Fujita).

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cumulation in the abdominal cavity. Transabdominal US showed a tumor with a mixture of hypoechoic and hyperechoic areas. Contrast-enhanced US combined with SMI revealed dendritic blood flow signals and no abnormal vascular network within the tumor. Macroscopic examination of the resected specimen revealed a white tumor with relatively clear boundaries. Microscopic examination revealed spindle cells with poor atypia proliferating in bundles with collagenous stromal cells. Immunohistochemistry showed nuclear localization of beta-catenin within the tumor cells. From these findings, we finally diagnosed intra-abdominal desmoid-type fibromatosis. Contrast-enhanced US combined with SMI is useful for diagnosing intra-abdominal desmoid-type fibromatosis.

Introduction

Desmoid-type fibromatosis is extremely rare, with an incidence of 2 to 5 cases per million inhabitants in European countries. The median age at diagnosis of desmoid-type fibromatosis is approximately 35 years, and most patients are women [1,2]. Desmoid-type fibromatosis can be intra-abdominal, within the abdominal wall, extra-abdominal, or located at multiple sites. Extra-abdominal desmoid-type fibromatosis can develop in any musculoaponeurotic structure throughout the body, such as the shoulder, chest wall, breast, back, thigh, head, or neck. Intra-abdominal desmoid-type fibromatosis arises within the mesentery or pelvis. By contrast, desmoid-type fibromatosis within the abdominal wall arises from musculoaponeurotic structures of the abdominal wall, especially the rectus muscle, internal oblique muscle, and their fascial coverings [3,4]. This type of desmoid-type fibromatosis has a higher rate in women with a recent pregnancy [5].

Intra-abdominal desmoid-type fibromatosis is usually diagnosed using abdominal computed tomography (CT) and/or abdominal magnetic resonance imaging (MRI) [6,7]. Transabdominal ultrasonography (US) and contrast-enhanced US are also useful for diagnosing intra-abdominal lesions [8–11]. These radiation-free modalities can be performed repeatedly and have excellent real-time performance. In some past case reports, intra-abdominal desmoid-type fibromatosis was observed using transabdominal US [12,13]. However, no reports have described using contrast-enhanced US for this purpose. We herein present a case of intra-abdominal desmoid-type fibromatosis in which contrast-enhanced US was useful for preoperative diagnosis.

Case report

A woman in her 70s visited a physician at our institution for a detailed examination of a tumor in the left lower lung field pointed out during a medical examination. A lung tumor was diagnosed as adenocarcinoma by CT-guided percutaneous lung biopsy. 18-Fluorodeoxyglucose positron emission tomography (FDG-PET) performed for systematic screening revealed an abnormal accumulation in the abdominal cavity. Therefore, she underwent a consultation for gastro-intestinal surgery. Her operative history included laparoscopic cholecystectomy for gallstones and hysterectomy for uterine myoma. Her family history was unremarkable. Physical examination revealed a fist-sized mass without tenderness in the umbilical region. Laboratory tests showed a carcinoembryonic antigen level of 4.3 ng/mL (reference range, 0.0–5.0 ng/mL) and carbohydrate antigen 19-9 level of <2.1 U/mL (reference range, 0.0–37.0 U/mL). Her white blood cell count, red blood cell count, platelet count, liver function, and renal function were normal.

FDG-PET showed FDG accumulation in the abdominal tumor below the abdominal wall at the level of the umbilicus. The FDG accumulation had a maximum standardized uptake value of 4.41 (Fig. 1, red arrow). Transabdominal US (Aplio i700 TUS-I700; Canon Medical Systems, Otawara, Japan) showed a tumor with a mixture of hypoechoic and hyperechoic areas (35.0 × 28.7 mm) in the abdominal cavity (Fig. 2A, red arrow). A sliding sign between the abdominal wall and the tumor and between the gastrointestinal tract and the tumor was confirmed. No ascites or peritoneal nodules were observed in the abdominal cavity. Contrast-enhanced US using perflubutane (Sonazoid; GE Healthcare, Oslo, Norway) showed no echogenic areas suggestive of necrosis within the

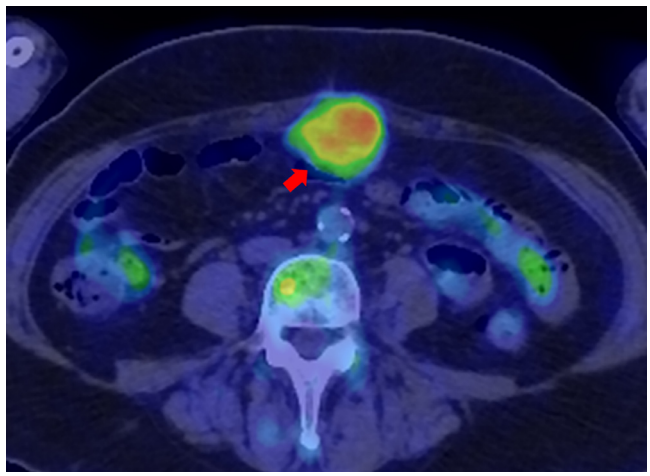

Fig. 1 – FDG-PET findings. FDG accumulation was observed in the abdominal tumor below the abdominal wall at the umbilical level (red arrow). FDG-PET, 18-fluorodeoxyglucose positron emission tomography.

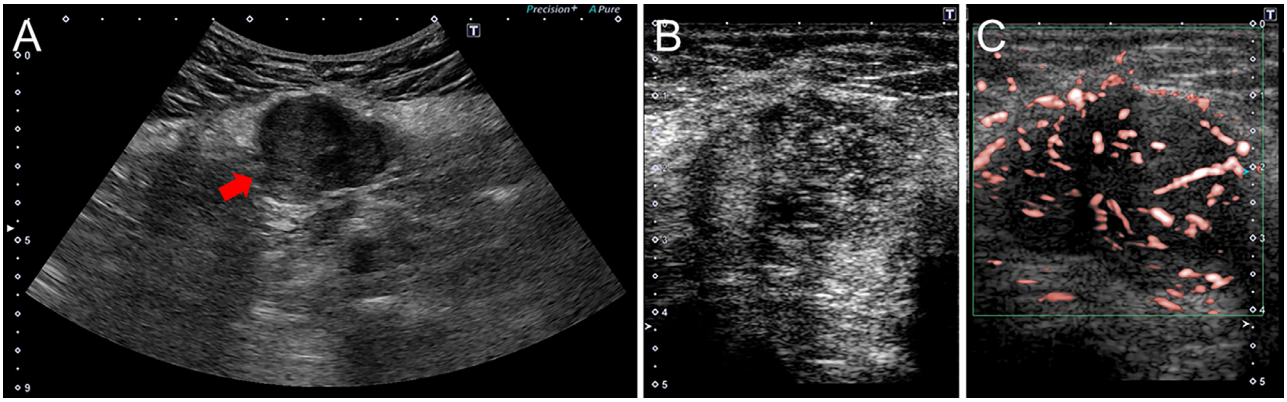

Fig. 2 – Transabdominal US and contrast-enhanced US findings. (A) Transabdominal US showed a tumor with a mixture of hypoechoic and hyperechoic areas in the abdominal cavity (red arrow). (B) Contrast-enhanced US showed no echogenic areas suggestive of necrosis within the tumor. (C) Contrast-enhanced US combined with superb microvascular imaging revealed dendritic blood flow signals and no abnormal vascular network within the tumor. US, ultrasonography.

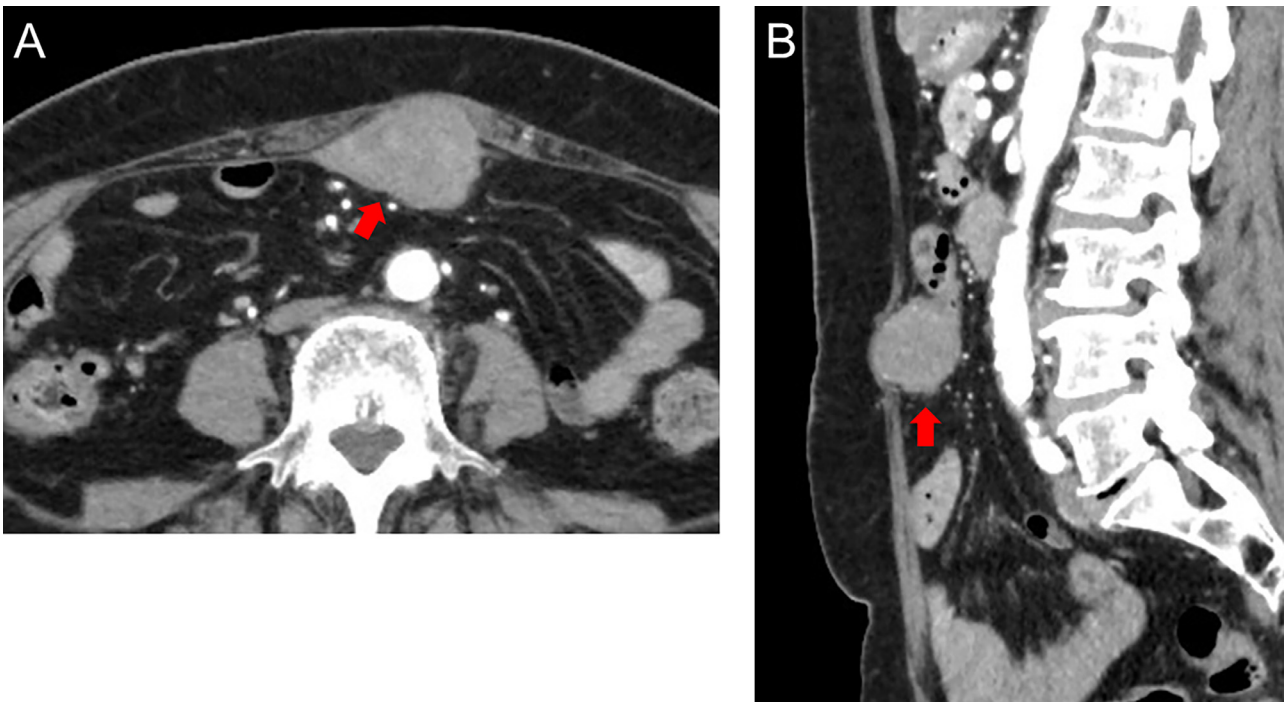

Fig. 3 – Contrast-enhanced computed tomography findings. A tumor with clear borders and contrast enhancement was observed in the abdominal cavity at the level of the umbilicus (red arrows). (A) Axial image. (B) Sagittal image.

tumor (Fig. 2B). Contrast-enhanced US combined with superb microvascular imaging (SMI) (Canon Medical Systems) revealed dendritic blood flow signals and no abnormal vascular network within the tumor (Fig. 2C). Contrast-enhanced CT demonstrated a tumor of 45 mm in diameter with clear borders and uniform contrast enhancement at the level of the umbilicus. Part of the tumor had invaded the abdominal wall and transverse colon (Fig. 3A, axial image, red arrow; Fig. 3B, sagittal image, red arrow). On abdominal MRI, the tumor showed isointense signals relative to muscle on T1-weighted images (Fig. 4A, axial image, red arrow) and mild high-intensity signals on T2-weighted images, (Fig. 4B, axial image, red arrow). The tumor showed a gradual and uniform contrast enhancement effect on fat-suppressed gadolinium-enhanced T1-weighted images (Fig. 4C, sagittal image, red arrow). Colonoscopy showed no neoplastic lesions.

We preoperatively diagnosed intra-abdominal desmoid-type fibromatosis and performed peritoneal resection and partial transverse colectomy. The patient resumed eating on postoperative day 3. She developed a fever of 38.0°C or higher but was discharged 26 days after surgery.

Macroscopic examination of the resected specimen demonstrated a well-circumscribed, whitish tumor

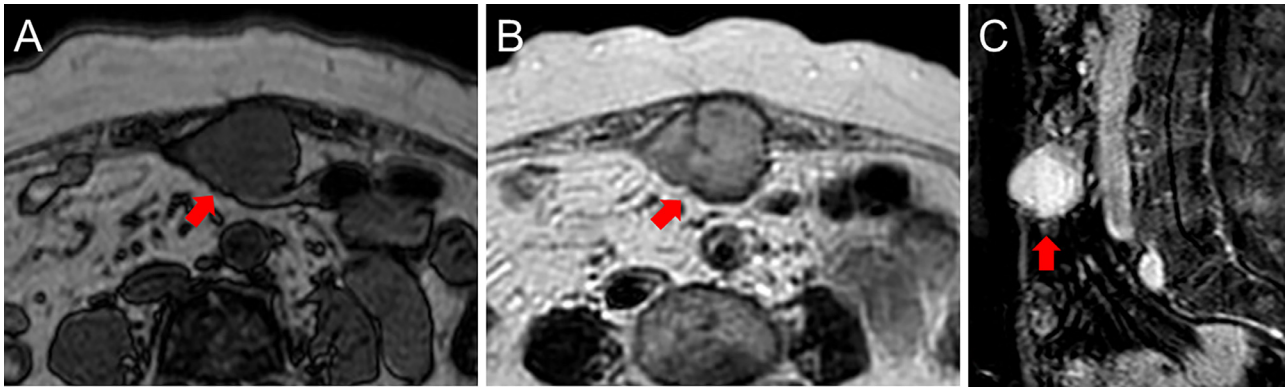

Fig. 4 – Magnetic resonance imaging findings. A mosaic pattern was observed, with a mixture of (A) isointense signals within the tumor on a T1-weighted axial image (red arrow) and (B) mild high-intensity signals on a T2-weighted axial image (red arrow). (C) A sagittal fat-suppressed gadolinium-enhanced T1-weighted image showed a gradual and uniform contrast enhancement effect (red arrow).

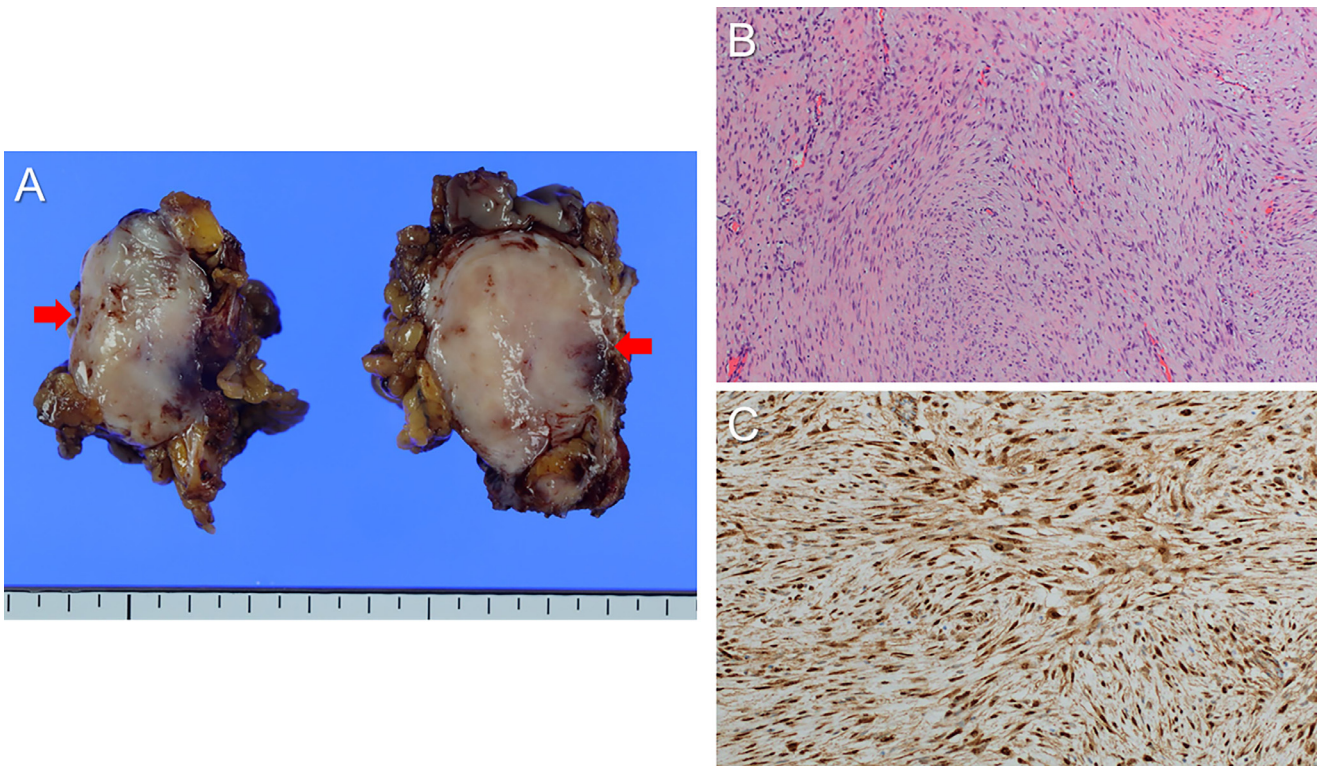

Fig. 5 – Macroscopic and microscopic findings of the resected specimen. (A) Macroscopic examination demonstrated a well-circumscribed, whitish tumor (red arrows). (B) Microscopic examination of the resected specimen revealed spindle cells with poor atypia proliferating in bundles with collagenous stromal cells. (C) Immunohistochemistry showed nuclear localization of beta-catenin within the tumor cells.

(57 × 45 × 25 mm) (Fig. 5A, red arrows). Microscopic examination revealed spindle cells with poor atypia proliferating in bundles with collagenous stromal cells (Fig. 5B). Immunohistochemistry showed nuclear localization of beta-catenin within the tumor cells (Fig. 5C) and negativity for c-kit, DOG-1, desmin, S-100, STAT6, and CD34 (data not shown). The median Ki-67 index was 5.5%. The surgical margin was negative. From these findings, we pathologically diagnosed intra-abdominal desmoid-type fibromatosis.

Abdominal CT performed 6 months after surgery revealed no findings of recurrence.

Discussion

Most cases of desmoid-type fibromatosis develop sporadically. However, some cases occur in the context of Gardner's syn-

drome (a variant of familial adenomatous polyposis), and their location is more often intra-abdominal than is the sporadic form [14]. Tumorigenesis is linked to beta-catenin stabilization involving beta-catenin/Wnt/T-cell factor signaling [15,16]. In the present case, colonoscopy showed no evidence of familial adenomatous polyposis, and beta-catenin was immunohistologically positive in the resected tumor specimen. Therefore, we diagnosed sporadic intra-abdominal desmoid-type fibromatosis.

Contrast-enhanced US can be used to evaluate vascular structure. Onji et al. [17] reported that the microvascular structure in colon cancer was irregular and characterized by large vessels, whereas acute inflammation was not observed. Alternatively, SMI is a new technology that can distinguish Doppler signals generated by low-velocity blood flow from those generated by tissue movement. This technique can reduce motion artifacts and simultaneously provide a high level of sensitivity and imaging resolution at a high frame rate [18]. The effect of SMI is further enhanced by combining it with contrast-enhanced US [10]. For preoperative diagnosis in the present case, it was necessary to differentiate between intra-abdominal desmoid-type fibromatosis and intra-abdominal metastasis of lung cancer because FDG-PET showed FDG accumulation within the tumor. We found no vascular network abnormalities within the intra-abdominal tumor as visualized by contrast-enhanced US combined with SMI. Additionally, transabdominal US and other imaging examinations revealed no ascites or disseminated nodules, thus ruling out a metastatic tumor.

Complete surgical removal remains the optimal treatment for intra-abdominal desmoid-type fibromatosis. However, it is sometimes impossible to radically remove the tumor because of the tumor's involvement with the base of the mesentery and its proximity to vital neurovascular structures. In some previous reports, the local recurrence rate was 15% to 30% even after complete resection [2,3]. Salas et al. [16] reported that an age of >37 years, tumor size of <70 mm, and tumors located in the abdominal wall or intra-abdominally have a lower risk of recurrence after surgery. Our patient was in her 70s and had an intra-abdominal tumor of <70 mm in size, and complete resection was possible. Although the risk of local recurrence appears to be low, careful follow-up is necessary.

Conclusion

We have reported a case of intra-abdominal desmoid-type fibromatosis in which contrast-enhanced US combined with SMI was useful for preoperative diagnosis. Contrast-enhanced US combined with SMI can depict the vascular structure more clearly and is useful for qualitative diagnosis of intra-abdominal tumors.

Compliance with ethical standards

This study was conducted in accordance with the principles of the Declaration of Helsinki. This study was reviewed and approved by the Institutional Ethics Committee of Kawasaki Medical School (approval number: 6342). This study included no experiments involving human participants performed by any authors.

Patient consent

Written informed consent was obtained from the patient to publish this case report and any accompanying images.

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