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Degenerated Serous Cystic Tumor of the Pancreas: Case Report and Literature Review of an Aggressive Presentation of a Benign Tumor

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Data Interpretation D
Manuscript Preparation E
Literature Search F
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Patient: Female, 79-year-old
Final Diagnosis: Degenerated serous cystic tumor of the pancreas
Symptoms: Colon polyps
Medication: —
Clinical Procedure: Pancreatectomy
Specialty: Surgery

Objective: Rare coexistence of disease or pathology
Background: Serous cystic tumors of the pancreas are known to present a benign nature and course, not requiring surgery in the absence of symptoms. In rare cases, these benign tumors may present aggressive characteristics such as local infiltration and lymph node and distant metastases. In such cases, a surgical approach may be necessary.
Case Report: We present the case of a 79-year-old woman with an asymptomatic cytologically suggested caudal serous cystic tumor infiltrating the spleen and the splenic vein. This tumor was discovered in a computed tomography scan in the setting of evaluating distant spreading of a primary malignant neoplasm of the rectum. Suspicious malignant signs on imaging dictated a surgical approach and a distal splenopancreatectomy was carried out in the same operative time as the transanal resection of the rectal lesion. The nature of the pancreatic neoplasm was confirmed by histology, but 2 lymph nodes out of 4 retrieved were positive. The postoperative course was uneventful. No adjuvant treatment was proposed. Imaging control 6 months after surgery was not indicative of relapse.
Conclusions: Serous cystic adenomas of the pancreas, although generally considered benign neoplasms, may present with characteristics of malignancy. Moreover, they may prove difficult to differentiate from other malignant neoplasms by non-surgical modalities. Although current guidelines and data from the literature provide controversial information regarding management of these clinical entities, in the presence of suspicious radiological aspects, surgical resection could be considered.

Keywords: Cystadenocarcinoma, Serous • Cystadenoma, Serous • Pancreatic Cyst

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Background

Serous cystic tumor of the pancreas is a rare clinical entity falling into the category of pancreatic cystic neoplasms, along with mucinous cystic tumors and intraductal papillary mucinous neoplasms [1,2]. It can be sporadic or associated with Von-Hippel-Lindau disease. In contrast to its mucin-producing counterparts, malignancy of serous cystic tumors is exceedingly uncommon [1] and this type of tumor tends to be considered as a completely benign lesion. Nonetheless, serous cystic tumors with locally aggressive behavior have been observed. The scarce epidemiological data regarding locally invasive or metastasizing tumors make the proposition of an optimal treatment difficult. In the present study, we present a case of an asymptomatic serous cystic tumor with invasion of nearby tissues without metastasis.

Case Report

A 79-year-old female patient was diagnosed with a 4.2 cm low rectal polyp during screening colonoscopy, 13 cm from the anal verge, and transanal resection was planned. She was a non-smoker, with past light alcohol consumption and her medical history consisted of hypercholesterolemia and hiatal hernia, without family history of cancer. A subsequent computed tomography (CT) scan in search of potential metastases showed a mass located at the tail of the pancreas measuring 12×6.8×9.8 cm with invasion of the hilum of the spleen and splenic vein thrombosis with segmental portal vein hypertension (Figure 1). Blood chromogranin A level was negative, as were tumor markers (CA 19.9 <9 kU/l, CEA <1.8 µg/l). A pancreatic magnetic resonance imaging (MRI) confirmed the presence of a mass at the tail of the pancreas, which is typically associated with a serous cystic adenoma. However, this mass showed invasion of the splenic parenchyma and splenic vein (Figure 2). A second, smaller lesion was found at the uncinete process, and was considered to be an intraductal papillary mucinous neoplasm. An endoscopic ultrasound with fine-needle aspiration was then performed, confirming the presence of these 2 lesions (one in the pancreatic body and the other in the pancreatic tail) with cytopathology of the caudal lesion showing serous cystic adenoma. After discussion in a multidisciplinary tumor board, surgical resection was proposed, based on the locally invasive characteristics. Distal splenopancreatectomy by laparotomy was performed, together with transanal excision of the rectal polyp (TAMIS). Operative time was 240 min and intraoperative blood loss was estimated at 250 ml. Postoperative course was uneventful and the patient was discharged at postoperative day 8.

Histopathology of the specimen showed a 12 cm serous microcystic tumor of the pancreatic tail without atypia, with a partially calcified fibrous stroma invading the spleen and splenic



Figure 1. Axial CT scan showing the masses of the pancreatic body and tail.

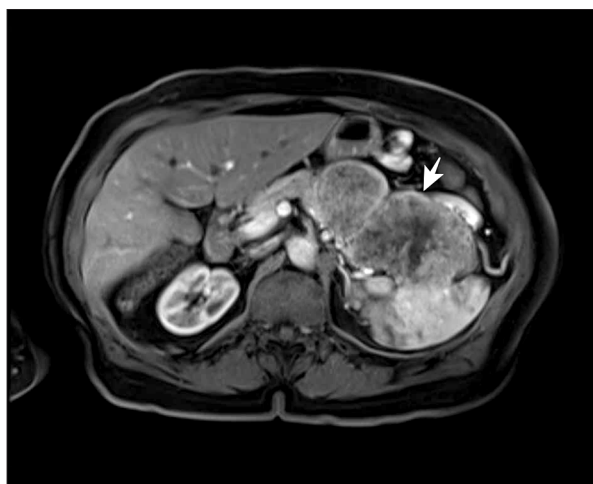


Figure 2. Axial pancreatic MRI (T1) showing the lesion of the pancreatic body and the larger lesion of the pancreatic tail, with invasion of the spleen.

vein (Figure 3). Of the 4 retrieved lymph nodes, 2 were positive, 1 of which appeared to be infiltrated in continuity with the tumor. Whether the second lymph node was also an infiltration in continuity with the tumor or a lymphatic metastasis could not be specified. The second lesion at the pancreatic body was a mixed gastric subtype intraductal papillary mucinous neoplasm without signs of malignancy. Analysis of the rectal polyp showed moderately differentiated adenocarcinoma developing on a tubulovillous adenoma with low- and high-grade dysplasia with negative resection margins.

After a new multidisciplinary oncologic discussion, the pancreatic tumor was classified as a locally invading serous cystic tumor and no adjuvant treatment was proposed. Followup with a thoraco-abdominal CT scan at 6 months postsurgery showed no recurrence.

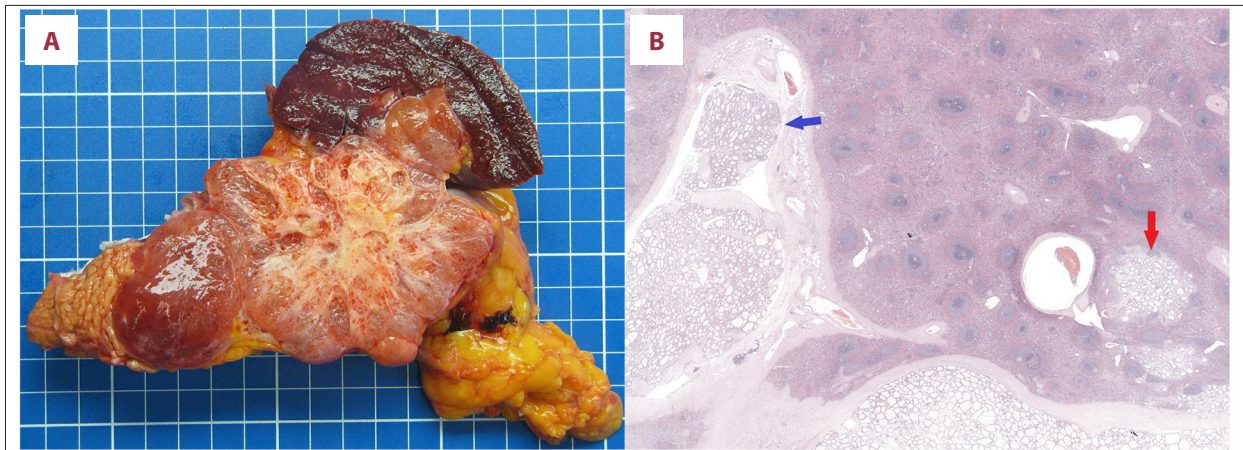


Figure 3. (A) Large lobulated mass of the pancreatic tail with a central calcified scar invading the splenic hilum. (B) Serous cystadenoma invading the splenic parenchyma on the right and the splenic vein on the left. Red arrow: splenic parenchyma invasion, blue arrow: splenic vein invasion. Magnification: 10x; H&E.

Discussion

Although serous cystic tumors are rare lesions, their prevalence remains unknown. One retrospective review of 24 039 patients undergoing CT scan and MRI showed a 0.7% combined incidence of serous cystic adenomas and mucinous cystic adenomas in the general population [3], while serous cystic adenomas may represent 10-16% of total pancreatic cystic neoplasms [4,5].

The macroscopic appearance of serous cystic adenomas may vary, with 4 typical types: microcystic (multiple cysts <2 cm), macrocystic (multiple cysts >2 cm), mixed type (variable-sized cysts) and solid (no visible cysts in imaging) [6], with microcystic being the most common morphology and solid being the rarest variant [7]. Microscopically, serous cystic adenomas demonstrate monomorphous cuboidal-shaped epithelium. Cells are glycogen-rich with cellular cytoplasm and small regular nuclei; mitotic activity is absent. Classification of these tumors as benign, borderline, or malignant depends on the cells' nuclear features [6].

In general, serous cystic adenomas are considered benign lesions with almost nonexistent malignant degeneration. Larger lesions may sometimes infiltrate local lymph nodes or adjacent organs (spleen, stomach, colon) [1]. A noteworthy observation is that such neoplasms with the same monomorphous histological appearance may present signs of malignancy such as invasion of nearby tissues (such as the spleen, splenic vein, or lymph nodes) or distant metastases. Serous cystadenocarcinoma is a term employed for these aggressively behaving variants. Khashab et al mention that histology of serous cystadenoma is insufficient to confirm malignancy, and it is therefore defined by the neoplasm's invading or metastasizing behavior [8]. In 1996, the World Health Organization's *Histological*

Typing of Tumors of the Exocrine Pancreas defined serous cystadenocarcinoma as an "extremely rare tumor that resembles serous cystadenoma but shows invasive and metastatic growth" [2], while the 2019 WHO classification of tumors of the digestive system reserves this term only for neoplasms presenting distant metastases [1]. This relative discrepancy in the details of definition of malignant traits of serous cystic adenomas is highlighted in a large multicenter retrospective study [9]. In our case, the pancreatic tail serous cystic adenoma appeared as a locally invasive mass with infiltration by continuity of the spleen and lymph nodes; it was not considered as a distant metastasis. According to the latest WHO classification, it does not fulfill the criteria for serous cystadenocarcinoma. **Table 1** presents all case reports or case series up to date, reporting pancreatic serous cystadenomas with malignant features as well as treatment choice per case.

The optimal management of serous cystic adenomas remains controversial to this day. Generally, watchful waiting is proposed, due to their benign nature, with surgical resection being reserved for symptomatic tumors or when malignancy cannot be formally excluded [10]. In the latter cases, oncological resection with followup, as is done for pancreatic cancer, is recommended. Conversely, guidelines from the European Study Group on Cystic Tumors of the Pancreas completely discard the possibility of a malignant serous cystic adenoma and reserve operation only for symptomatic patients, proposing no followup for serous cystic lesions when diagnosis is certain [11]. In our case, radiological invasion of the spleen and splenic vein compelled the surgical resection of this potentially degenerated lesion.

Finally, to date, there are no cases describing the coexistence of rectal adenocarcinoma and serous cystic tumors of the pancreas. Data in the pertinent literature do not support a

Table 1. Studies involving degenerated serous cystadenomas of the pancreas and selected treatment.

Author/Year	Symptoms/signs	Localization	Malignant feature(s)	Surgical treatment	Outcome
Kameh et al, 1991 [11]	Jaundice	Head	Focal perineural invasion	Total pancreatectomy	Not disclosed
Ohta et al, 1993 [12]	Incidental finding	Body	Vascular and perivascular invasion	Enucleation	Cured
Widmaier et al, 1996 [13]	Abnormal liver tests	Head	Lymph node metastasis	Pylorus-preserving partial duodenopancreatectomy	Cured
Abe et al, 1998 [14]	Palpable mass at the left hypochondrium	Body and tail	Lymph node invasion	Distal pancreatectomy with splenectomy	Cured
Friebe et al, 2005 [15]	Anorexia, weight loss, abdominal pain	Body and tail	Spleen invasion	Distal pancreatectomy with splenectomy	Cured
Matsumoto et al, 2005 [16]	Palpable mass at the left hypochondrium	Tail	Spleen and colonic mesentery invasion, Single lymph node metastasis	Distal pancreatectomy, splenectomy, and segmental resection of the colon	Cured
Shintaku et al, 2005 [17]	Fatigue and diarrhea	Body and tail	Spleen and neural invasion	Distal gastrectomy, distal pancreatectomy with splenectomy	Not disclosed
King et al, 2009 [18]	Gastric bleeding and abdominal pain	Head	Duodenal invasion	Pancreaticoduodenectomy	Cured
Cho et al, 2011 [19]	Hematochezia	Tail	Spleen and transverse colon invasion	Distal pancreatectomy, splenectomy, and segmental resection of the colon	Not disclosed
Kadhirvel et al, 2015 [20]	Abdominal pain	Body and tail	Spleen invasion by contiguity	Distal pancreatectomy with splenectomy	Not disclosed
Gao et al, 2016 [21]	Back pain	Body	Spleen vein encasement	Distal pancreatectomy and splenectomy	Not disclosed
Kawai et al, 2020 [22]	Palpable mass at the left hypochondrium	Head	Superior mesenteric vein invasion	Pancreaticoduodenectomy with resection of limited superior mesenteric vein	Liver and peritoneal metastases after 4 years
Yagi et al, 2020 [23]	Diabetes mellitus	Tail	Spleen invasion	Laparoscopic distal pancreatectomy with splenectomy	Cured

potential connection, via genetic predisposition, between the 2 neoplasms, although these data are scarce and dedicated studies do not exist [12,13].

Conclusions

A grey area exists between aggressively behaving serous cystic adenomas and serous cystic adenocarcinomas. Although rare entities, these tumors pose a medical riddle for the practitioner, posing the question of whether surgical resection only

would suffice to avoid malignancy-associated sequelae like disease relapse or metachronous metastases. A unanimous agreement is needed to better define these entities and evaluate their malignant potential, which is the deciding factor for their management.

Declaration of Figures' Authenticity

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.

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