JOP. J Pancreas (Online) 2009 May 18; 10(3):332-334.

MULTIMEDIA ARTICLE - Clinical Imaging

Enormous Serous Microcystic Adenoma of the Pancreas

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Summary

Serous microcystic adenomas or serous cystadenomas are rare and account for 1-2% of the exocrine neoplasms of the pancreas. Recently, due to the improvements in imaging techniques, they have been identified more frequently. We present the case of a patient in whom a serous microcystic adenoma, 26 cm in diameter, was found incidentally in the pancreas, the largest reported in the literature thus far. Pylorus-preserving pancreaticoduodenectomy was performed, combined with a right hemicolectomy because of the tumor localization. Despite their size, the resection of serous microcystic adenomas, even of that magnitude, is operatively efficient and should always be taken into consideration when examining therapeutic options.

Currently, there are five recognized subtypes of serous cystic neoplasms of the pancreas: serous microcystic adenoma, serous oligocystic ill-demarcated adenoma, solid serous adenoma, von Hippel-Lindau-associated cystic neoplasm and serous cystadenocarcinoma [1]. Serous microcystic adenomas are benign tumors of the pancreas and are also known as glycogen-rich cystadenomas or serous cystadenomas [2]. Two-thirds of the patients present with symptoms, such as abdominal pain, abdominal mass, nausea, and vomiting or weight loss. The remaining one-third of the tumors are discovered incidentally on routine physical examination or autopsy. Serous microcystic adenomas make up approximately 1-2% of all exocrine pancreatic tumors and 25% of cystic neoplasms. They usually occur in elderly women in the seventh to eighth decade of life (range 34-91 years; mean age 66 years) [3, 4]. There is also a female predominance (70% of the tumors occur in females). Most of them are located in the body or tail of the pancreas; however, they are also described in the head of the pancreas. In previously reported cases, their size ranged from 1 to 25 cm, with an average of 6-10 cm [2, 3, 4, 5]. Despite their rarity, serous microcystic adenoma of the pancreas should be kept in mind when making a differential diagnosis of cystic pancreatic lesions. In our case, we describe the largest serous microcystic adenoma reported in the world literature, 26 cm in diameter.

Received February 9th, 2009 - Accepted April 17th, 2009

Key words Adenoma; Cystadenoma, Serous; Pancreas; Pancreatic Cyst; Pancreatic Neoplasms

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Document URL http://www.joplink.net/prev/200905/25.html

Case Report

A palpable mass in the right upper abdominal quadrant was incidentally detected in a 66-year-old female during a routine physical examination. An abdominal ultrasound examination was performed and a cystic tumor with solid parts involving the head of the pancreas was observed. She was referred to our clinic for further investigation and treatment. The patient was asymptomatic on admission. There was no history of laparotomy, abdominal trauma, gallstones, pancreatitis or alcohol consumption. Physical examination was unremarkable despite the palpable tumor mass. A complete blood count, biochemical parameters and CA 19-9 were all within the reference range as well. A

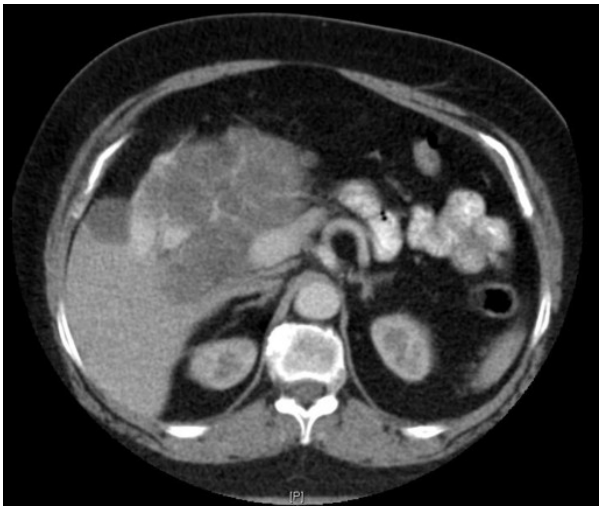

Image 1.

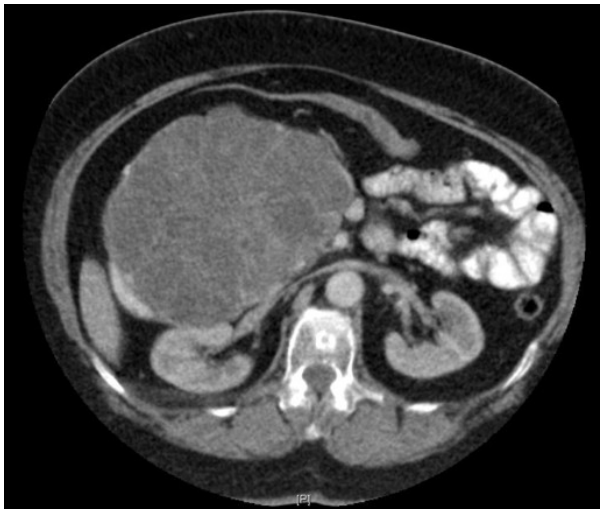

Image 2.

contrast-enhanced dynamic CT scan of the patient demonstrates an enormous 26x20x15 cm mass with lobulated contours and containing multiple tiny cysts separated by thin septa localized in the head of the pancreas reached the aortal bifurcation (Images 1, 2, and 3). The splenic artery, superior mesenteric artery and superior mesenteric vein were normal with no signs of invasion.

A pylorus-preserving pancreaticoduodenectomy was performed, combined with a right hemicolectomy since the tumor surrounded the colonic vessels. At surgery, a cystic mass, 26 cm in diameter, showing an expansive growth surrounding the right colonic vessels and compressing the vena cava inferior was identified in the pancreatic head (Image 4). Histologically, the multilocular cyst was lined with single-layered cuboidal cells without atypia (Image 5). No infiltration of the tumor into the interstitium was observed. The bowel wall was not infiltrated by the tumor. Immunohistochemically, there was a strong positive

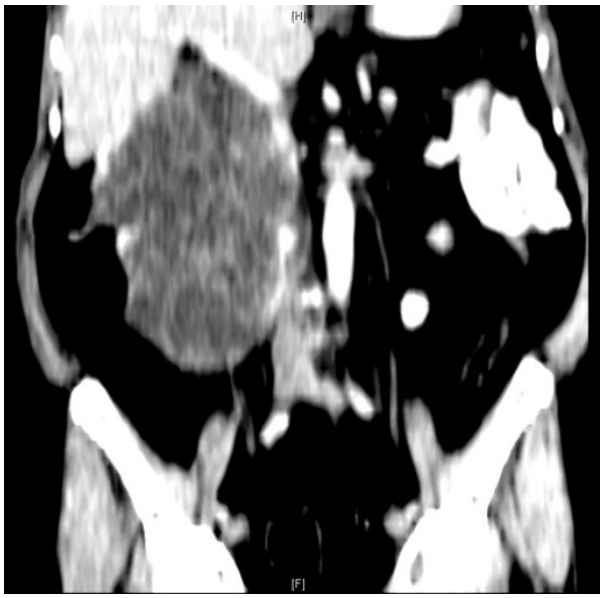

Image 3.

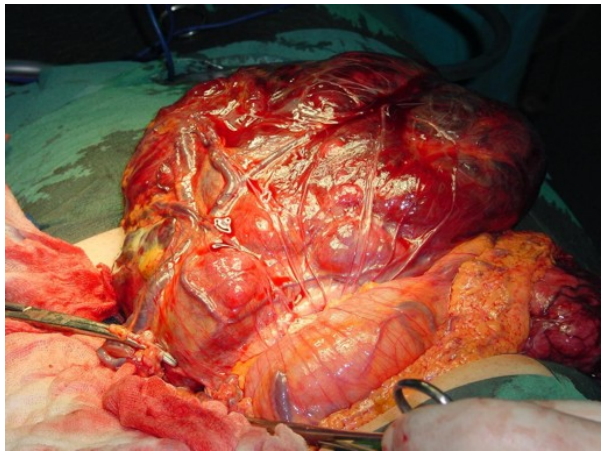

Image 4.

expression of cytokeratin 18. The cysts were lined by a flattened columnar epithelium showing granular PAS-positive staining.

Based on these findings, a diagnosis of serous microcystic adenoma of the pancreas head was reached. The patient is doing well one year after surgery. No postoperative complications occurred.

Discussion

Serous microcystic adenomas are benign cystic tumors of the pancreas. They usually appear as a large single asymptomatic mass in elderly women and are often incidentally detected during abdominal imaging for unrelated reasons. Grossly, they are large and multiloculated, and range in size from 1 to 25 cm [2, 3, 4].

A diagnosis of serous microcystic adenoma relies mainly on imaging techniques such as US, CT and EUS. While they may occur in any portion of the pancreas, they are mainly located in the body and tail [3, 4, 5, 6]. Except for centers having great experience in pancreas imaging, an accurate preoperative diagnosis of serous microcystic adenoma can be made only in about 40% of the patients [4, 7].

On CT scan, they can have a varied appearance. The morphological patterns of serous microcystic adenoma

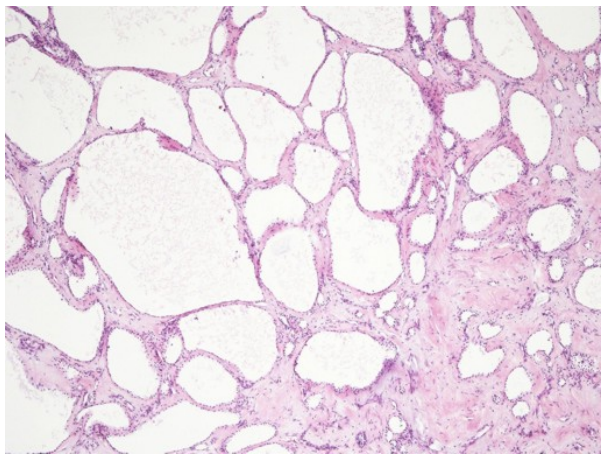

Image 5.

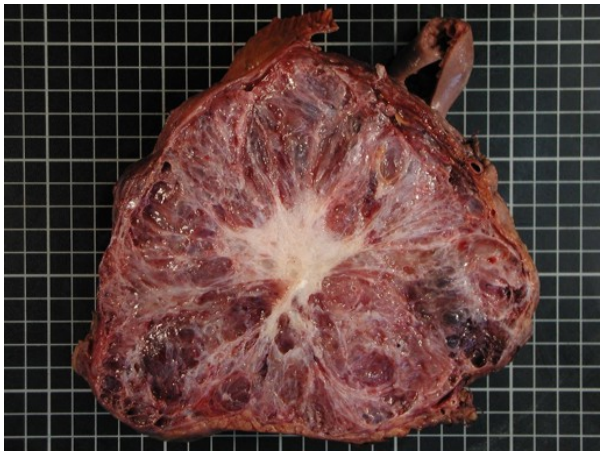

Image 6.

can be classified as polycystic, honeycomb and oligocystic [5, 8]. Serous microcystic adenomas are typically solitary but may be multiple in case of von Hippel-Lindau syndrome, causing an appearance of disseminated involvement. They can also appear as a solid mass and can overlap with a variety of other types of pancreatic pathologies. When the distinction is less clear, fine-needle aspiration of cystic lesions for cytological and biochemical analysis of the fluid can be performed and may help in reaching a diagnosis [1, 2, 3, 4]. Grossly, they are well demarcated and have a central scar (Image 6). Immunohistochemically, they are positive for cytokeratins 7, 8, 18 and 19 [2, 9, 10].

Symptoms, accuracy of the preoperative diagnosis, safety of resection and risks of conservative management play a role in deciding how to treat serous microcystic adenomas. The majority of symptomatic patients undergo pancreatic resection. Drainage of these tumors is inappropriate. Tumors of the body and tail of the pancreas require distal pancreatectomy. The surgical procedure of choice for tumors in the head or uncinate process of the pancreas remains a Whipple resection. Patients with cystic lesions of undetermined nature undergo resection because there is a risk of mistaking a mucinous tumor for serous microcystic adenoma [2, 4, 11].

Since the potential of malignant transformation is very low, the prognosis of serous microcystic adenomas of the pancreas is considered to be excellent. Therefore, some authors suggest that serous microcystic adenomas may be conservatively managed and closely followed-up in asymptomatic patients without vascular obstruction, in elderly patients or in those who have high operative risks [3, 4]. But conservative management has the risk of progressive growth of the tumor and development of complications, such as hemorrhage, erosion into the adjacent structures and gastrointestinal or biliary obstruction. In our case, although the patient was asymptomatic, she was operated on because of the enormous size of the tumor and not because of the preoperatively established diagnosis. Surgical excision of the mass should be performed when the tumor is symptomatic or very large. An intraoperative pathological consultation may obviate the need for more radical surgery which could reduce morbidity in elderly and high-risk patients [3, 4, 6, 10, 11, 12, 13].

In conclusion, serous microcystic adenomas of the pancreas have recently been identified more frequently due to advances in imaging techniques. Differentiation from other cystic tumors as well as from non-neoplastic cysts is very important because of the difference in their management. Finally, the resection of serous microcystic adenomas, even of extreme magnitude, is operatively efficient and should always be taken into consideration when examining therapeutic options.

Conflict of interest The authors have no potential conflicts of interest

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