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Multiple synchronous serous cystadenomas of the pancreas: uncommon CT and MRI findings

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Abstract
Serous cystadenomas (SCAs) of the pancreas are benign cystic tumors. Although still controversial, asymptomatic SCAs, in contrast to their mucinous counterparts, can be treated conservatively. This attitude is mostly defendable for lesions of the pancreas head or multiple lesions distributed throughout the entire pancreas, as their surgical resection is still associated with significant morbidity. Thus, correct diagnosis is essential, and this relies on radiological and biological characteristics. Asymptomatic multiple SCAs are rare. Most cases described in the literature are either symptomatic, degenerated, or both. We present a case of huge (>10 cm), multiple asymptomatic SCAs, not associated with von Hippel-Lindau disease, involving the entire pancreas. The patient has been followed up for 3 years and remains asymptomatic. Tumor markers were within normal ranges. On abdominal computed tomography (CT) and magnetic resonance imaging (MRI), the lesions showed pathognomonic characteristics of SCAs, and their uncommon dimensions and number remained stable over time. 18-Fluorodeoxyglucose positron emission tomography (PET) scan performed at 3-year follow-up did not show hypermetabolic lesions.

Key words Pancreas · Cystic tumor · Serous cystadenoma · Pancreatectomy

Introduction

Serous cystadenomas of the pancreas (SCAs), also called microcystic adenomas, are benign cystic tumors and must be differentiated from their potentially malignant mucinous counterparts. Although still controversial, asymptomatic SCAs, in contrast to mucinous cystadenomas, can be treated conservatively.[1–4] Most SCAs appear as single lesions of the pancreas. Multiple pancreatic SCAs are described in von Hippel-Lindau disease.[5] Multifocal SCAs in patients not affected by this inherited disease are uncommon. We present a case of huge multiple asymptomatic SCAs of the pancreas in a patient treated expectantly with a 3-year follow-up.

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Case report

A 63-year-old woman, with a medical history of appendectomy, was admitted for acute intestinal obstruction requiring an emergent laparotomy. Small-bowel obstruction was found to be related to adhesions in the right iliac fossa secondary to the previous appendectomy. During the exploration phase, a huge retro-peritoneal mass was fortuitously discovered. It appeared to be firm on palpation, and was polylobar, arising from the entire pancreas, without gross invasion of adjacent structures and obviously not related to the intestinal obstruction. The small bowel was successfully freed from adhesions as therapeutic management, but no attempt was made to dissect the pancreatic tumor. Retrospectively, no symptoms related to the abdominal mass were found. Abdominal computed tomography (CT) and magnetic resonance imaging (MRI) were performed postoperatively.

Nine multilocular cystic lesions with innumerable small cysts of less than 2 cm in diameter, giving a typical honeycomb pattern, were observed on CT. Six lesions, the largest having a diameter of 11 cm, were located in the body and tail of the pancreas, with smaller satellite cysts. Three smaller cysts were visible in the head of the pancreas. A central fibrous scar with calcification foci was clearly identified as a hypointense area in most of the tumors, The walls of the cysts were thin (Fig. 1).

MRI confirmed the presence of these nine masses, which appeared as mixed microlacunar and macrolacunar cystic tumors, homogeneously hyperintense on T2-

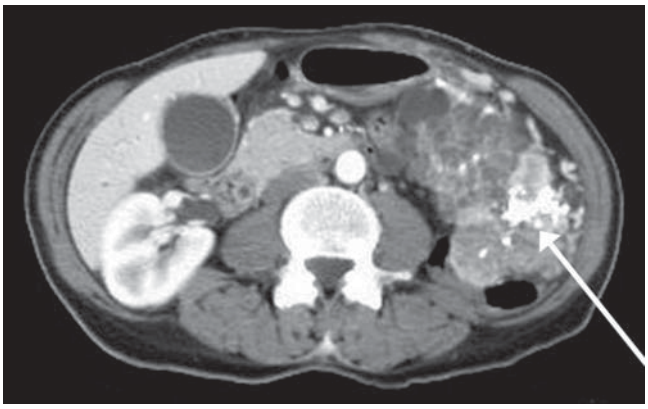

Fig. 1. Contrast-enhanced abdominal computed tomography (CT), showing a huge multilocular cystic mass of the pancreas. Honeycomb appearance and calcifications within a central fibrous scar (*arrow*) are visible. The thin cystic walls are enhanced by the intravenous contrast

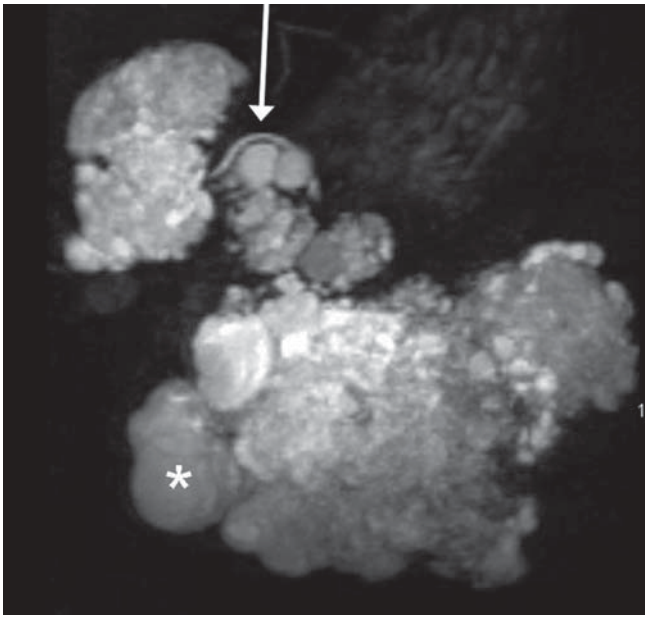

Fig. 2. T2-weighted magnetic resonance (MR) image, showing the huge microcystic lesions surrounded by larger cysts (>2 cm; *asterisk*) The pancreatic duct does not show any signs of obstruction (*arrow*)

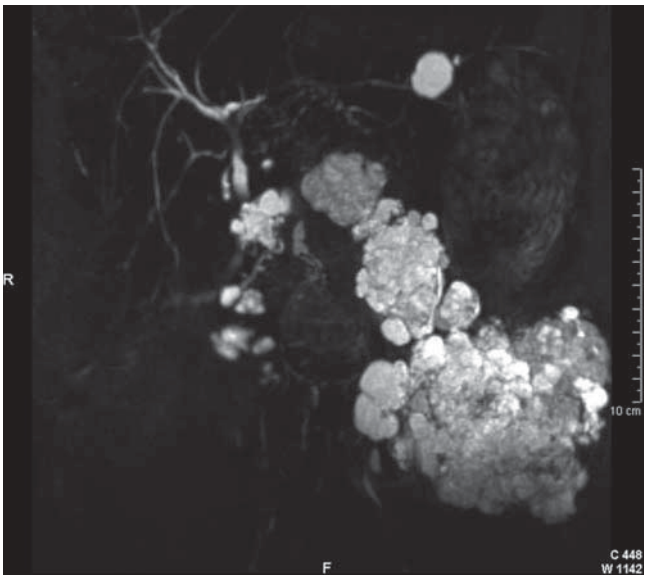

Fig. 3. MR image (T2-weighted) showing the cystic lesions affecting the whole pancreas at the time of diagnosis

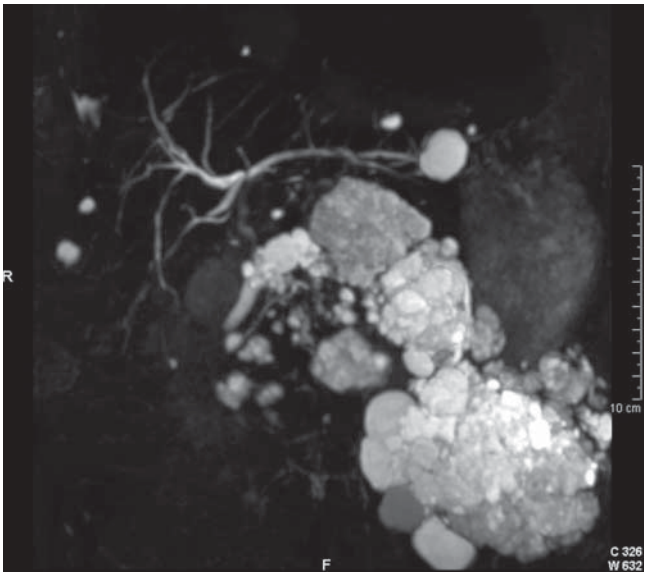

Fig. 4. MR image (T2-weighted), performed at 3-year follow-up, showing the absence of noticeable tumor growth. Lesions in the head are particularly well discernible on this image

weighted images, with lobulated contours. Neither direct nor indirect signs of obstruction were seen in the pancreatic duct, which was clearly identified (Fig. 2).

Tumor markers, including carcinoembryonic antigen (CEA) and carbohydrate antigen (CA) 19.9, were within normal ranges, and the diagnosis of multiple synchronous SCAs of the pancreas was retained. The patient was placed under regular clinical, biological, and radiological (MRI) surveillance. At the 3-year follow-up, she still remained asymptomatic. Tumor markers stayed normal and MRI confirmed the absence of tumor growth (Figs. 3 and 4). Furthermore, the patient underwent an 18-fluorodeoxyglucose positron emission tomography (PET) scan in order to assess the metabolic behavior of the tumor. The patient's fasting glucose was 5.3 mmol/l at the time of the investigation, and full-body, three-dimensional PET scan was undertaken 1 h after the intravenous injection of 285 mBq

18-fluorodeoxyglucose (body weight, 58 kg). This PET scan did not show any hypermetabolic region in the pancreatic lesions or at distant locations.

Discussion

Approximately 70% of patients suffering from SCAs are symptomatic and should be submitted to surgical resection, whereas asymptomatic SCAs can be managed expectantly.[1-4] This attitude is mostly defendable for lesions located in the head of the pancreas or for multiple lesions distributed in the entire pancreas, as their surgical resection is still associated with significant morbidity.[1-4] In our patient, due to the involvement of the entire pancreatic gland, the only appropriate surgical procedure would have been a total pancreatectomy. Thus, determination of the type of cystic tumor, in order to rule out mucinous cystadenoma or intraductal papillary mucinous neoplasm (IPMN) of the pancreas appears to be essential.

The radiological diagnosis of SCA relies on precise criteria previously reported. Numerous small cysts of less than 2 cm, conferring a typical honeycomb pattern, and thin fibrous septa converging into a central scar, sometimes forming a "sunburst calcification" are the major characteristics of SCAs, whereas mucinous cystadenomas are typically unilocular or multilocular, with a small number of discrete compartments. Furthermore, wall and septa are thickened and the uncommon finding of peripheral eggshell calcification on CT appears to be specific to a mucinous cystic neoplasm and highly predictive of cancer.[4,5] IPMN of the pancreas, especially branch duct type, can also look like cystadenoma of the pancreas. In IPMN though, the multicystic mass communicates with the pancreatic duct and appears to be more frequently located in the head or in the uncinate process of the pancreas. Additionally, the presence of intraluminal filling defects, i. e., papillary growths or thickened mucin, also represents an important criterion for the accurate diagnosis of IPMN. In our patient, none of these characteristics could be found on imaging workup and "sunburst" calcifications, as described in our patient, are not typically observed in IPMN.[6,9]

In our patient, the classical features of SCA were present on imaging studies, although most SCAs present as single lesions of the pancreas. The classical imaging procedures give reliable information about these tumors. In a blind retrospective review, Procacci et al.[10] showed that the positive predictive value of CT reached 87.5% in the SCA subgroup.

Multiple pancreatic SCAs are described in von Hippel-Lindau disease, with the pancreatic gland sometimes being totally replaced by the cystic lesions.[11] Nevertheless, the presence of a large number of synchronous multifocal SCAs in patients not affected by this inherited disease has seldom been reported. Most reported SCAs were either symptomatic or degenerated and, in contrast to the management in our patient, were resected or aspirated.[12-17]

The risk of malignant SCA was estimated by Strobel et al.,[18] in a review of the literature, to be less than 3%, but this value was probably overestimated, due to publication bias. Although giving no information about the histological type of cystic tumor, PET scan has recently been proposed as a possible investigation to determine the benign or malignant behavior of cystic lesions of the pancreas. In a series of 56 patients, Sperti et al.[19] found a sensitivity and a specificity of 94% and 95%, respectively.

In conclusion, we have reported a case of an unusual multiple huge asymptomatic SCA involving the whole pancreas. The typical radiological appearance, the normal tumor marker levels, the absence of clinical and radiological progression, and the negative PET scan findings led us to pursue conservative management for our patient.

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