

[CASE REPORT]

A Rare Case of Pancreatic Endometrial Cyst and Review of the Literature

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Abstract:

Pancreatic cysts include a variety of benign, premalignant, and malignant lesions. Endometrial cysts in the pancreas are extremely rare lesions that are difficult to diagnose before surgery. We report the case of a 26-year-old patient with a recent episode of left abdominal pain who presented with a large cyst in the pancreatic body. Laboratory results showed white blood cell and C-reactive protein elevation, whereas the patient's tumor marker levels were within the normal range. Distal pancreatectomy with splenectomy was performed. The final histopathological examination confirmed the presence of endometriotic cysts within the pancreas. Only 13 cases of endometriotic cysts of the pancreas have been previously reported. The preoperative diagnosis is challenging, and most patients undergo pancreatic resection because of suspected neoplasms. This case report reviews previous studies and discusses the clinicopathological features, pathogenesis, and appropriate treatment for pancreatic endometrial cysts.

Key words: pancreas, endometrial cyst

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Introduction

Endometriosis, in which endometrial tissue grows outside the uterus and attaches to various organs in the body, is a relatively common condition, affecting approximately 5-10% women of reproductive age. Extragenital manifestations of endometriosis occur in up to 40% of patients with pelvic endometriosis, which most commonly manifests in the intestines, urinary tract, abdominal wall/peritoneum, and thoracic cavity. In contrast, intrapancreatic endometrial cysts are a rare entity, with a literature review reporting 13 cases of pancreatic endometrial cysts (1-12). All patients with pancreatic endometrial cysts underwent surgical resection because malignant or premalignant lesions could not be ruled out. We herein report the case of a patient with a cyst in the pancreatic body. A pathological examination revealed that

endometriotic tissue lined the cystic wall.

Case Report

A 26-year-old woman with an irregular menstrual cycle was hospitalized because of acute left abdominal pain, which was not related to her menses. She had no history of biliary disease, alcoholism, previous surgery, or traumatic abdominal injury. Her family history was also unremarkable. On admission, a laboratory examination revealed the following: white blood cell count, 12,700/mm³; hemoglobin level, 14.4 g/dL; platelet count, 427,700/mm³; total protein level, 8.1 g/dL; albumin level, 5.2 g/dL; total bilirubin level, 0.8 mg/dL; aspartate aminotransferase level, 22 IU/L; alanine aminotransferase level, 27 IU/L; alkaline phosphatase level, 245 IU/L; γ -glutamyl transferase level, 34 IU/L; amylase level, 77 U/L; C-reactive protein level, 0.69 mg/dL; carci-

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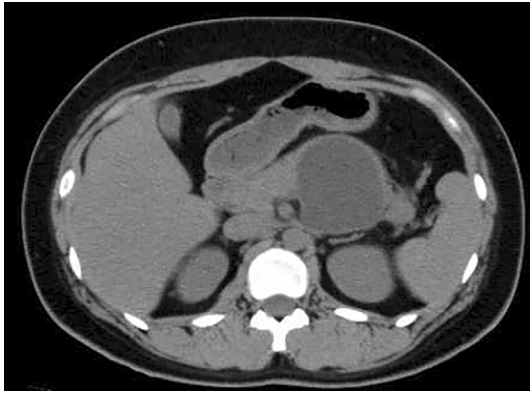


Figure 1. Computed tomography without contrast enhancement revealed a cystic lesion measuring 7 cm in diameter in the pancreatic body.



Figure 2. Magnetic resonance imaging (MRI; T2WI) revealed a cystic lesion without cyst in the cyst structure, and dilatation, stenosis, or displacement of the main pancreatic duct.



Figure 3. Endoscopic ultrasonography (EUS) showed a hypo-echoic mass of 6.7×5.3 cm in size without a septum, no solid elevated lesions, and no areas of focal hemorrhage.

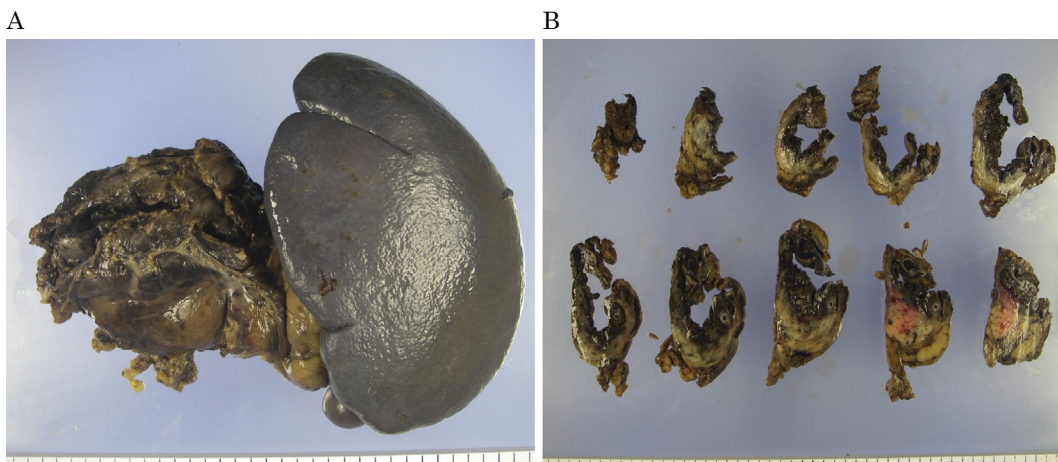


Figure 4. A: Laparoscopic distal pancreatectomy with splenectomy. B: The gross examination of the surgical specimen revealed a circumscribed cyst measuring 7.0×5.0×2.5 cm that was located in the pancreatic body. The cyst wall appeared smooth, trabeculated, and gray-brown with areas of focal hemorrhage. No papillary projections were observed.

noembryonic antigen (CEA) level, 0.6 ng/mL; CA19-9, 6.7 U/mL; DU-PAN-2, <25 U/mL; and s-pancreatic antigen-1 level, <10 U/mL. Computed tomography without contrast enhancement revealed a cystic lesion measuring 7 cm in diameter in the pancreatic body (Fig. 1). Magnetic resonance imaging (MRI) revealed a cystic lesion without any dilatation, stenosis, or displacement of the main pancreatic duct (Fig. 2). Endoscopic ultrasonography (EUS) showed a hypo-echoic mass of 6.7×5.3 cm in size without a septum and no solid papillary lesions (Fig. 3). After a preoperative diagnosis of mucinous cystic adenoma, laparoscopic distal pancreatectomy with splenectomy was performed (Fig. 4A).

The gross examination of the surgical specimen revealed a circumscribed cyst measuring 7.0×5.0×2.5 cm that was located in the pancreatic body. The cyst wall appeared smooth, trabeculated, and gray-brown with areas of focal hemorrhage. No papillary projections were observed (Fig. 4B).

A histological examination revealed that the cyst was cov-

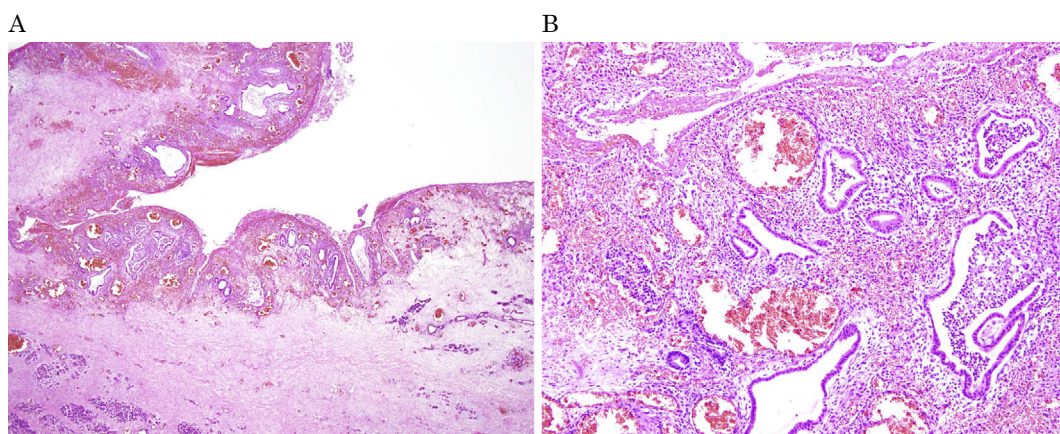


Figure 5. A histological examination revealed that the cyst was covered with fibrous tissue containing elongated endometrial glands consisting of cuboidal and columnar cells with basal nuclei and amphophilic cytoplasm.

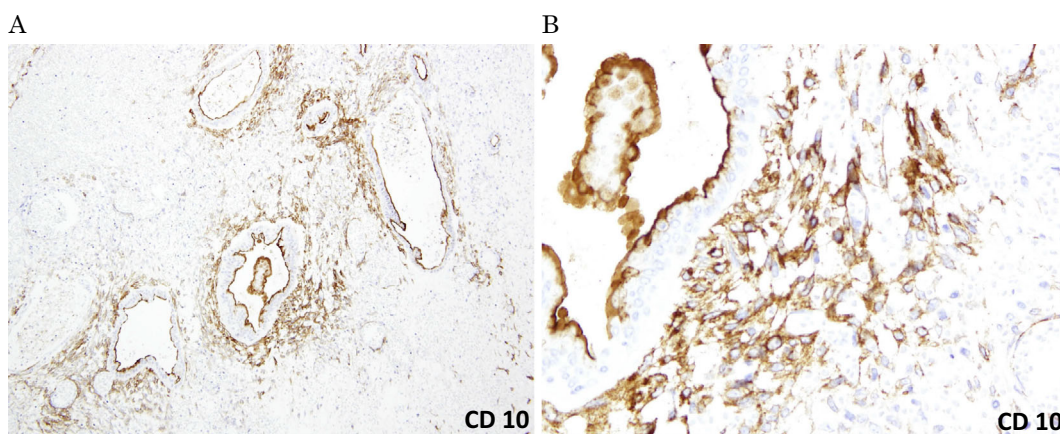


Figure 6. Immunohistochemistry revealed that the spindle cell stroma was positive for CD10.

ered with fibrous tissue containing elongated endometrial glands consisting of cuboidal and columnar cells with basal nuclei and amphophilic cytoplasm (Fig. 5). The glands were admixed with well-vascularized spindle cell stroma containing multiple areas of focal hemorrhage and hemosiderin-laden macrophages.

Immunohistochemistry revealed that the spindle cell stroma was positive for CD10 (Fig. 6), estrogen receptor-related protein (Fig. 7C and D), and nuclear progesterone receptor protein (Fig. 7B), and negative for inhibin (Fig. 7A). Taken together, these features were consistent with a final diagnosis of cystic endometriosis in the pancreatic body.

The postoperative course was uneventful, and the patient did not develop any complications, including pancreatic fistula.

Discussion

Endometriosis, defined as the presence of endometrial glands and stroma outside the uterus, typically manifests as recurrent pain that is commonly associated with the men-

strual cycle. Several underlying mechanisms for the pathogenesis of endometriosis have been proposed, including the direct extension of the endometrial tissue to the neighboring compartments, menstrual regurgitation, lymphatic/vascular spread, and celomic metaplasia (13). The most common sites of endometriosis are the pelvic organs; however, endometriosis of the upper abdominal organs has also been described (10). Conversely, pancreatic endometrial cysts are extremely rare; our search of the PubMed database revealed only 13 cases that had been reported in the English literature since 1984, when the first case was reported by Marchevsky et al. (1). The characteristics of the 14 patients are shown in Table. All reported cases involved female patients (mean age at presentation, 38.5 years; range, 21-68 years). The mean cyst size was 76.9 mm (range, 30-165 mm). In the current case, the patient did not have a gynecologic history of endometriosis, but had a history of cesarean section during her childbirth. She had never required any medical or surgical therapy for abdominal pain. In less than half of the reported cases of pancreatic endometrial cyst, pain was correlated with the menstrual cycle; there was only one case in which endometriosis was the presumed di-

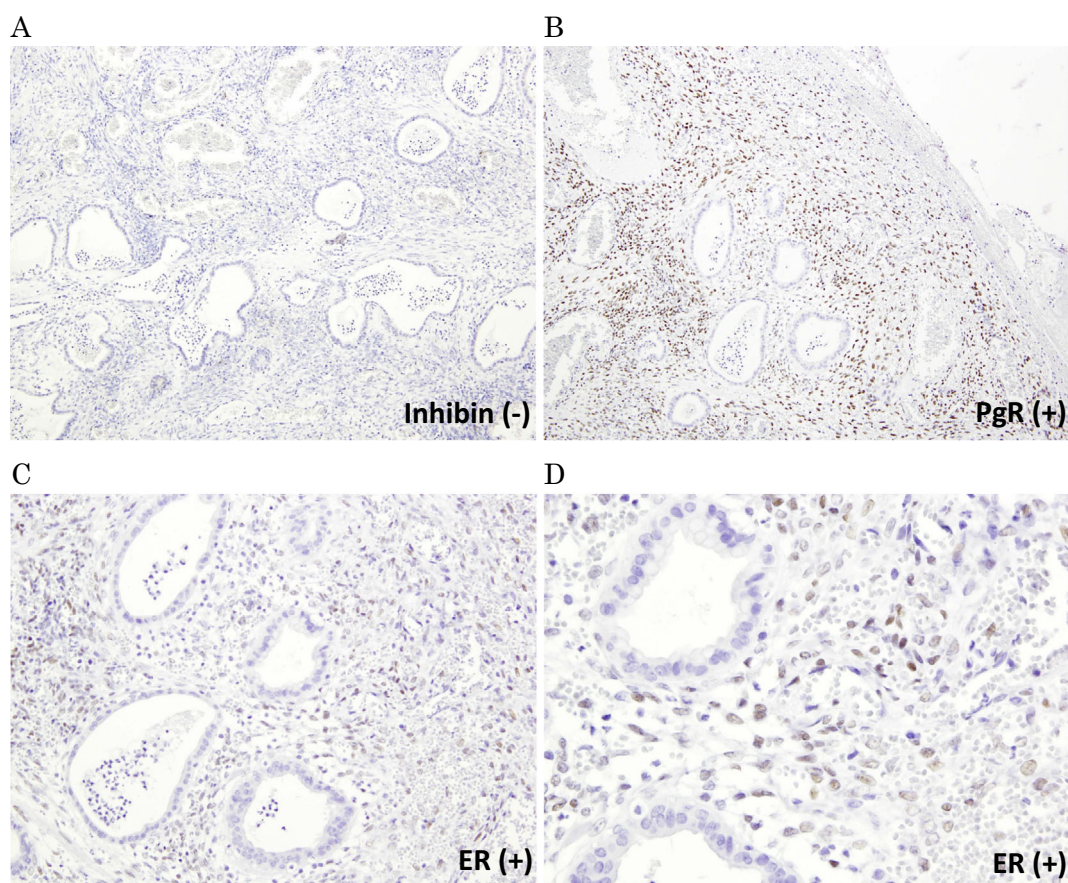


Figure 7. A-D: The cyst was positive for estrogen receptor-related protein (C, D), and nuclear progesterone receptor protein (B), and negative for inhibin (A).

Table. The Characteristics of the 14 Patients.

Reference	Case	Symptom	Size of cyst (cm)	Location of cyst	Treatment	Outcome
1	36F	epigastric pain	8×6	tail	distal pancreatectomy	alive
2	40F	left upper quadrant pain	8×6	tail	distal pancreatectomy	unknown
3	28F	epigastric pain	10×4.5	body	distal pancreatectomy	unknown
4	47F	epigastric pain	5×4.5	tail	distal pancreatectomy	unknown
5	21F	epigastric pain	4.5×4	body	distal pancreatectomy	alive
6	34F	left upper quadrant pain	8×8	tail	distal pancreatectomy	unknown
7	23F	epigastric pain	10×10	tail	distal pancreatectomy	unknown
8	35F	epigastric pain	3×2.5	body	distal pancreatectomy	alive
9	43F	epigastric pain	unknown	tail	distal pancreatectomy	unknown
10	32F	epigastric pain	13×10	tail	distal pancreatectomy	unknown
10	63F	unknown	5×5	body	distal pancreatectomy	unknown
11	68F	left upper quadrant pain	3×2.4	tail	distal pancreatectomy	alive
12	43F	epigastric pain	16.5×11.1	tail	distal pancreatectomy	alive
our case	26F	left upper quadrant pain	6.1×5.7	body	distal pancreatectomy	alive

F: female

agnosis (9). The clinical findings from previous reports are similar to those in the present case. The patient did not relate any of her abdominal symptoms to her menstrual cycle or hormones, and the patient had no previous episodes of pancreatitis. The initial assessment of the lesion suggested MCN of the pancreas due to its location in the pancreatic body. MCN defines lesions with malignant potential that are predominantly observed in middle-aged women (14). The

typical imaging features of pelvic endometriosis with hemorrhage are hyperintense foci on T1-weighted MRI with or without fat saturation; in the absence of bleeding, these lesions are hypointense on T1- and T2-weighted MRI. There are few reports on endometriotic cysts in the liver or pancreas; hence, no typical imaging features have been established (11). In the current patient, the cyst was 12 cm in its largest dimension with loculation. EUS with fine needle as-

piration is a reasonable approach when imaging findings are equivocal. In a previous report (12), surgical resection was indicated due to an elevated CEA level. An elevated cyst fluid CEA level (940 ng/mL) was only reported in one other case of pancreatic endometrial cyst (10). Thus, pancreatic endometrial cysts can present elevated cyst CEA levels. However, in Japan, EUS-FNA is not commonly applied to pancreatic cystic lesions based on the fear of needle tract seeding, which has been reported to cause pseudomyxoma in cases of MCN, or IPMN. Due to the diagnostic similarities between rare endometrial cysts and mucinous cystic lesions, resection is considered to be the best diagnostic and therapeutic approach.

The composite theory of histogenesis of endometriosis combines the implantation, vascular/lymphatic metastasis, and direct extension theories (15). In a previous report, the pancreas was located in the retroperitoneum and had no clear connection with the peritoneal cavity, suggesting that endometrial fragments could be transported into distant sites through lymphatic or blood vessels (1). Because of the variable manifestations of endometriosis, it is likely that several mechanisms are involved in its pathogenesis. Further clinical and experimental studies are needed to evaluate the pathogenesis underlying the occurrence of endometriosis in unusual sites.

Given that the malignant potential of the rarely reported pancreatic cysts is variable, there are currently no reliable preoperative detection methods for endometrial disease of the pancreas, and the diagnosis and treatment of endometriosis at unusual sites remains rather difficult due to its extreme rarity.

As this case illustrates, the differential diagnosis of cystic lesions of the pancreas in women of reproductive age should include endometriosis.

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

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