A Rare Case of an Intraductal Papillary Mucinous Neoplasm of Pancreas Fistulizing Into Duodenum With Adult Polycystic Kidney Disease

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Abstract

Intraductal papillary mucinous neoplasm (IPMN) accounts for 20-50% of all cystic neoplasms of the pancreas. Rarely, IPMN, whether benign or malignant, can fistulize into adjacent organs like duodenum, stomach or common bile duct. IPMN can be associated with other diseases like Peutz-Jeghers syndrome and familial adenomatous polyposis. Association with adult polycystic kidney disease (ADPKD) is extremely rare. We report a case of a 60-year-old male with a large IPMN in the head of the pancreas diagnosed by magnetic resonance imaging, endoscopic ultrasound and cyst fluid analysis. It was complicated by fistula formation into the second part of the duodenum. Patient was simultaneously having adult polycystic kidney disease. There is only one case report of uncomplicated IPMN with ADPKD in the literature so far. And even rarer, there is no any case report of fistulizing IPMN with ADPKD reported so far, to the best of our knowledge.

Keywords: Fistulizing IPMN; ADPKD; Duodenum

Introduction

Cystic neoplasms of the pancreas are rarely encountered, diagnosed in 10% of pancreatic cysts detected on imaging. They consist of intraductal papillary mucinous neoplasm (IPMN) of the pancreas, mucinous cystic neoplasm, serous cystadenoma, papillary cystic tumors and cystic islet cell tumors [1]. IPMN accounts for 1-3% of all exocrine pancreatic neoplasms and 20-50% of all cystic neoplasms of the pancreas [2].

Though rare, IPMN, whether benign or malignant, can fistulize into adjacent organs like duodenum or stomach. Moreover, IPMN can be associated with many other conditions like Peutz-Jeghers syndrome and familial adenomatous polyposis but association with adult polycystic kidney disease (ADPKD) is extremely rare and is almost unheard of. Only one case of IPMN (non-complicated) with ADPKD has been reported in the literature [3].

We report a case of a 60-year-old male diagnosed to have an IPMN complicated by a fistula formation into second part of the duodenum associated with ADPKD. There is not a single case report of similar combined presentation in an individual patient reported in the literature so far, to the best of our knowledge.

Case Report

A 60-year-old male, chronic smoker, non-alcoholic, presented with 3 months history of pain in upper abdomen which was epigastric, dull aching, continuous, mild to moderate intensity and radiating to back. He had significant anorexia and weight loss of 8 kg. He did not have similar pain in the past. On physical examination, the patient was pale, malnourished, afebrile, with a blood pressure of 150/90 mm Hg, pulse rate of 90 beats per minute and a respiratory rate of 22 breaths per minute. Abdominal examination revealed soft and non-tender abdomen with non-palpable liver and spleen. There was an approximately 8 × 8 cm ballotable non-tender cystic lump with ill defined margins noted in left lumbar region which did not move with respiration.

Laboratory examination showed hemoglobin of 9.2 g/dL, total leukocyte count 8,800/mm\textsuperscript{3}, and platelet count of 3.3 lakh/mm\textsuperscript{3}. Renal function tests showed serum creatinine of 3.3 mg/dL and blood urea nitrogen of 54 mg/dL. Liver function tests were normal. Serum amylase and lipase were 90 IU/L and 80 IU/L, respectively. Serum CA 19-9 was 20 IU/mL.

Ultrasonography of the abdomen was suggestive of dilated pancreatic duct (7 mm in head) and a 6 × 5 × 5 cm well defined cystic collection with wall thickness of 4 mm with multiple mobile internal echoes seen in the head and body of pancreas which was communicating with pancreatic duct. It also showed bilateral innumerable renal cortical cysts, largest measuring 10 × 10 cm in left kidney. Magnetic resonance cholangiopancreatography (MRCP) with magnetic resonance imaging (MRI) abdomen showed large 8 × 5 × 5 cm cystic...
irregular collection with septations involving head and body of the pancreas, almost replacing them with 7.5 mm pancreatic duct. MRI also suggested bilateral polycystic kidney disease (Fig. 1). Endoscopic ultrasound (EUS) suggested ill defined cystic lesion in the head of the pancreas with some solid component and 7 mm pancreatic duct (Fig. 2). Second part of duodenum seemed to be ulcerated and the cyst was in communication with D2. EUS guided aspiration of pancreatic cyst fluid revealed high viscosity fluid with fluid amylase 945 IU/L and fluid CEA of 312 ng/mL. Cytology revealed clusters
of columnar epithelial cells with mild degree of dysplasia in
a background of mucin-like material, histiocytes and cellular
debris. Subsequent esophagastroduodenoscopy revealed a
fistulous opening on the medial wall of the second part of duode-
num (D2) just distal to the papilla with infiltrated mucosa.
The opening was discharging mucus material and necrotic de-
bris. The tumor cavity was visible through the opening (Fig.
3). Biopsy from the fistulous tract was taken. It turned out to be
negative for malignancy. We referred the patient for pancreati-
coduodenectomy. But the patient was declared high risk by the
anesthetist in view of chronic renal failure, hypertension, poor
respiratory reserve and ischemic heart disease. Patient chose
not to undergo surgery.

Discussion

IPMN is defined as an intraductal epithelial tumor composed
of mucin-producing columnar cells showing papillary prolif-
eration, cyst formation, and variable degrees of cellular atypia,
even within an individual neoplasm.

IPMN, whether benign or malignant, can become compli-
cated in 6.6% cases forming a fistula into adjacent organs [4].
The fistulizing behavior is of two types: penetrating/invasive
type and automatic/mechanical type. In the penetrating/inva-
sive type, tumor cells invade the adjacent organs by forming a
malignant fistula. This pattern is seen mostly in malignant tu-
mors only. In the automatic type, rising intratumoral pressure
causes the tumor to rupture into adjacent organ releasing the
content into it [5]. This type is seen mostly in benign IPMN,
though a large malignant IPMN can also have this type of fis-
tula. Kobayashi et al reported that automatic/mechanical type
was accounted for 67% cases of fistulizing IPMN [4]. In our
case, biopsy from fistula site in the duodenum was negative
for malignancy. So, it was classified as automatic type rupture
resulting in fistula formation.

The most common organ fistulized is the duodenum
(64%), followed by the common bile duct (56%) and the stom-
ach (17%) [5]. Few case reports of intraperitoneal rupture re-
sulting in pseudomyxoma peritonei have been reported [6].
The 5-year survival rates of patients with benign and mali-
gnant IPMNs have been reported to be 85-100% and 25-65%,
respectively. However, the 5-year survival of patients with IP-
MNs fistulizing into other organs cannot be calculated because
the case reports of fistulizing IPMN are rare according to the
literature review.

Association of IPMN with ADPKD is very rare. Only one
case has been reported in 2009 by Yasunori Sato from Japan.
Simple pancreatic cysts can be found in as many as 7-10% pa-
tients with ADPKD, but rarely a cyst in the pancreas can turn
out to be a cystic neoplasm of the pancreas. Other diseases that
are associated with IPMN are Peutz-Jeghers syndrome and fa-
miliar adenomatous polyposis [7].

In case of fistulizing IPMN, a biopsy specimen should
be taken from the fistula endoscopically which would reveal
whether the tumor has invaded an organ adjacent to the pancre-
as and enable determination of extent of surgery and progno-
sis. There are no clear guidelines to manage IPMN complicat-
ed by a fistula formation. However, pancreaticoduodenectomy
is the standard of care for an IPMN fistulizing into duodenum
in most of the centers.

Our patient had a large IPMN involving the head of the
pancreas diagnosed on MRI, EUS and cyst fluid analysis with
unusual feature of fistula formation into second part of duode-
um and simultaneous association with ADPKD. This is the
first case, to the best of our knowledge, of a large IPMN with
ADPKD and a fistula formation into duodenum.

Conflict of Interest

None declared.

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