CASE REPORT

Co-existence of Diffuse Serous Cystadenoma and Pancreatic Neuroendocrine Tumor

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Abstract Diffuse serous cystic neoplasm (SCN) associated with pancreatic neuroendocrine tumor (PNET) is a rare finding reported previously in only three patients to the best of our knowledge. We herein present one such interesting report of a diffuse serous cystic adenoma (SCA) and co-existent PNET in a 25-year old lady who presented with abdominal pain for past 6 months. A triple-phase pancreatic protocol computed tomography (CT) scan revealed multiple cysts involving the entire pancreas. The cysts were thin walled, ranging from 2 to 8 cm in width, with no calcification or central scar that was confirmed at laparotomy. A frozen section revealed a neuroendocrine tumor and she underwent total pancreatectomy. Diffuse SCA with co-existent PNET infiltrating nerve bundles of the pancreatic parenchyma was made upon histopathology further verified by chromogranin-A immunostaining. The patient is insulin dependent and doing well at 2 years of follow-up. The origin of endocrine tumors from multipotent ductular stem cells has been suggested.

Keywords Serous cystadenoma · Pancreatic neuroendocrine tumor · Total pancreatectomy · Pathology

Introduction

Diffuse serous cystic neoplasm (SCN) is a rare presentation with only eight cases that have been reported this far [1]. Some have

been seen in Von Hippel-Lindau disease. Associated pancreatic neuroendocrine tumor (PNET) in diffuse SCN is further rarely reported earlier in three patients to the best of our knowledge [1–3]. We herein present one such interesting report of a diffuse serous cystic adenoma (SCA) and co-existent PNET.

Case Report

A 25-year old lady presented with complaints of abdominal pain for the past 6 months. Clinical examination was unremarkable. Blood investigations were normal including her blood sugar, liver function tests, serum lipase, and amylase, CA 19-9 and CEA. She had an ultrasound of the abdomen that indicated the presence of cystic lesions in the pancreas. A triple-phase pancreatic protocol computed tomography (CT) scan revealed multiple cysts involving the entire pancreas. The cysts were thin walled, ranging from 2 to 8 cm in width, with no calcification or central scar (Fig. 1). The pancreatic duct was normal in size and there appeared no communication of the cysts with the pancreatic duct. A diagnosis of multifocal mucinous cystic neoplasm (MCN) was made. No lesions were seen in the kidney and her retina examination was normal.

At laparotomy, the pancreas was seen to have multiple cystic lesions with larger cysts on the body and tail of the pancreas. A frozen section revealed infiltrative nests and acini of the neuroendocrine tumor (Fig. 2). Total pancreatectomy was performed. The patient developed diabetes postoperatively that was controlled on insulin. The postoperative recovery was otherwise uneventful and the patient was discharged on postoperative day 6.

Histopathology revealed cystic lesions lined by cuboidal epithelial cells with clear cytoplasm and osseous metaplasia of the wall. A diagnosis of diffuse SCA with co-existent PNET infiltrating the nerve bundles of the pancreatic parenchyma

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Fig. 1 **a** Computed tomography scan showing multiple cystic lesions of the pancreas (yellow arrows). **b** Gross specimen showing pancreas replaced by multiple cystic lesions—the largest being in the body of the pancreas (yellow arrows). **c** Cystic lesions in the tail of the pancreas (yellow arrow). **d** Cystic lesions in the head and uncinate process of the pancreas (yellow arrows)

was made. This was further verified by immunostaining using chromogranin-A that was strongly positive (Fig. 2).

Discussion

SCNs represent about 10–16% [4] of all resected cystic tumors of the pancreas. These neoplasms are composed of non-

atypical cuboidal glycogen-rich epithelial cells that produce a watery fluid. Typically, they do not produce mucin and do not communicate with the pancreatic ductal system. Five macroscopic variants of SCN have been described: microcystic, macrocystic or oligocystic, mixed micro-macrocytic, Von Hippel-Lindau associated, and solid.

SCNs are commoner in women, usually benign, although rare malignant examples have been reported [5]. Microcystic

Fig. 2 **a** Frozen, shows infiltrative nests and acini of neuroendocrine tumor (hematoxylin and eosin stain, $\times 200$). **b** $\times 20$, photomicrograph showing a serous cyst lining with undersurface showing tumorlets of atypical neuroendocrine tumor and congested blood vessels (hematoxylin and eosin stain; $\times 200$). **c** $\times 40$, shows nerve bundle infiltrated by nests and acini of neuroendocrine tumor (hematoxylin and eosin stain; $\times 400$) **d** 20×1 , shows strong cytoplasmic positivity in the nests and acini of tumor cells (chromogranin immunostain $\times 200$)

serous cystadenoma appears like a “honeycomb” with a typical central calcification named “central scar” on CT scan. However, less common macrocystic variants may be more difficult to differentiate from a mucinous tumor. No communication of the cysts with the pancreatic duct was seen in our patient making the diagnosis of intraductal papillary mucinous neoplasm (IPMN) unlikely on preoperative imaging. A magnetic resonance cholangiopancreatography (MRCP) is often the preferred technique if confusion persists, as MRCP is capable of detecting the lack of a communication between the cyst and the ductal system with a higher accuracy.

Unifocal, asymptomatic, small (cut off 4 cm has been suggested) SCN may be managed expectantly with close follow-up [6]. In diffuse serous cystadenomas, it is impossible to draw conclusions due to their extreme rarity, although most surgeons would find it safe to operate on large and symptomatic tumors.

The origin of endocrine tumors from multipotent ductular stem cells was suggested by Bordi and Bussolati in 1974 [7] by the presence of nesidioblastosis in the ductules surrounding endocrine tumors, the origin of islets and the frequent finding of ductular cells in pancreatic endocrine tumors [8, 9].

To our knowledge, a PNET arising in association with a serous cystadenoma was described first by Heresbach et al. in 1993 [10]. Three patients have previously been reported in literature to have a mixed tumor with a diffuse or multifocal serous cystadenoma and a neuroendocrine component to the best of our knowledge [1–3].

Compliance with Ethical Standard

Conflict of Interests The authors declare that they have no conflict of interest.

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