An IPMN Inside a Serous Cystadenoma: Is That Possible?

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Abstract

Objectives: Although rare, the pancreatic cystic neoplasms are detected with increased frequency lately. The treatment of these modalities however remains controversial.

Methods: In this case reports a multifocal Intraductal Papillary Mucinous Neoplasm (IPMN) along with Pancreatic Serous Cystadenoma (PSC) were identified in the pancreatic specimen following a distal pancreatectomy. The results of the histopathology confirmed the existence of an IPMN inside a large PSC.

Results: In our case, the patient with this extremely rare type of mixed cystic neoplasm was managed successfully with extensive distal pancreatectomy.

Conclusions: The preoperative evaluation of the extent of the pancreatic involvement and the definition of the histologic type of the involved neoplasms before the pancreatic resection is challenging.

Keywords: Papillary mucinous neoplasm; Serous cystadenoma; MultiFocality

Introduction

Due to the development of the imaging and endoscopic procedures, an enormous increase in the incidental identification of asymptomatic pancreatic neoplasms has occurred lately during scans for investigations for other modalities [1]. Intraductal Papillary Mucinous Neoplasms (IPMN) and Pancreatic Serous Cystadenomas (PSC) belong to the category of cystic neoplasms which comprise only 1% of the pancreatic cancers [2,3]. Inspite of the evolution in the preoperative evaluation of the patients with these neoplasms, difficulties in the oncologic risk assessment and the operative planning still remain. Distinguishing the invasive IPMN from the benign cases before the operating day is challenging. Moreover the management of - large cysts with almost no malignant potential such as large PSC - IPMNs with multiple involvements of branch ducts is still under debate mainly due to the risk of recurrence as well as the significant rates of complications and the functional pancreatic insufficiency (exocrine and endocrine) that pancreatic resections can cause.

Materials and Methods

A 83 year old lady presented in our department in June of 2013 with a 2 month history of epigastric pain, weakness and loss of appetite. The pain was getting better after the meals. The results of a gastroscopy. Another cyst with similar morphology was detected at the tail of the pancreas (19.5 mm×14.9 mm) (Figure 2). The results of the gastroscopy). Two years before, she had undergone endoscopic resection of a polyp of the stomach with normal epithelium giving the impression of the cecum which was proved to be non malignant.

Results

The first imaging test carried after admission an Endoscopic Ultrasound (EUS) with Fine Needle Aspiration (FNA). The EUS was indicative of a spheric entity with size 45.5 mm×34.2 mm. A biopsy was taken from a small cystic wall projection which was recognized inside this cyst (Figure 1). The cyst was adherent to the splenic vessels and was applying pressure to the gastric wall (confirming the finding of the gastroscopy). Another cyst with similar morphology was detected at the body of the pancreas (19.5 mm×14.9 mm) (Figure 2). The results of the FNA were indicative of IPMN with medium grade epithelial dysplasia.

Almost ten days after the EUS–FNA, an extensive distal pancreatectomy and splenectomy were performed in an open procedure. The histopathological examination showed a specimen of pancreas with size of 12 cm×6 cm×2.5 cm. macroscopically in the tail of the pancreas were identified the following:

- i. a cystic lesion of 4.5×4×4 cm filled with a liquid of low viscosity
- ii. smaller cysts (max diameter: 0.5 cm) around the main cystic lesion described above

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still unknown [4,5]. The main duct IPMN carry a significant higher
reported in the literature but the prevalence of this diffused type is
ducts along with involvement of the main pancreatic duct has been
the pancreatic cystic neoplasms. Multiple involvement of the branch
was a PSC. This is an extremely rare case of simultaneous existence of
confirms that the cyst from which the sample of the FNA was taken
results of the histopathological examination
results of the cytology that followed the initial FNA were indicative
of IPMN whereas the results of the histopathological examination
and MUC5AC vary in small case series [6]. CDX2 and MUC1 seem
to be promising for the identification of IPMN and MCN. Moreover
MUC1 and HER2 have been reported to be key biomarkers in order to
distinguish invasive and non invasive types of IPMN [7].
In spite of the accumulated experience, the morbidity and the
mortality of the pancreatic resections remain high; consequently
the selection of patients with cystic neoplasms for pancreatectomies should
be performed with high level of responsibility and from specialized
surgical teams. The patients with IPMNs or PSCs are individuals with
benign neoplasms at the time of the operation and as always the benefits
should always outweigh the risks.

Conclusion
In this case we presented a very rare type of mixed cystic pancreatic
neoplasm with both IPMN and PSC in the pancreatic specimen.
The lady was treated with extensive distal pancreatectomy. The post
operative course was uneventful regarding gastrointestinal
symptoms and the patient was transfer to a pneumonology clinic due
to deterioration of her respiratory function which was attributed to her
past medical history and exacerbation of the lung aspergillosis.

Discussion
Maybe the most interesting finding in our case is the fact that, the
results of the cytology that followed the initial FNA were indicative
of IPMN whereas the results of the histopathological examination
confirms that the cyst from which the sample of the FNA was taken
was a PSC. This is an extremely rare case of simultaneous existence of
multifocal IPMN and PSC. Multifocality is a novel characteristic of
the pancreatic cystic neoplasms. Multiple involvement of the branch
ducts along with involvement of the main pancreatic duct has been
reported in the literature but the prevalence of this diffused type is
still unknown [4,5]. The main duct IPMN carry a significant higher
risk of malignancy compared to those located in the side branches.
Pancreatic resections have been suggested widely for the managements
of these entities due to the malignant potential. The management of
PSCs is more challenging, and varies in current practice; very few cases
serous cystadenocarcinomas have been described [3]. A diameter of
4 cm or greater and the resulting “pushing” effects in organs, such as
the stomach in our case, have been discussed as the main reasons for
offering patients the choice an operation [1]. The utilization of all the
appropriate imaging techniques during the diagnostic work up, along
with the intraoperative US for remnant cysts can help the relative
complex management of these cases. The available surgical choices at
the moment are the Whipple procedure, as well as the distal, the middle
and the total pancreatectomy. The immunohistochemical staining on
the surgically resected specimens that follows or even on the biopsies
following EUS and FNA can help to differential diagnosis of mucinous
non neoplastic cysts, IPMN and MCN. The expression frequencies
regarding the expression of CK7, CDX2, MUC1, MUC2, MUC6 and
MUC5AC vary in small case series [6]. CDX2 and MUC1 seem
to be promising for the identification of IPMN and MCN. Moreover
MUC1 and HER2 have been reported to be key biomarkers in order to
distinguish invasive and non invasive types of IPMN [7].

Additionally, in the body of the pancreas were found many cysts
with diameter ranging 0.2-1.5 cm, fulfilled with a similar type of liquid.
The microscopical examination revealed: macroscopic variants of
serous cystadenoma in the tail both for the main cyst as well as for
the smaller cysts closely attached, whereas the vast majority of the
cysts in the area of the body were proved to be IPMNs with middle
grade epithelial dysplasia. The immunohistological examination was
performed with CK7, CK19, CEA, CA19-9, CDX2, MUC1, MUC2,
MUC6, MUC5AC and Ki67 which confirmed the abovementioned
findings. All the excised lymph nodes (seven) were negative for
metastasis.
The postoperative course was uneventful regarding gastrointestinal
symptoms and the patient was transfer to a pneumonology clinic due
to deterioration of her respiratory function which was attributed to her
past medical history and exacerbation of the lung aspergillosis.

References