SEROUS CYSTADENOMA OF PANCREAS WITH STROMAL AMYLOID DEPOSITS, VERY RARE ENTITY

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ABSTRACT
Serous cystadenoma (SCA) of pancreas is a rare tumour and unique for its benign nature with little malignant potential. Classically the tumor is riddled with innumerable small cysts around a stellate scar. The quintessential histological features are closely placed small cysts lined by glycogen rich cuboidal epithelium. In view of its excellent prognostic outcome, this tumor needs to be accurately diagnosed. Very rarely the tumor is associated with amyloid material. This report documents a case of SCA with stromal amyloid deposits occurring in a 72-year-old female, SCA with amyloid deposits has been reported in only one of the literature.

KEYWORDS: Pancreatic Exocrine Tumors, Pancreas and Amyloid.

INTRODUCTION
Microcystic adenoma or serous cystadenoma (SCA) of the pancreas is an uncommon, benign tumor that accounts for 1–2% of pancreatic exocrine neoplasms and 25% of cystic neoplasms.[1] Their prevalence is increasing, primarily due to imaging technique improvements. These tumors are usually unifocal, mostly arising in the body and tail of the pancreas and are often incidentally detected. They present as single, large, well demarcated, and multiloculated cystic tumors, 1–25 cm in size. They are composed of cysts lined by epithelial cells that produce serous fluid and show ultrastructural evidence of centroacinar differentiation.[2]

Pancreatic serous cystadenomas were referred as microcystic cystadenoma or glycogen-rich cystadenoma. These terms are no longer considered appropriate. The preferred name is now serous cystadenoma and are more common than mucinous cystic neoplasm, with a ratio of about 2:1.[3]

Serous cystadenoma may be associated with other disorders which include Von Hippel Lindau (VHL), Evans syndrome, intestinal hemangiomas and rare entities like mediastinal lipoma.[4]

CASE HISTORY
A 72-year-old female presented with vague abdominal pain and loss of appetite since six months. On clinical examination a mass in the left upper quadrant was noted. An endoscopy-guided FNAC revealed; scattered benign epithelial cells in the proteinaceous background on microscopy, this was reported as benign epithelial lesion of pancreas. CT scan of the abdomen showed multiseptated heterogeneously enhancing mass lesion in the tail of pancreas and diagnosed as cystadenoma of pancreas.

PATHOLOGIC FINDINGS
Gross examination showed a large, encapsulated mass measuring 9.5X8X8 cm in the body and tail of pancreas along with spleen [Figure-1 A]. Cut sections of the mass revealed a 'honey comb' appearance imparted by innumerable small cysts ranging from 0.1-2.0cm in diameter [Figure 1]. Most of the cysts contained clear serous, watery or blood-tinged fluid. Also observed was a central 2x2cm irregular stellate scar[Figure -1 B].

Spleen measured 10X6X3 cms. External surface and cut surface were normal. No lymph node were found in the splenic hilum.

Microscopy revealed numerous closely placed , variable sized ,thin walled cysts lined by a single row of cuboidal epithelium with moderate to abundant clear cytoplasm and relatively monomorphic nuclei with inconspicuous nucleoli ; no nuclear atypia or increase in mitosis was noted, the lumen showed pink granular material [Figure-3] [Figure-2 A].

Staining with periodic acid–Schiff demonstrated PAS-positive, diastase–sensitive intracytoplasmic glycogen [Figure-5]. The intervening stroma was loose, hypocellular with lamellar hyalinized areas which were positively stained by congo red [Figure-2 B] and showed
green birefringence, dichroism with polarized light confirming the amyloid deposits. Compressed pancreatic tissue was seen at the periphery. Sections from spleen showed no significant pathology. Section from proximal surgical and proximal portion of pancreas margin was free.

Legends to Figures

Figure-1 [A&B].

A. Pancreatic mass with spleen.
B. Pancreas showing honey comb appearance with central stellate scar.

A. Cysts lined by flattened to cuboidal epithelium.
Magnification -High power (40X)
Stain used - Hematoxylin and Eosin

B. Magnification -High power (40X).
Stain used -PAS-reaction – positive

DISCUSSION

The classification of cystic pancreatic tumors was beset with confusion among pathologists till the landmark paper of Compagno and Oertel in 1978 that defined and separated serous cystic pancreatic tumors, which are almost always benign, from mucinous tumors which are potentially or frankly malignant.
According to the World Health Organization (WHO) the serous cystic neoplasms are subclassified into two groups, serous microcystic adenomas and serous oligocystic adenomas. The serous oligocystic adenoma is an often ill-demarcated tumor composed of a few cysts with diameters of 1–2 cm. Serous cystic tumors showing larger cysts have been referred to as serous macrocystic adenomas. A third type, the disseminated variant (DV), has been recognized because of its rare involvement of the entire pancreas. Although DVs are rare, they are more commonly reported in association with Von Hippel-Lindau (VHL) disease. In a rare case report by Bilal O Al-Jifty et al, have reported a disseminated variant of pancreatic serous cystadenoma causing obstructive jaundice in a 73 year old man, the tumor was completely involving the whole pancreas, without association with VHL disease.

The head of the pancreas is an uncommon location for SCA as compared with the body and tail. However, a case of SCA occurring in the head of pancreas has been reported.

SCA increasingly being diagnosed at an asymptomatic stage due to improvement in imaging modalities like computerized tomography (CT), endoscopic ultrasound (EUS), and magnetic resonance cholangiopancrea to graphy (MRCP). These tests will reveal a cystic mass within the pancreas. The cysts do not communicate with the larger pancreatic ducts SCA. However in some cases a fine needle aspiration (FNA) biopsy can be obtained to confirm the diagnosis.

Histochemistry (periodic acid-schiff [PAS] and mucin) and immunohistochemistry also play significant role in diagnosis of SCA. They show positive staining for PAS that is diastase sensitive. Typically, they stain positive for cytokeratins (CK7 & CK19), neuron-specific enolase (NSE), α-inhibin, MUC-1 and MUC-6, and negative for α-smooth muscle actin, S-100 protein, carcinoembryonic antigen (CEA), chromogranin and synaptophysin. Very rarely stromal amyloid can be seen in these neoplasms which can be picked up by Congo red staining and confirmed by polarized light. Immunohistochemical studies for amyloid protein done by using antibodies like anti-beta amyloid protein and anti amyloid precursor pre-A4695, the former antibody diffusely stains tumour stroma, while the latter stains scattered stroma cells.

Differential diagnosis of SCA includes other cystic lesions of the pancreas, lymphangioma and metastatic renal cell carcinoma. Differentiation from mucinous pancreatic neoplasms is of paramount importance considering the latter's potential for malignant behaviour.

To conclude, pancreatic serous cystadenoma is an uncommon tumor with a unique gross and microscopic morphology with generally benign course. Hence it needs to be accurately diagnosed and differentiated from other malignant or potentially malignant pancreatic tumors. However, in the light of the recent findings of rare instances of malignant transformation and of co-existent potentially malignant tumors, a thorough sampling of the specimen and postoperative follow-up by regular CT surveillance is advocated. The case reported here is rare for the association of serous cystadenoma with stromal amyloid deposition which has been reported in only one of the literature.

REFERENCES


