Mucinous Cystic Neoplasm of Pancreas in a Male Patient: A Case Report and Review of the Literature

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Mucinous cystic neoplasms (MCNs) are among the most common primary cystic neoplasms of pancreas. These lesions usually occur in body and tail of the pancreas and are characterized by the presence of ovarian type stroma in the pathological evaluation. Mucinous cystic neoplasms have significant malignant potential; therefore, their diagnosis and resection is of utmost importance. Mucinous cystic neoplasms typically occur in women. Only a few cases have been previously reported in male patients. In this case report, we present a 48-year-old man who was referred to our center due to an incidentally found cystic lesion in the tail of the pancreas that was increasing in size in serial evaluation. The patient underwent open distal pancreatectomy. The pathology showed mucinous cystic neoplasm with characteristic ovarian type stroma and positive staining for estrogen and progesterone receptors. This case report shows that mucinous cystic neoplasms can occur in men and should be considered in differential diagnosis of cystic pancreatic lesions in this population.

INTRODUCTION

Mucinous cystic neoplasms (MCNs) are one of the three most common cystic neoplasms of pancreas and comprise nearly half of these cases.[1] MCNs are usually located in the body and tail of the pancreas and are characterized by the presence of the ovarian type stroma in the pathological evaluation.[1,2] Due to the significant malignant potential of mucinous cystic neoplasms, their diagnosis and resection is of utmost importance.[1,2] MCNs almost always occur in women.[1] Occurrence of MCN in males is very rare and only a few cases have been previously reported.[2-8] In this article, we present a case of MCN in a male patient who was treated at our center.

CASE PRESENTATION

A 48-year-old Caucasian male was referred to our clinic because of a cystic mass in the tail of the pancreas. This mass was incidentally found one year prior to the referral to us on a CT scan performed due to an abdominal gunshot wound that resulted in splenectomy. The trauma surgeon did not resect the pancreatic lesion and decided to monitor its progression. On serial abdominal CT scans, the mass increased in size from 4.3 x 3.7 cm at the initial CT scan to 4.7 x 4.6 cm one year later (Figure 1). Although the patient was asymptomatic due to the increase in the size and the suspicion of the malignant nature of the lesion, he was referred to us and underwent open distal pancreatectomy. Grossly, the lesion consisted of a 5 x 3 x 2 cm multilocular cyst in the tail of the pancreas. Microscopically, the cyst demonstrated a flat epithelium, which varied from a mucin-producing columnar epithelium to a cuboidal epithelium (Figure 2, Panel A). No significant atypia was identified. Subjacent to the epithelium, a distinctive ovarian-type stroma was present, composed of densely packed spindled cells with regular elongated and wavy nuclei (Figure 2, Panel A). These nuclei showed strong immunoreactivity with estrogen and progesterone receptor stains (Figure 2, Panels B and C). After finding this type of pathology, male karyotyping was ordered for the patient. Cytogenetic analysis of bone marrow aspirate revealed a normal male karyotype.

The patient recovered well postoperatively and is symptom-free three months after the surgery.

DISCUSSION

Based on World Heath Organization (WHO) criteria, MCNs are defined as cystic epitheilal neoplasms composed of mucin producing columnar epithelium and an ovarian-type stroma which forms a band of densely packed stromal cells beneath the epithelium.[1,3] Immunohistochemial staining of the ovarian-type stroma in MCN is usually positive for estrogen and progesterone receptors.[1,2] Presence of this

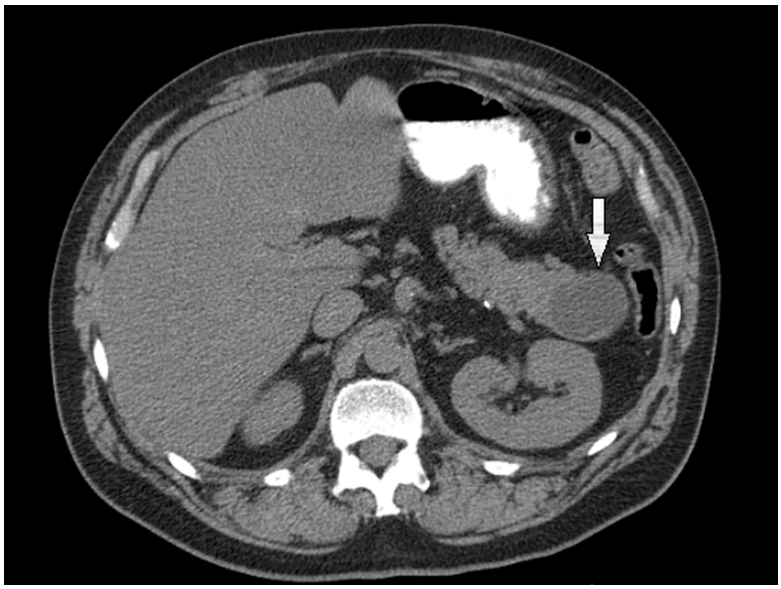

Figure 1: Abdominal CT scan showing a cystic mass in the tail of the pancreas (arrow).

ovarian-type stroma is essential for pathological diagnosis of MCNs.[1,3] MCNs typically occur in premenopausal women.[1,2] To our knowledge, only 10 cases of MCN in men diagnosed based on WHO criteria have been previously reported in the literature.[2-8] Our case is the 11th case of MCN reported in a man.

Because MCNs rarely happen in men, to rule out any chromosomal abnormality in our patient, we performed karyotyping, which showed normal male XY karyotype. This shows that MCNs can happen in men without any chromosomal abnormality.

As in our patient, MCNs are usually asymptomatic and are incidentally found during abdominal imaging for evaluation of another often unrelated clinical indication.[1] However, MCNs can occasionally present with abdominal pain and fullness, nausea, vomiting, recurrent pancreatitis, or gastric outlet obstruction.[1,5,9] Presence of symptoms such as weight loss or jaundice should raise the suspicion of the malignant nature of the MCNs.[1,9]

In cross-sectional imaging by ultrasonography, CT, or MRI, MCNs are usually found as spherical macrocystic masses.[1] MCNs usually do not communicate with pancreatic ducts but could result in the dilatation of the pancreatic ducts through pressure effect and obstruction.[1,10] The following findings in the imaging of MCNs are in favor of malignant nature of these lesions: large size (>5cm), the presence of calcification, multiple papillary invaginations, a mural node or asymmetrically thickened wall, an eccentrically located mass within a cystic area, local vascular invasion, pericystic reaction, extrahepatic biliary obstruction, splenic vein obstruction or ascites.[1,5] ERCP could be very helpful in differentiation of MCNs which usually do not communicate with pancreatic ducts versus psuedocysts or branch duct IPMNs which communicate with pancreatic ducts.[1] Moreover, ERCP could show distortion of pancreatic or biliary ducts by MCNs, which is in favor of malignant nature of MCNs. ERCP could also relieve the biliary obstruction in patients with jaundice. However, ERCP is not necessary for diagnosis and is not used in most patients with MCNs. Like ERCP, MRCP could also be helpful in differentiation of pseudocysts or branch duct IPMNs from MCNs.[1] By providing a clear imaging of the tail of pancreas and its ductal system, EUS could also be helpful in differentiating MCNs from other cystic lesions.[1] Moreover, EUS could be used for FNA. FNA could also be achieved percutaneously under CT-guidance. FNA analysis of MCNs shows honeycomb sheets and clusters of mucin-producing columnar cells. Abundant mucin in the background of the MCNs could be helpful in differentiation of these lesions from serous cystic neoplasms or pseudocysts.[11] The degree of cellular atypia in FNA could also be predictive of malignancy.[12]

Because of the possibility of malignant transformation of MCNs, they should be resected irrespective of their size or location.[1,5] MCNs are usually located in the body and tail of the pancreas;[5] open, laparoscopic, or robotic distal pancreatectomy with or without splenectomy is the operation of choice for these lesions.[1] MCNs could also rarely happen in the head of the pancreas; formal pancreatoduodenectomy is the operation of choice for these lesions.[1] Extra caution should be implemented during the surgery not to rupture the cyst, as it could lead to intra-abdominal seeding of tumor cells.[1] Moreover, in order to provide an optimal sample for pathologist, the cyst should be removed intact and not marsupialized.[1]

Complete resection of MCNs with benign nature is considered a complete cure.[1,10] These lesions do not recur; therefore, no follow-up imaging is needed.[1,13] However, patients with malignant lesions have poor prognosis; their five-year survival is 15%-35%.[13] The most important predictor of prognosis in malignant MCNs is the extent of tumor invasion.[14] After resection, patients with malignant MCNs are recommended to have follow-up imaging with CT or MRI for evaluation of possible recurrences or metastases.[1]

In summary, we present a case MCN of pancreas with characteristic pathological finding of ovarian-type stroma in a man. Our study shows that MCN could occur in men and should be considered in the differential diagnosis of pancreatic lesions in men, as well as women.

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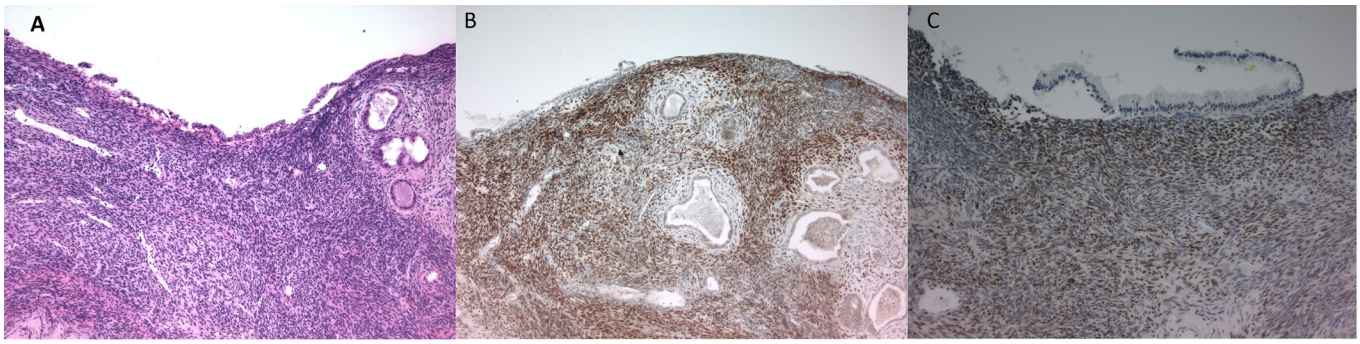

Figure 2: Panel A: Photomicrograph illustrating the epithelial lining with prominent underlying ovarian-type stroma (Hematoxylin and Eosin stain, 100x original magnification). **Panel B:** Photomicrograph illustrating strong nuclear immunoreactivity of the ovarian-type stroma with estrogen receptor stain (100x original magnification). **Panel C:** Photomicrograph illustrating strong nuclear immunoreactivity of the ovarian-type stroma with progesterone receptor stain (100x original magnification).

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