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【 CASE REPORT 】

An Intra-abdominal Solid-cystic Desmoid That Emerged after Distal Gastrectomy

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Abstract:
Desmoid is a locally aggressive fibroblastic neoplasm, typically showing a heterogeneous solid mass, and its pathogenesis is multifactorial, including surgical scars. We herein report a rare case of an intra-abdominal desmoid, consisting of solid and cystic components covered with epithelial linings, that emerged after distal gastrectomy. The preoperative diagnosis was inconclusive, so laparotomy was performed. Histopathology of the solid component showed proliferating spindle cells, which were positive for beta-catenin in their nuclei. Clinicians need to bear in mind that desmoids can appear in a solid-cystic form, and immunostaining of beta-catenin should be applied for tumors that emerge around postoperative wounds.

Key words: post-operative wound, desmoid, beta-catenin, diagnosis

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Introduction

Desmoid is a locally aggressive fibroblastic neoplasm that lacks metastatic potential but tends to show local recurrence. The histology characteristically demonstrates infiltration and proliferation of elongated and uniform spindle cells within a collagenous stroma and vascular networks (1). It typically presents as a heterogeneous solid mass, depending on the amount of collagenous stroma, fibrosis, and cellular components (2), and cyst formation of desmoid is rarely seen.

We herein report a rare case of solid-cystic desmoid mimicking a gastrointestinal stromal tumor and pancreatic cystic lesion that emerged after distal gastrectomy.

Case Report

A 60-year-old asymptomatic man was referred to our hospital to investigate an abdominal cyst detected by abdominal ultrasound sonography at a medical checkup. He had a history of early-stage gastric cancer that had been treated by distal gastrectomy and Billroth-I reconstruction three years earlier and for which follow-up had been completed because of no lymph-vascular invasion and no lymph node metastasis. His family history was not remarkable, including no cases of hereditary disease.

No abnormal mass was palpable, and no tenderness was recognized in the abdomen. Laboratory data showed no abnormal finding, including serum tumor markers: white blood cell count, 6,070/mm^3; hemoglobin, 15.5 g/dL; C-reactive protein, 0.01 mg/dL; carcinoembryonic antigen (CEA), 1.3 ng/mL; and carbohydrate antigen 19-9 (CA19-9), 3 IU/mL. Computed tomography (CT) demonstrated a well-demarcated lesion consisting of a heterogeneous solid component and a round cystic component, 5 cm in size, widely attached to both the remnant stomach and the pancreas (Fig. 1a). Endoscopic ultrasonography (EUS) of the stomach showed a variable echogenic solid component and an anechoic cystic component (Fig. 2). The inside of the cyst was clear without debris. ^{18}F-Fluorodeoxyglucose-positron emis-

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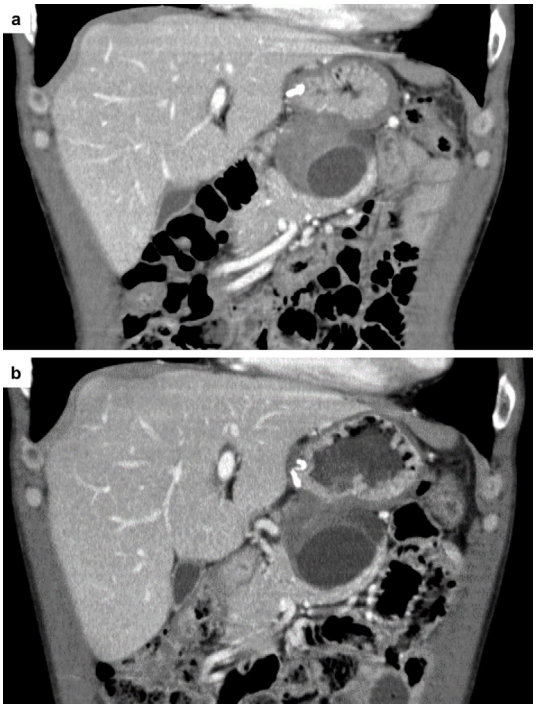

Figure 1. **Computed tomography (CT) of the lesion. a) The coronal view at the referral showed a lesion consisting of a cystic component and an ill-enhanced solid component, located between the stomach and the pancreas tail. b) The coronal view at four months of follow-up showed that the cystic lesion had increased in size.**

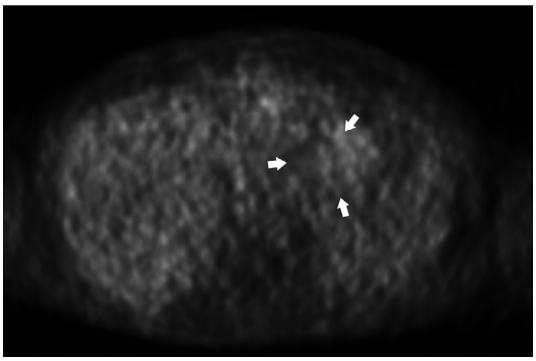

Figure 3. **^{18}F-Fluorodeoxyglucose-positron emission tomography (FDG-PET) demonstrating a weak uptake of FDG at the lesion (SUV_{max}: 3.1).**

sion tomography (FDG-PET) demonstrated a weak uptake of FDG at the lesion [maximum standardized uptake value (SUV_{max}): 3.1] (Fig. 3). As the post-gastrectomy CT images demonstrated no obvious lesion at the corresponding site (Fig. 4), a tumor with aggressive oncological potential was suspected, such as a gastrointestinal stromal tumor with cystic degeneration. Alternatively, an inflammatory condition was another candidate, such as a pseudocyst of the pancreas, although the patient did not complain of pain suggestive of pancreatitis. Careful follow-up was performed for four months, and CT showed the enlargement of the cystic lesion

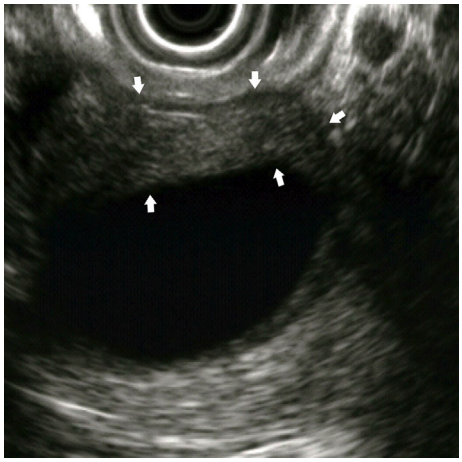

Figure 2. **Endoscopic ultrasonography (EUS) scan from the stomach showing a slightly echogenic solid component (arrows) and an anechoic cystic lesion.**

(Fig. 1b). EUS-guided fine-needle aspiration (FNA) was first considered to obtain a histological diagnosis, but the patient refused the procedure because of the risk of seeding neoplastic cells. He therefore underwent partial gastrectomy and distal pancreatectomy.

The mass adhered strongly to the gastroduodenal anatomy and pancreatic parenchyma, measuring 75 mm in size (Fig. 5a). The cyst contained clear serous liquid. Histopathology showed spindle cell proliferation in the background of abundant collagenous stroma (Fig. 5b) and epithelial cells lining the cyst (Fig. 5c). An immunohistochemical study showed the tumor cells to be positive for cytoplasmic muscle specific actin (MSA) and intranuclear beta-catenin (Fig. 5d) but negative for CD34 and S-100. The Ki-67 proliferation index was <1% in the tumor. The epithelial cells were positive for immunostaining of MUC5AC and MUC6 but negative for MUC2 and CDX2. The diagnosis was an intra-abdominal desmoid with a cystic component that had emerged after the initial gastrectomy. Both the stomach and the pancreatic parenchyma were largely involved with the tumor, so that the origin of the epithelia was difficult to conclude as either the stomach or pancreas (gastric-type epithelial metaplasia of the pancreatic epithelia).

He was discharged 14 days after the surgery. No recurrence has been recognized for four years.

Discussion

Desmoid is a rare tumor, estimated to occur in about 2-4 per million population per year and accounting for 3% of soft-tissue tumors (3). This tumor arises from anywhere in the body and is classified into abdominal wall (49%), extra-abdominal (43%), and intra-abdominal (8%) types, depending on its location (4). Intra-abdominal desmoid typically arises in patients with Gardner-type familial adenomatous polyposis (5). However, trauma (most often surgery) (6-8) is a common pathogenesis in addition to genetic, endocrine,

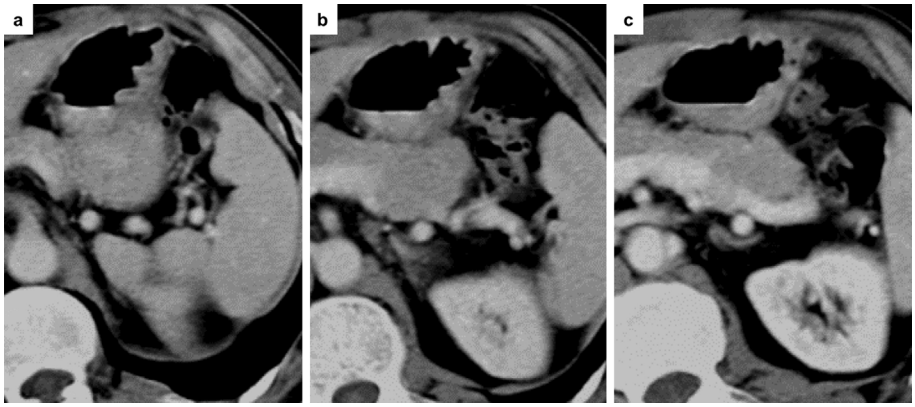

Figure 4. **Computed tomography (CT) before distal gastrectomy against early-stage gastric cancer three years earlier showing no cystic lesion.**

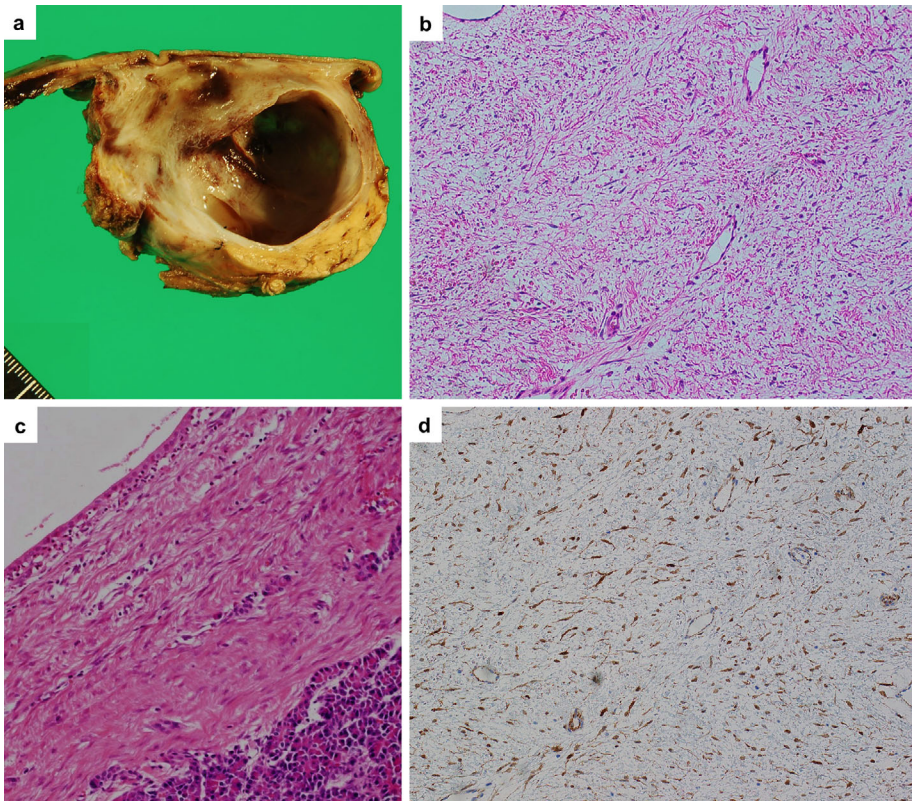

Figure 5. **Pathology of the resected tumor. a) A gross section showed that the tumor was widely attached to both the remnant stomach and the pancreas and contained a heterogeneous solid component and an adjacent cystic component. b) An Hematoxylin and Eosin staining section showed spindle cells, collagenous stroma, and vessels (×200). c) The cystic wall was covered by flattened columnar epithelium (×200). d) Immunostaining of beta-catenin showed intranuclear expression in the fibromatosis cells (×200).**

and other physical factors (9), so careful follow-up with clinical images is needed after surgery.

Intra-abdominal desmoid presents as a well-defined soft tissue mass with variable attenuation or as an ill-defined infiltrative lesion (10, 11). The differential diagnosis of desmoid tumor includes neuroendocrine tumor, lymphoma, retractile sclerosing mesenteritis, gastrointestinal stromal tumor, and mesenteric metastases (11). Since desmoids scarcely cause necrosis (12) and bleeding (13), cystic formation of desmoids has rarely been reported; indeed, only 13 cases have been published in the English literature, as shown in Table (origin: 12 pancreas and 1 mesenterium) (14-26). Due to its rarity, preoperative diagnoses of solid-cystic desmoids are very difficult, and these lesions are often misdiagnosed as malignant tumors, such as a pancreatic cancer (14-16), a neuroendocrine tumor with cystic

Table. **Reported Cases of Solid-cystic Desmoid (English Literatures, 2008-2018).**

No.	Ref.	Age	Sex	Surgical history	Location	Size (cm)	Preoperative diagnosis	Mucosal lining*
1	14	15	M	none	pancreas	20	pancreatic cancer	ND
2	15	11	M	ND	pancreas	10	pancreatic carcinoma	ND
3	16	68	M	none	pancreas	5	pancreatic cancer	ND
4	17	46	M	none	pancreas	21.5	pseudocyst, neuroendocrine tumor, mucinous cystadenoma	ND
5	18	19	F	ND	pancreas	37	pseudopapillary tumor	(+)
6	19	21	F	ND	pancreas	6	mucinous cystadenoma	ND
7	20	41	M	neck lipoma resection	pancreas	1.9	pancreatic cystic neoplasm	(+)
8	21	17	M	ND	pancreas	8.6	solid pseudopapillary neoplasm	(+)
9	22	51	F	hysterectomy	pancreas	6	mucinous cystadenocarcinoma	ND
10	23	58	F	ND	mesenterium	4.5	teratoma, lymphangioma	ND
11	24	19	F	none	pancreas	4	ND	ND
12	25	20	F	ileocolectomy	pancreas	7.5	ND	ND
13	26	13	M	ND	pancreas	10	splenic cyst	ND

F: female, M: male, ND: not described, *mucosal lining inside of the cyst.

changes (17), a cystic neoplasm of the pancreas (18-22), and an inflamed teratoma (23). These cystic formations were thought to be caused by pancreatic duct dilation associated with tumor infiltration, tumor infection, or rupture. Information on the previous surgery was available for three cases (Table). As the desmoids in all three cases developed away from the operation site, these cases were thought to be sporadic desmoids. Three of the 13 reported cases with cystic desmoids histologically demonstrated epithelial lining inside of the cysts, probably originating from the pancreatic epithelia entrapped by the stromal infiltration (Table) (15, 17, 18). Clinicians need to consider the possibility of desmoids potentially accompanied by a cystic component.

The development of desmoids is based on the Wnt/β-catenin-dependent pathway, which controls key developmental gene expression programs. Nuclear staining of beta-catenin was detected in 80% of cases of sporadic desmoid tumor, and nuclear immunopositivity is supportive for desmoid fibromatosis in the differential diagnosis of spindle cell lesions (27). The current stromal tumor was negative for CD34, which is often positive in a variety of fibrosarcomas, angiosarcomas, and epithelioid sarcomas, and it was also negative for S-100, which is frequently expressed in malignant peripheral nerve sheath tumors, rhabdomyosarcomas, and clear cell sarcomas (27). In the current case, the final diagnosis of solid-cystic desmoid was made based on these pathological findings and the clinical course, although the origin of the tumor and epithelial cells was unable to be concluded as either the stomach or the pancreas. We refrained from performing EUS-FNA, as it carried a risk of causing leakage of the cyst content, potentially leading to peritoneal dissemination or gastric seeding (28). However, by retrospectively reviewing the CT images (Fig. 1a) with the possible development of postoperative desmoids in mind, we concluded that EUS-FNA could be performed safely for the solid component. By further applying beta-catenin immunohistochemistry on the obtained samples, an accurate diagnosis of a desmoid could have been made without surgery.

Unfortunately, we were unable to reach the accurate diagnosis preoperatively. However, the tumor was fully resected by surgery, and the patient has remained free from recurrence for more than four years. This outcome was compatible with the desmoid tumor staging of the current case (Stage I: low risk of recurrence, in asymptomatic cases with a non-growing tumor, <10 cm in size) (29). However, if the accurate diagnosis could have been obtained by EUS-FNA preoperatively, we could have avoided unnecessary lymph node dissection and performed minimally invasive surgery.

Conclusion

Desmoid is a locally aggressive fibroblastic neoplasm typically showing a heterogeneous solid mass but rarely accompanied by cystic changes and/or epithelial lining. Clinicians need to consider the possibility of desmoid occurrence near postoperative scars and perform beta-catenin immunohistochemistry for the differential diagnosis.

The authors state that they have no Conflict of Interest (COI).

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