

Desmoid type fibromatosis of the distal pancreas: A case report

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A 23-year-old Korean female presented epigastric pain of two-months' duration. She had a laparoscopic ovarian cyst excision 8 months previously. Clinical examination was normal. An abdominal computed tomogram (CT) demonstrated a 10-cm solid mass in the distal pancreas, with signs of splenic artery and vein occlusion, gastric and transverse colon invasion. Operative findings showed a mass involving distal pancreas, invasive to the posterior wall of the antrum of the stomach and transverse colon and 4th portion of the duodenum without lymph node involvement. The surgery consisted of a distal pancreatectomy, splenectomy and combined partial resection of the stomach, transverse colon and 4th portion of the duodenum. The immunohistochemistry and histopathological features were consistent with a confirmed diagnosis of intra-abdominal desmoid type fibromatosis (DTF). The prognosis of pancreatic DTF is not known and she showed no recurrence or distant metastasis during a 3 year follow-up. Herein we report a rare case with an isolated, sporadic, and non-trauma-related DTF, located at the pancreatic body and tail. ([Ann Hepatobiliary Pancreat Surg 2021;25:276-282](#))

Key Words: Pancreatic desmoid type fibromatosis; Desmoid tumor; Aggressive fibromatosis

INTRODUCTION

Desmoid type fibromatosis (DTF), also known as a desmoid tumor, deep fibromatosis, or aggressive fibromatosis, is a rare neoplastic tumor arising from proliferation of mesenchymal stem cell progenitors.¹ It is locally invasive but without metastatic potential, and is slow-growing.² Although not metastatic, DTF tends to destroy surrounding tissues and organs and recurs frequently after radical resection. It can arise sporadically at any anatomical site throughout the body or be associated with familial adenomatous polyposis.³ DTF is categorized by location as extra-abdominal, abdominal wall, and intra-abdominal. Desmoid tumors account for 0.03% of all neoplasms and 3% of soft tissue tumors, and their annual incidence has been estimated to be 2-4 cases per million per year in the general people.⁴ Moreover, the incidence of sporadic intra-abdominal DTF is low (5%); fewer than 100 cases have been reported since the 1960s. Of these, intra-abdominal DTF of pancreatic origin is particularly rare, with only thirteen individual cases described in the English lit-

erature since the 1980s (Table 1).⁵⁻¹⁶ Herein we report a case with an isolated, sporadic, and non-trauma-related DTF, located at the pancreatic body and tail and invading the stomach, the transverse colon and the duodenum.

CASE

A 23-year-old Korean female presented to our surgical department with epigastric pain of two-months' duration. Her past surgical history included laparoscopic ovarian cyst excision 8 months previously. No history of prior abdominal trauma was noted. There was no family history to suggest a genetic hereditary disease. Clinical examination was normal. The liver function test was normal except elevated serum amylase 250 U/L (reference range: 28 to 100 U/L) and lipase level 708 U/L (reference range: 13 to 60 U/L). The levels of serum tumor markers were within normal limits: carcinoembryonic antigen, 0.6 ng/ml (reference range: 0 to 5 ng/ml); carbohydrate antigen 19 to 9, 6.27 IU/ml (reference range: 0 to 37 IU/ml) (Table 2). An abdominal computed tomogram (CT) demonstrated

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Table 1. Review of pancreatic desmoid type fibromatosis (DTF)^a

	Reference	Gender	Age (years)	Symptoms and signs	Lab findings	CT features	Tumor location	Size (cm)	Previous surgery	Surgery	Adjuvant treatment	Recurrence	Follow-up
01	Roggli et al. (1980) ⁵	M	0.3	Tachypnea, fever, anorexia, weight loss	NA	Solid	Diffuse	NA	None	Biopsy	None	NA	DOD 7 days
02	Bruce et al. (1996) ⁶	M	38	Abdominal pain	NA	Solid	Tail	5×4.5×2.1	Partial pancreatectomy	Resection	NA	No	ANED 24 months
03	Sedivy et al. (2002) ⁷	F	68	Weight loss, nausea	Normal	Solid	Head	1.5	Pancreatic biopsy	Resection	NA	NA	NA
04	Nursal et al. (2003) ⁸	F	25	Epigastric pain	NA	Solid	Tail	8.5×5.0	None	Biopsy	Symptomatic	NA	NA
05	Nursal et al. (2003) ⁸	M	39	Epigastric pain	NA	Solid	Tail	7.5×4.0	None	Biopsy	Symptomatic	NA	NA
06	Pho et al. (2005) ⁹	M	17	Epigastric pain	NA	Cystic	Tail	2.8×4.2	None	Resection	Sulindac, tamoxifen, methotrexate, vinblastine	Yes	AWD 24 months
07	Weiss et al. (2006) ¹⁰	M	63	Epigastric pain, abdominal fullness	Normal	Solid	Tail	6.5×5.3	Partial pancreatectomy	Resection	None	No	ANED 9 months
08	Amiot et al. (2008) ¹¹	F	51	Epigastric pain, weight loss	Normal	Cystic	Tail	6	None	Resection	None	No	ANED 12 months
09	Polistina et al. (2010) ¹²	M	68	None	Normal	Cystic	Tail	5	None	Resection	None	No	ANED 60 months
10	Jia et al. (2013) ¹³	M	41	Epigastric pain, weight loss	Normal	Cystic	Head	1.9	None	Resection	None	No	ANED 24 months
11	Xu et al. (2013) ¹⁴	M	17	Epigastric pain	Normal	Cystic solid	Body	8.6	None	Resection	None	No	ANED 20 months
12	Slowik-Moczydlowska et al. (2015) ¹⁵	M	13	Left upper quadrant abdominal pain	Normal	Cystic	Tail	10	None	Resection	None	No	ANED 19 months
13	Wang and Wong (2016) ¹⁶	F	57	Epigastric pain, weight loss	Normal	Solid	Head	10	None	Resection	Cox-2 inhibitor	No	ANED 24 months
14	Present study (2017)	F	22	Epigastric pain	Normal	Solid	Body, Tail	10	Ovarian cystectomy	Resection	None	No	ANED 8 months

^aAll cases were sporadic except for one patient with complication FAP reported by Pho et al.⁹ CT, computed tomography; NA, not available; ANED, alive with no evidence of disease; AWD, alive with disease; DOD, dead of disease

Table 2. Laboratory finding

Hgb (g/dl)	14.2 (12-16)	Sodium (mEq/L)	140 (135-145)
WBC (/μl)	5,700 (4,000-105,000)	Potassium (mEq/L)	3.5 (3.5-5.3)
PLT (/μl)	237K (150K-450K)	Chloride (mEq/L)	102 (98-110)
AST (U/L)	39 (7-38)	CA 19-9 (U/ml)	6.27 (0-37)
ALT (U/L)	79 (4-43)		
BUN (mg/dl)	12 (8-20)		
Creatinine (mg/dl)	0.9 (0.6-1.2)		
Total protein (mg/dl)	7.3 (6-8.1)		
Albumin (mg/dl)	4.7 (3.5-5.3)		
Amylase (U/L)	250 (28-100)		
Lipase (U/L)	708 (13-60)		

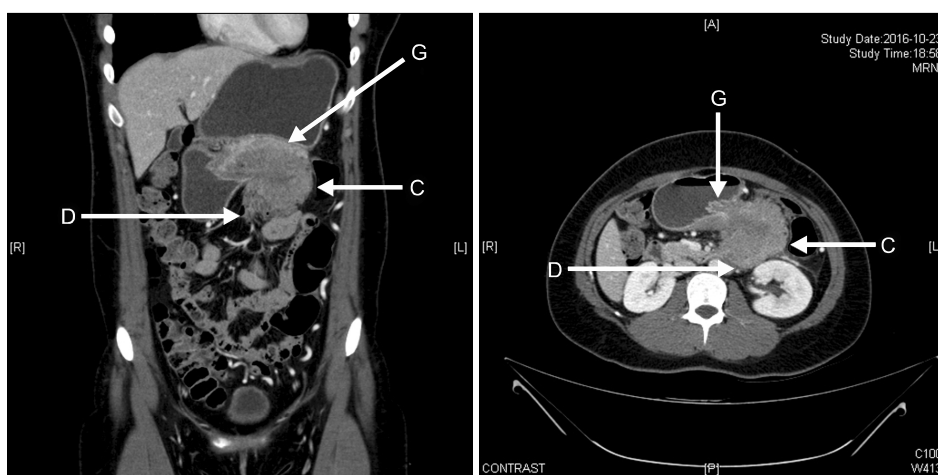


Fig. 1. Abdominal computed tomogram (CT) showed a 7.7-cm solid mass in the distal pancreas, with signs of splenic artery and vein occlusion, duodenum ‘D’, gastric ‘G’ and transverse colon invasion ‘C’.

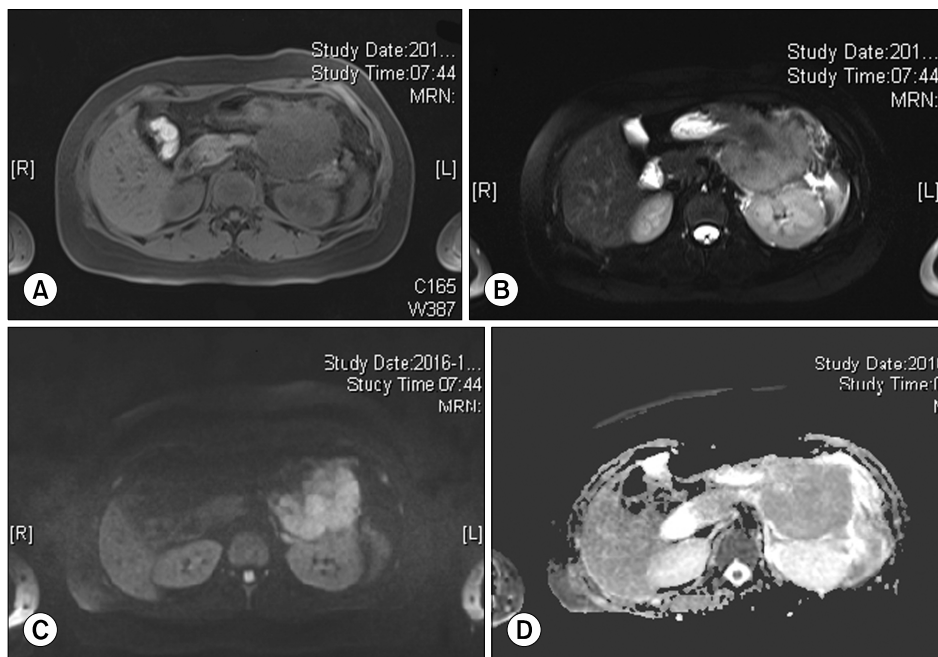


Fig. 2. A huge lobulated contour mass was found in the body and tail of the pancreas, showing low signal intensity on T1-weighted image (A) and slightly high T2-weighted image (B) with diffusion restriction (C) and delayed enhancement pattern (D).

a 10-cm solid mass in the distal pancreas, with signs of splenic artery and vein occlusion, gastric and transverse colon invasion (Fig. 1). The pancreatic mass was ill-de-

linedated with no evidence of metastasis. On MRI scan, a huge lobulated contour mass was found in the body and tail of the pancreas, showing low signal intensity on T1-

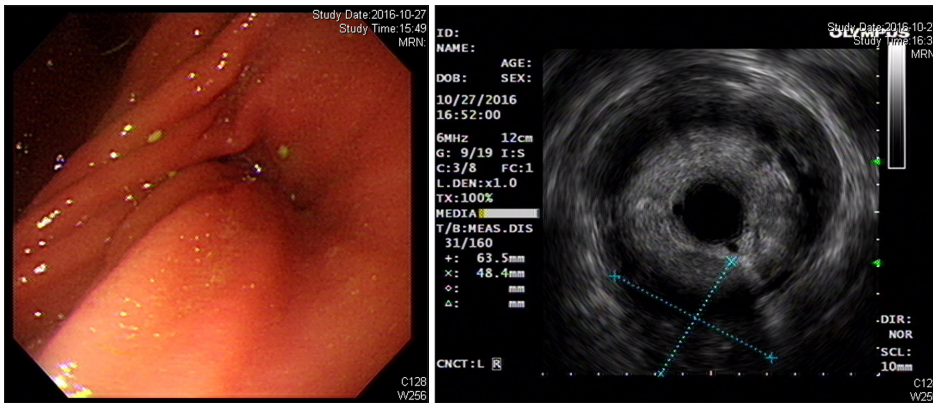


Fig. 3. Endoscopic ultrasonogram shows a huge soft tissue mass with no cystic portion but multiple echogenic foci in the distal pancreas.

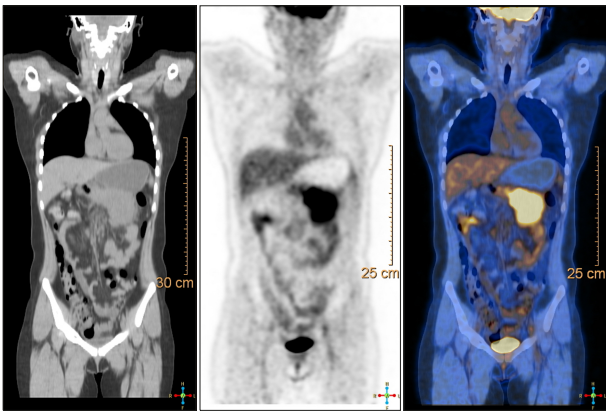


Fig. 4. PET-CT showed heterogeneous hypermetabolic tumor in left upper abdomen.

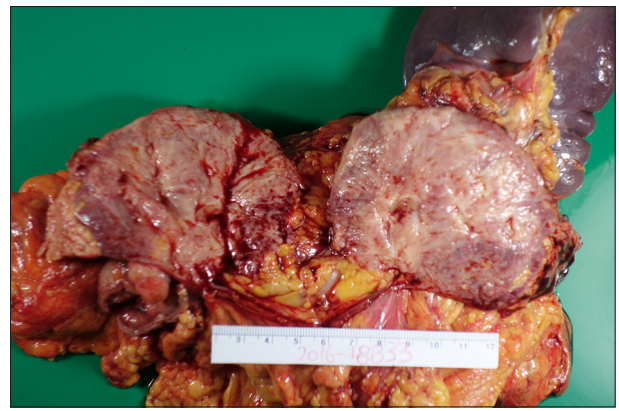


Fig. 5. Gross specimen of the pancreatic body solid mass measuring 10 cm in size.

and slightly high T2-weighted images with diffusion restriction and delayed enhancement pattern (Fig. 2). On EUS a showed an huge soft tissue mass with no cystic portion but multiple echogenic foci located in the distal pancreas (Fig. 3). PET CT showed heterogeneous hypermetabolic lesion in left quadrant abdomen (Fig. 4). Operative findings showed a mass involving distal pancreas, invasive to the posterior wall of the antrum of the stomach and transverse colon and 4th portion of the duodenum without lymph node involvement. The surgery consisted of a distal pancreatectomy, splenectomy and combined partial resection of the stomach, transverse colon and 4th portion of the duodenum. On gross pathology the tumor appeared as a grayish, white, dense and firm mass, without necrosis or hemorrhage (Fig. 5). On pathologic microscopic examination, the surgical margins were negative for a tumor. Histological sections showed a large number of spindle-shaped cells, with a regular nuclear pattern within a background of massive collagen fibers.

No marked cell death but mitosis 8/50 HPF was observed (Fig. 6). Immunohistochemical analysis revealed that the tumor cells were diffuse immune-positive for vimentin and β -catenin (Fig. 7), but immune-negative for smooth muscle actin, S-100, and CD34. The immunohistochemistry and histopathological features were consistent with a confirmed diagnosis of intra-abdominal DTF. The patient received no adjuvant therapy after surgery. She exhibited good health and complained of no discomfort. No local recurrence or distant metastasis in CT was found during a 3 year follow-up.

DISCUSSION

Pancreatic DTFs are extremely rare and encountered incidentally in surgical practice. To the best of our knowledge only thirteen previous cases of pancreatic DTFs have been reported within the last three decades in the English medical literature (Table 1).⁵⁻¹⁶ Of these cases, all oc-

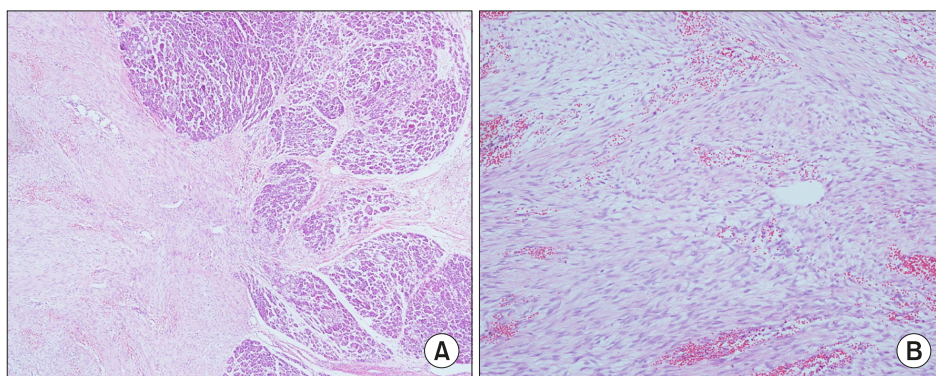


Fig. 6. H&E staining showed spindle cell mass in the pancreatic gland (A: H&E $\times 40$ magnification, B: H&E $\times 100$ magnification).

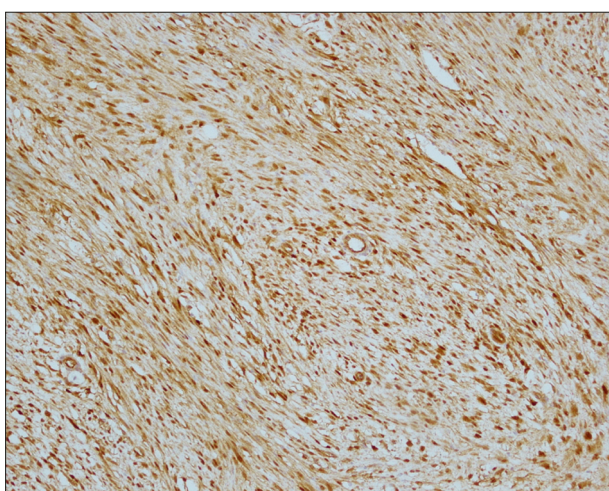


Fig. 7. Immunohistochemical staining showed in strong positive beta-catenin.

curred in middle-aged patients except for one infant.⁵ Six patients had cystic pancreatic tumors and one patient had concomitant FAP. Nine of the pancreatic DTFs were located in the pancreatic tail, three in the head, and one case was diffuse throughout the pancreas.

DTF etiology has not been well defined. A history of trauma to the site of the tumor, often surgical in nature, may be elicited in approximately 25% of cases.^{17,18} Because women are more prone to sporadic intra-abdominal DTF than men, (female-to-male ratio of 2:1 to 5:1), estrogen might have a pathogenetic role. However, for diagnosed pancreatic DTF in particular, the literature review and our report identified nine males and five females. FAP is frequently associated with intra-abdominal DTF, but only one pancreatic DTF patient was reported to have concomitant FAP in the current literature.⁹ A somatic mutation in the genes 3'-adenomatous polyposis coli (APC) or β -catenin (CTNNB1) is the most significant risk factor

for intra-abdominal DTF in FAP patients.^{19,20}

Previous abdominal surgery is said to initiate the proliferation of fibrous tissue and is therefore implicated in the occurrence and recurrence of intra-abdominal DTF. However, of the reported pancreatic DTF patients, only three had undergone previous abdominal surgery, one of whom developed a sporadic DTF at the pancreatic suture line,⁶ and another a DTF of the pancreatic stump following a distal pancreatectomy for pancreatic neuroendocrine tumors.¹⁰ Our case is sporadic and not associated with FAP but has the history of ovarian cystectomy previously and we do not assure whether the ovarian surgery develop pancreatic DTF.

The clinical presentation of most patients with DTF is usually asymptomatic and otherwise presents nonspecific abdominal pain. Our patient presented epigastric pain and had a mild elevation in amylase and lipase which suggest pancreatitis.

DTF is typically suspected on imaging studies, although a pathological specimen is required for definitive diagnosis. Computed tomography demonstrates a well-defined mass with variable enhancement pattern. The mass is typically iso-attenuating to hyper-attenuating to muscle on contrast-enhanced imaging.²¹ DTF on MRI typically returns low signal on T1-weighted sequences, with variable signal on T2-weighted sequences. EUS is very useful for visually defining the pancreatic tumor located in the pancreatic head, and to determine whether the major vessels and the duodenum are infiltrated by the tumor. Additionally, EUS allows fine needle aspiration biopsy for cytological examination of superficially located tumors.^{22,23}

Differentiating DTF from other types of soft tissue neoplasms may be difficult based on histological analysis alone. Expression was negative for CD34, CD117, S-100

Table 3. Diagnosis and treatment

Immunohistochemistry	Treatment
Mesenchymal cell (VIMENTIN) (+)	Watching and waiting
Marker for stromal cell CD117(-) CD34(-)	Radical resection: recurrence 19-77%
Neural cell S-100(-)	Non-steroidal anti-inflammatory drugs (NSAIDs)
Beta-catenin (+)	Hormonaltherapy non-steroidal benzothiophene (tamoxifen or raloxifen)
	Interferon
	Chemotherapy(vinbrastine, methotrexate, doxorubicin and dacarbazine)
	Imatinib

and desmin, which excluded a gastrointestinal stromal tumor, solitary fibrous tumor, schwannoma, leiomyoma and leiomyosarcoma, respectively.^{24,25} Recent efforts in immunostaining of intranuclear β -catenin and the gene responsible for its mutation proved efficient in discriminating DTF from other benign and malignant fibroblastic and myofibroblastic lesions (Table 3).²⁶

Multimodal therapy has been reported for the treatment of intra-abdominal DTF, including pancreatic DTF (Table 3). The prognosis of pancreatic DTF is not known.

CONCLUSION

Sporadic pancreatic DTF is a rare nonmetastatic and locally invasive soft tissue tumor. Diagnosis is based on medical imaging and confirmed via immunohistochemistry to differentiate DTF from other uncommon soft tissue tumors originating from the pancreas. Radical resection is recommended as first-line treatment for pancreatic DTF. Long-term follow-up studies are required to establish the prognosis of pancreatic DTF.

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AUTHOR CONTRIBUTIONS

Conceptualization: WYK. Writing - original draft: CGP, WYK. Writing - review & editing: CGP, YNL, WYK

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