Macrocystic Serous Cystadenoma of the Pancreas in a Young Patient Resembling a Pseudocyst: Case Report and Literature Review

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Macrocystic serous cystadenoma is an unusual and essentially benign pancreatic tumor. Ages of reported cases are usually 60 years and over, with a mean age of 54 years. Herein, we report on a 26-year-old man who presented with upper abdominal pain. A cystic lesion in the mid-portion of the pancreas was revealed by abdominal computed tomography, and a pseudocyst was suspected. A distal pancreatectomy was performed with a splenectomy due to intractable abdominal pain and being unable to rule out to be a mucinous cystic neoplasm, which has a malignant potential. The histopathological diagnosis was macrocystic serous cystadenoma of the pancreas. To our knowledge, this patient is the youngest person to present with such tumor. Clinical and pathologic features including complete immunohistochemical studies are presented, and we review the relevant literature. *(Chang Gung Med J 2003;26:602-6)*

Key words: **macrocystic serous cystadenoma, pancreas.**

Serous cystadenoma of the pancreas is a cystic epithelial neoplasm composed of serous-type, glycogen-rich epithelial cells. It is an uncommon and essentially benign pancreatic tumor. The majority of serous cystadenomas are microcystic. The macrocystic subtype of serous cystadenoma has been reported only on rare occasions.[1,2] The radiologic features of a macrocystic serous cystadenoma (MSC) may resemble a pseudocyst or a mucinous cystadenoma.[1,3-6] Thus, it is often difficult to make a correct diagnosis. MSC usually occurs in patients over 60 years preoperatively, with a mean age of 54 years.[7] Herein, we report on an MSC in a 26-year-old male patient, with a preoperative diagnosis of pancreatic pseudocyst by imaging study. To our knowledge, our patient is the youngest person to present with such tumor in the literature.

CASE REPORT

A 26-year-old man was admitted to our institution in June 2001 with a 5-month history of a cystic lesion in the body of the pancreas. During a prior admission for a crushing injury to his right hand, abdominal ultrasonography and computed tomography were performed to investigate his complaint of recurrent upper abdominal discomfort. This led to the discovery of a cystic lesion in the body of the pancreas. He had experienced intermittent upper abdominal pain after that. The patient had no history of alcohol consumption, abdominal trauma, or pancreatitis. The physical examination was unremarkable. Laboratory data, including hematology, general biochemistry, and urinalysis, were within normal limits. Values of the carcinoembryonic antigen (CEA; 1.69 ng/ml) and CA19-9 (< 2 mg/ml) were

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also within normal limits (normal values, < 5 ng/ml and < 37 mg/ml, respectively). Abdominal ultrasonography showed a pancreatic cystic lesion at the junction of the body and tail, measuring 2.3 cm in diameter. An abdominal CT scan showed a well-defined hypodense cystic lesion of 2.5×2.5×2.5 cm in the mid-portion of the pancreas (Fig. 1). Percutaneous needle aspiration revealed only foamy histiocytes; no epithelial cells were present. While the clinical diagnosis was a pancreatic pseudocyst as suggested by the abdominal CT, a mucinous neoplasm with a definite malignant potential could not be ruled out. Therefore, as the patient was complaining of intractable abdominal pain, he underwent an exploratory laparotomy with a distal pancreatectomy and splenectomy with the intent to establish a pathologic diagnosis.

Macroscopically, there was a well-defined, 2.5-cm unilocular cyst in the pancreas, which was filled with a clear watery fluid, with no solid part identified. There was no communication between the cyst and pancreatic duct. Microscopically, the cystic wall lining was composed of simple cuboidal or flattened epithelial cells with clear cytoplasm. The nuclei were rounded to oval in shape, uniform, and centrally located (Figs. 2, 3). Periodic acid-Schiff (PAS) staining was focally positive in the cytoplasm of the epithelium, and was negative after diastase digestion. Mucin staining was negative. Immunohistochemically, the epithelium was positive for CA19-9, CAM5.2, and epithelial membrane antigen (EMA), and negative for CEA and neuron-specific enolase (NSE). Therefore, the tumor was diagnosed to be a macrocystic serous cystadenoma of the pancreas. The postoperative course was uneventful. The patient was well for 1 year after surgery and is currently being followed-up through our outpatient department.

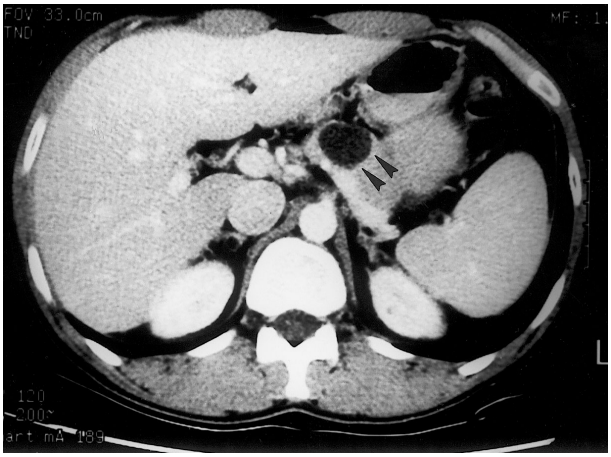

Fig. 1 Abdominal CT with contrast showing a well-defined hypodense, cystic lesion in the mid-portion of the pancreas.

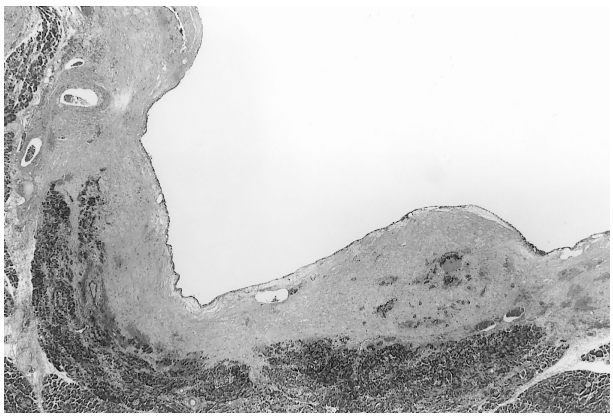

Fig. 2 A unilocular cyst within an unremarkable pancreas. (H&E, ×25)

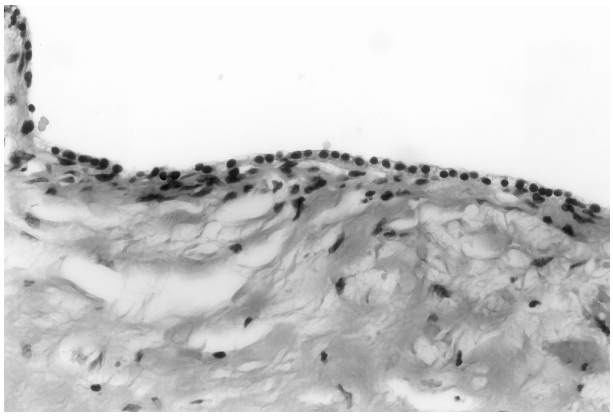

Fig. 3 Cyst lined by simple cuboidal serous epithelial cells, with clear cytoplasm and uniform, rounded nuclei. (H&E, ×400)

DISCUSSION

Serous cystadenoma of the pancreas is an uncommon and essentially benign tumor. It consists of a predominantly microcystic architecture lined by a layer of cuboidal epithelium with rounded and uniform nuclei, and clear cytoplasm containing a large

amount of glycogen.[8] The current case is of a macrocystic variant of a serous cystadenoma. It has been reported only on rare occasions. These cases and our own are summarized in Table 1. This entity was first reported by Lewandrowski et al.[1] The lining epithelial cells generally have the same histological features as a microcystic serous cystadenoma, but the tumor is composed of only a few relatively large cysts or even a unilocular cyst. It is considered that an MSC is a subtype of serous cystadenoma of the pancreas. There is a slight female preponderance. Ages of reported cases are usually 60 years and over, ranging from 35 to 69 years, with a mean of 54 years. The youngest patient was 35 years old.[4] Our case, a 26-year-old man, was much younger than those described in the literature. The presenting symptom of our patient was intermittent abdominal pain, similar to other reported cases.

The macroscopic features of a macrocystic serous cystadenoma can be unilocular or oligocystic with each cyst having a diameter of 2.5-8 cm. Our case presented a 2.5 cm unilocular cyst. Microscopic examination revealed simple cuboidal or flattened serous epithelial cells with clear cytoplasm and rounded nuclei.[1-5] These cells were PAS positive and were digested by diastase. In addition, the mucicarmine stain was negative, excluding the possibility of a mucinous cystic neoplasm. The immunohistochemical study showed that the tumor cells were positive for CA19-9, CAM5.2, and EMA and negative for CEA and NSE. These findings were compatible with those reported by Spertic et al.[5]

The radiological features of MSC may resemble those of a pseudocyst or a mucinous cystadenoma; the latter has the risk for malignant progression, whereas the prognosis for an MSC is much better. Therefore, it is important to make a correct diagnosis before surgery so that appropriate surgical management can be followed. However, it seems difficult to make the distinction by imaging studies preoperatively, and almost all cases are diagnosed after surgery by histologic examination. Procacci et al.[12] reviewed 30 cases of serous cystadenoma of the pancreas based on their imaging findings and concluded that it was impossible to diagnose macrocystic serous cystadenoma correctly by preoperative imaging. Lewandrowski et al.[1] also found that the radiological features of their 5 cases of macrocystic serous

Table 1. Summary of Reported Cases of Macrocystic Serous Cystadenoma of the Pancreas

No.	Age	Gender	Symptoms	Tumor location	Tumor size (cm)	Preoperative diagnosis	Operative methods	Outcome	Reference no.
1	66	F	Abdominal fullness	Head and body	8×6.5×4	MCN	SP	No recurrence at 6 months to 5 years (cases 1-5)	1
2	46	M	ND	Body	2.5×2.5×2.5	Tumor or pseudocyst	DP		1
3	69	M	Dull epigastric pain	Head	2.5×2.5×2.5	MCN or pseudocyst	WP		1
4	60	F	Upper abdominal discomfort	Head	8×8×7	MCN	SP		1
5	67	F	Upper abdominal pain	Body and tail	3.5×3.5×3.5	MCN	DP		1
6	35	F	Upper abdominal pain	Body	ND	MCN	E	ND	4
7	48	F	Abdominal pain	Tail	5×5×5	MCN	DP	No recurrence at 2.5 years	3
8	47	F	ND	Body	5×4.5×3	MCN	DP	ND	9
9	63	F	Upper abdominal movable mass	Body	6×5×3	Mucinous cystadenoma	E	No recurrence at 1 year and 7 months	10
10	42	F	No symptoms	Head	4.8×4×3.8	MCN	PPPD	No recurrence at 1 year	11
11	26	M	Upper abdominal pain	Body and tail	2.5×2.5×2.5	Pseudocyst or MCN	DP	No recurrence at 1 year	Our case

ND, not documented; MCN, mucinous cystic neoplasm; SP, subtotal pancreatectomy; DP, distal pancreatectomy; WP, Whipple procedure; E, enucleation; PPPD, pylorus preserving pancreaticoduodenectomy.

cystadenoma were indistinguishable from those of mucinous cystic neoplasms, and when unilocular, could be confused with pseudocysts. As in our case, a preoperative diagnosis of a pseudocyst was made according to the radiological features.

The role of preoperative percutaneous needle aspiration of the pancreatic cyst remains controversial. Nguyen et al.[13] encountered a case of an MSC of the pancreas, and they made a correct diagnosis using preoperative percutaneous needle aspiration. They saw small monolayered sheets of benign cubic epithelial cells in the needle aspirates. Borgne et al.[14] reported that in their series, cytologic analysis was acellular in 50% of cases, and did not provide a reliable means of distinguishing serous cystadenomas from mucinous cystic tumors. The aspiration cytology of our patient was similar to the result of Borgne et al.[14] It revealed only scattered foamy histiocytes, and no epithelial cell was present. Therefore, we believe that preoperative percutaneous needle aspiration has limited utility since the aspirated material is usually insufficient for a diagnosis. In addition, the procedure may also lead to tumor cell dissemination, unless a benign diagnosis can be made with reasonable certainty. Thus, an intraoperative frozen section remains the only means to establish a diagnosis before the operation.

In our opinion, when a unilocular pancreatic cyst is found, MSC, as well as pseudocyst and mucinous cystic neoplasms, should be considered in the differential diagnosis, especially in those patients without a history of pancreatitis or gallstones, and whose related serum tumor markers are not elevated. An intraoperative frozen section should be performed so that appropriate surgery can be given. It is important to acknowledge that an MSC can occur in young people such as in the present case, although most patients are over 60 years of age. Therefore, when clinical findings characteristic for pancreatitis are not present, even in a young patient, the possibility of an MSC should still be considered.

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發生在一個年輕人像假囊腫的胰臟巨囊型漿液性的囊腺瘤：病例報告及文獻回顧

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巨囊型漿液性的囊腺瘤 (macrocystic serous cystadenoma) 是一種非常罕見而且完全良性的胰臟腫瘤。大部分被報告過的案例年齡都在60歲以上，平均年齡是54歲。這裡我們報告一位26歲的年輕男性，臨床表現是上腹部疼痛。在腹部電腦斷層檢查發現在胰臟的中間部分有一個囊腫，術前被懷疑是一個假囊腫 (pseudocyst)。病人因爲持續的腹部疼痛及無法完全排除是一個具惡性潛質的黏液性囊腫的可能性，因此接受了遠端胰臟切除手術合併脾臟切除。病理學上的檢查證實是巨囊型漿液性的囊腺瘤。就我們所知，在文獻上這個病人是罹患這個腫瘤最年輕的一例。本文包括了臨床、病理的表現，以及完整的免疫組織化學染色的結果，並針對胰臟巨囊型漿液性的囊腺瘤做一文獻回顧。(長庚醫誌 2003;26:602-6)

關鍵字： 巨囊型漿液性的囊腺瘤，胰臟。

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