INTRODUCTION

Desmoids are rare tumors, with a prevalence of 2.4-4.3 cases per million people each year. In one retrospective analysis of more than 50,000 patients admitted for neoplastic disease, only 17 desmoid tumors were found (0.03%) [1]. The term desmoid is derived from the Greek “desmos”, meaning band-like. The first description of a desmoid tumor was credited to McFarlane in 1832, when he observed a fibrous tumor in a woman after childbirth [2]. Intra-abdominal desmoids involve the mesentery and retroperitoneum and usually occur in association with familial adenomatous polyposis (FAP) or Gardner’s syndrome [3]. FAP and Gardner’s syndrome are autosomal dominantly inherited disorders arising from mutation of the adenomatous polyposis coli (APC) gene [4]. Several studies have shown that about 12.4-29% of individuals affected with FAP develop desmoids. Those not coupled with FAP or Gardner’s syndrome are termed sporadic and are much less common. Intra-abdominal desmoids involving the pancreas are extremely rare, and only eleven cases have been reported. Herein, we report the first case of a sporadic desmoid tumor involving the pancreatic tail, presenting as a cystic lesion. It is very difficult to distinguish this tumor from a primary pancreatic cystic lesion.

CASE REPORT

A 77-year-old female patient, who had a history of myasthenia gravis, diabetes mellitus, and hysterectomy for an unknown cause, visited a physician for abdominal pain. Physical examination was unremarkable without fever, nausea, vomiting, or rebounding tenderness. She denied any history of trauma. Routine blood investigation was within normal limits without elevation of the leucocyte count, serum lipase level, or serum levels of tumor markers (carcinoembryonic antigen and carbohydrate antigen 19-9). On abdominal ultrasonography (Fig. 1), a unilocular, ovoid, anechogenic cystic lesion with a regular wall and a maximum diameter of 6.7 cm was found in the pancreatic tail. On abdominal computed tomography (Fig. 2), a well-defined homogeneous ovoid cystic lesion, measuring 8.8 × 6.2 × 7.8 cm³, was observed in the pancreatic tail without a prominent papillary or solid component. No prominent peripancreatic fat infiltration was noted. After contrast...
administration, the lesion showed mural enhancement. The spleen was not involved in the tumor, but the splenic artery and vein were compressed by the lesion. Besides, the lesion also showed an indistinct margin to the stomach. Owing to the imaging characteristics, a pancreatic mucinous cystadenoma or pseudocyst was suspected initially. The patient received surgical treatment and the operation proceeded smoothly without immediate complications.

On gross examination (Fig. 3a), there was a soft tissue tumor measuring 7.5×6.6 cm\(^2\) in dimension with pancreas involvement. The tumor had a gray-white and firm cut surface. There was a unilocular and well-demarcated cystic lesion measuring 7.5×6.6 cm\(^2\) with green brownish watery fluids within the part of the pancreas involving the tumor. No papillary or solid component was observed on the cystic wall. Microscopically (Fig. 3b), the tumor comprised elongated, slender, spindle-shaped cells of a uniform appearance arranged in interlacing fascicles and focal storiforms separated by collagen bundles infiltrating into the pancreatic parenchyma. Immunohistochemical studies of the tumor cells showed β-catenin (+), ALK (-), CD34 (-), p53 (-), and...
bcl-2 (-). The final pathologic diagnosis was desmoid-type fibromatosis.

After the operation, the patient recovered well without any major complaints or evidence of tumor recurrence.

DISCUSSION

The term fibromatoses describes rare forms of connective tissue cellular dysplasia with monoclonal fibroblastic proliferation in an abundant collagen extracellular matrix. They form invasively-growing masses and frequent recur locally, but lack metastatic potential. Local aggressive behavior can lead to mechanical complications such as compression or obstruction of blood vessels, the gastrointestinal tract, or the urinary tract. In a reported case, tumor strangulated blood vessels causing life-threatening hemorrhagic shock [5]. The etiology of desmoid tumor development may be multifactorial. Genetic abnormalities, trauma (including surgery), endocrine, and physical factors are implicated in their pathogenesis, because abdominal wall tumors often arise in young women after childbirth or in a post-operation scar [6]. However, the exact mechanism is still not completely understood.

Fibromatoses can be studied as two groups, superficial (fascial) and deep (musculoaponeurotic), based on their location. Deep fibromatoses, also termed desmoid tumors, are now more commonly referred to as aggressive fibromatoses. In 2002, the World Health Organization designated the term desmoid-type fibromatosis for deep fibromatoses [7]. Desmoid-type fibromatoses of the abdomen can be further categorized as abdominal wall fibromatosis or intra-abdominal fibromatosis. Intra-abdominal fibromatosis has no gender or age predilection and has been reported in patients between 14 and 75 years of age [7].

In the abdomen, desmoid tumors rarely involve the pancreas, and only 11 cases have been reported (Table 1). In the reported 11 cases, 7 patients had solid desmoid tumors [1, 2, 8-11], 3 patients had solid tumors with intratumoral cystic components [3, 6, 12] and only 1 patient had a cystic desmoid tumor [4]. In the 3 patients who had solid desmoid tumors with intratumoral cystic components, the initial diagnoses were mucinous cystadenocarcinoma, cystic pancreatic cancer, and pancreatic adenocarcinoma, respectively. These cases confirmed the difficulties in diagnosis of incidental cystic pancreatic tumors.

Generally speaking, cystic pancreatic lesions can be categorized into three groups: pseudocysts, common cystic neoplasms (intraductal papillary mucinous neoplasm, serous/mucinous cyst neoplasm), and uncommon cystic neoplasms (solid and pseudopapillary tumor, tumor with cystic degeneration). When a cystic pancreatic lesion is detected, it is most important to decide whether the lesion is a pseudocyst, accounting for 70-90% of such lesions, or another cystic neoplasm. Our patient had no history of pancreatitis, alcohol abuse, or biliary stone disease. There was no evidence of acute or chronic pancreatitis on CT imaging. Although pseudocyst may not be the first consideration, the possibility of pseudocyst could not be completely excluded. Besides, the cystic lesion had no internal septa, solid component, central scar, or mural calcification. In addition, there was no prominent dilatation of the pancreatic duct, atrophy of the distal pancreatic parenchyma, or peripancreatic desmoplastic reaction on CT imaging. Intraductal papillary mucinous neoplasm, serous

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age/Gender</th>
<th>Sporadic form</th>
<th>Tumor form on CT</th>
<th>Location in pancreas</th>
<th>Size</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 [8]</td>
<td>4 months/Male</td>
<td>Yes</td>
<td>Solid</td>
<td>Diffuse</td>
<td></td>
</tr>
<tr>
<td>2 [9]</td>
<td>2 months/Female</td>
<td>Yes</td>
<td>Solid</td>
<td>Head</td>
<td>4.0 cm</td>
</tr>
<tr>
<td>3 [1]</td>
<td>38 years/Male</td>
<td>Yes</td>
<td>Solid</td>
<td>Tail</td>
<td>5.0 cm</td>
</tr>
<tr>
<td>4 [10]</td>
<td>68 years/Male</td>
<td>Yes</td>
<td>Solid</td>
<td>Head</td>
<td>1.5 cm</td>
</tr>
<tr>
<td>5 [11]</td>
<td>25 years/Female</td>
<td>Yes</td>
<td>Solid</td>
<td>Tail</td>
<td>8.5 cm</td>
</tr>
<tr>
<td>6 [11]</td>
<td>39 years/Male</td>
<td>Yes</td>
<td>Solid</td>
<td>Tail</td>
<td>7.5 cm</td>
</tr>
<tr>
<td>7 [4]</td>
<td>17 years/Male</td>
<td>No (FAP)</td>
<td>Cystic</td>
<td>Tail</td>
<td>4.0 cm</td>
</tr>
<tr>
<td>8 [2]</td>
<td>63 years/Male</td>
<td>Yes</td>
<td>Solid</td>
<td>Tail</td>
<td>1.5 cm</td>
</tr>
<tr>
<td>9 [3]</td>
<td>51 years/Female</td>
<td>Yes</td>
<td>Solid-cystic</td>
<td>Tail</td>
<td>6.0 cm</td>
</tr>
<tr>
<td>10 [12]</td>
<td>68 years/Male</td>
<td>Yes</td>
<td>Solid-cystic</td>
<td>Tail</td>
<td>5.0 cm</td>
</tr>
<tr>
<td>11 [6]</td>
<td>11 years/Male</td>
<td>Yes</td>
<td>Solid-cystic</td>
<td>Tail</td>
<td>10 cm</td>
</tr>
<tr>
<td>Our patient</td>
<td>78 years/Female</td>
<td>Yes</td>
<td>Cystic</td>
<td>Tail</td>
<td>8.8 cm</td>
</tr>
</tbody>
</table>
cyst neoplasm, or a tumor with cystic degeneration were excluded preliminarily. The lesion in our female patient was a unilocular cystic neoplasm. Mucinous cystadenoma was suspected preoperatively, and the patient underwent distal pancreatectomy with splenectomy. However, the diagnosis of desmoid-type fibromatosis was confirmed by positive β–catenin immunohistochemical analysis.

In the 11 reported cases, the only patient who had a pure cystic tumor was a 17-year-old male [4]. He had been clinically and genetically confirmed to be affected with FAP, an autosomal dominantly inherited disorder arising from mutation of the \( \text{APC} \) gene. Loss of \( \text{APC} \) gene function leads to accumulation of β–catenin in the cytosol and nucleus. Gardner syndrome is another inherited disorder with colonic findings of FAP, multiple osteomas, thyroid malignancies, epidermoid cysts, fibromas, sebaceous cysts, and desmoids. In a review article, the author pointed out the mutations in either the \( \text{APC} \) or β–catenin genes are likely to be a major driving force in the formation of these desmoid tumors [13]. However, despite continued surgical and medical treatments, recurrent desmoid tumor was still observed on follow-up CT imaging of this patient, and he was the only non-sporadic case in the reported 11 patients. In contrast, our patient was not affected with FAP or Gardner syndrome. The desmoid tumor involving the pancreas of our patient can be considered as a sporadic lesion. To our knowledge, never has a sporadic desmoid tumor involving the pancreas been reported in the literature presenting as a cystic lesion. This case indicates that desmoid-type fibromatosis may be considered as a possible etiology of cystic pancreatic lesions.

Regarding management, the optimal protocol for the treatment of desmoid tumors has not been well established. Medical treatment including non-steroidal anti-inflammatory drugs (sulindac or indometacin), anti-fibrotic agents, and anti-estrogens (tamoxifen) may be quite effective for intra-abdominal desmoids and lead to improvement in up to 50% of cases [4]. In addition, radiation therapy can be used as an adjunct to surgery, and cytotoxic chemotherapy may be used for unresectable tumors unresponsive to other

Figure 3. a. On gross examination, there was a soft tissue tumor (T), measuring 7.5×6.6 cm\(^2\) in dimension, with pancreas involvement. S: spleen. b. Light microscopic picture (Hematoxylin and eosin; original magnification x100). The tumor comprised elongated, slender, spindle-shaped cells (thin arrows) of a uniform appearance arranged in interlacing fascicles and focal storiforms separated by collagen bundles (background) infiltrating into the pancreatic parenchyma (arrows). c. Immunohistochemical study of the tumor cells showed immunoreactive for β-catenin (thin arrow). The pathologic diagnosis was desmoid-type fibromatosis.
Desmoid mimicking cystic pancreatic lesion

relatively benign treatments. 
Post-operative recurrence occurs in up to 90% of FAP cases with associated intra-abdominal desmoid tumors, and in 10% of sporadic cases [6]. Although surgical resection is the main therapy for extra-abdominal and abdominal wall desmoids, it must be kept only for rapidly growing and life-threatening tumors. Observation alone is recommended for static lesions. However, surgical resection was performed in 8 of the 11 patients with pancreatic desmoid tumors, and our patient also received surgical resection of the tumor. Post-operative recurrence occurred only in one patient who had a past history of FAP [4]. As regards our patient, there was no prominent recurrence evidence to be found in the follow-up imaging.

In conclusion, desmoid tumors rarely involve the pancreas, and the diagnosis is difficult to make if they do. Although they extremely rarely involve the pancreas, the possibility of desmoid-type fibromatosis should be kept in mind for an incidental cystic pancreatic lesion.

REFERENCES