Introduction

Desmoid tumor is known as deep fibromatosis, aggressive fibromatosis, or well-differentiated fibrosarcoma. It accounts 1.5 to 3% of all soft tissue masses and less than 1% of retroperitoneal tumors (1, 2). Desmoid tumor can be sporadic or be associated with previous operation history, familial adenomatous polyposis (FAP, as Gardner syndrome), estrogen or pregnancy showing female predilection (2). Herein, we present a sporadic case of a huge desmoid tumor in a young male patient.

Case Report

A 25-year-old men was referred to our institution for evaluation of ultrasound detected left upper quadrant mass (Fig. 1A) due to left flank pain. He suffered from left flank pain for one month. He had undergone appendectomy for acute appendicitis eight years ago. Except this medical history, he had no other underlying disease including familial adenomatous polyposis (FAP).

On pre-operative contrast-enhanced CT, a huge (more than 22 cm) solid mass was found at the left upper quadrant (Fig. 1B-E). The solid lesion showed heterogeneous enhancement on CT. It displaced spleen medially and left kidney caudally and closely abutted to splenic flexure colon. 18F-Fluorodeoxyglucose (FDG) PET-CT was performed and the mass showed FDG uptake on FDG PET-CT (Fig. 1F).

He underwent tumor excision, under suspicious of malignant mass such as lymphoma or myxoid liposarcoma. On surgical specimen, the huge yellowish mass showed adhesion to spleen, splenic flexure colon and abdominal wall. Therefore, retroperitoneal mass excision was performed with splenectomy and segmental colectomy. On histopathology, the solid lesion was filled with spindle cells on H&E staining, and was pathologically confirmed.
Fig. 1. Imaging findings of a desmoid tumor in a 25-year-old man. A more than 18 cm sized low echoic lesion was seen on ultrasound at the left upper quadrant area, during evaluation for left flank pain (A). On contrast enhanced CT, a more than 22 cm sized, huge solid lesion is seen at the left upper retroperitoneum (B-E). It shows heterogeneous enhancement, displaces the spleen and left kidney, and encases splenic flexure colon. On 18F-Fluorodeoxyglucose (FDG) PET-CT, FDG uptake was shown (F).
as a desmoid tumor, showing β-Catenin and smooth muscle actin (SMA) staining (Fig. 2).

**Discussion**

Desmoid tumor arises from musculoaponeurotic structures and disrupts adjacent muscular and soft-tissue planes. Desmoid tumor can appear as an ill-defined infiltrative soft-tissue mass in mesenteric or deep tumors, while it can be sharply margined tumors, in abdominal-wall (3).

For diagnosis and follow-up, CT is the most commonly used imaging modality. It shows variable attenuation or enhancement patterns according to its compositions (spindle cell, collagen and myxoid matrix) (2). With abundant myxoid elements, it can be hypoattenuated, while necrosis and calcifications are rare (4). MRI can reflects those component, with excellent soft-tissue contrast. T2 high signal intensity can be shown with myxoid matrix, while low T2 signal intensity with abundant

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**Fig. 2.** Pathologic findings of a desmoid tumor in a 25-year-old man. The huge solid tumor was hard and infiltrative to adjacent organ, thus splenectomy and segmental colectomy were performed with retroperitoneal tumor excision (A). The tumor composed of spindle cells on H&E staining (x 100) (B). On immunohistochemistry, β-Catenin (C) and smooth muscle actin (SMA) (D) staining showed positive, thus desmoid tumor was diagnosed.
collagen component (3). Desmoid tumor needs to be differentiated from other neoplasms, such as lymphoma, pleomorphic sarcoma, and fibrosarcoma with imaging findings, thus pathologic confirmation is needed before definitive treatment (3).

If possible, surgical resection with a wide margin is the treatment of choice for symptomatic desmoid tumors. When operation is not feasible due to tumor’s location, local invasion state or patient’s conditions, the other treatment option includes radiotherapy, conventional chemotherapy, hormonal agents, and newer molecular targeted agents (3, 5).

In conclusion, we report a case of the desmoid tumor, sporadically arisen in retroperitoneum, with imaging features on CT. In our case, a huge left retroperitoneal mass showed heterogeneous enhancement, with infiltrative margin encasing splenic flexure colon and showed mass effect displacing spleen and left kidney. Although it is a pathologically benign tumor, it should be differentiated from malignant retroperitoneal tumors such as lymphoma or liposarcoma, due to its local aggressive features.

References