

A Case of Intraductal Papillary Mucinous Neoplasia with Classical Imagings

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Abstract

Nowadays, with the help of improvements in scanning methods and increment of their application frequency, cystic lesions of pancreas especially in asymptomatic people with advanced age are detected with an increasing rate. IPMNs are originated from mucin-producing columnar cells and main pancreatic duct, branch duct or both may be involved in IPMNs. Intraductal Papillary Mucinous Neoplasia is a relatively rare condition that sometimes there is difficulty in diagnosis. Herein we presented an IPMN case with all classical appearances imaging modalities. Especially endoscopic view of the "fish-mouth" sign in ERCP of the patient was quite characteristic.

Introduction

Nowadays, with the help of improvements in scanning methods and increment of their application frequency, cystic lesions of pancreas especially in asymptomatic people with advanced age are detected with an increasing rate [1]. Intraductal Papillary Mucinous Neoplasms (IPMN) which are first described in 1982 are important among cystic neoplasms of pancreas for the clinical course and the difficulty in to diagnose, and they are characterized by cystic dilatations of pancreatic ducts [2]. IPMNs are originated from mucin-producing columnar cells and main pancreatic duct, branch duct or both may be involved in IPMNs [3]. Herein, we report a case of main duct IPMN with characteristic endoscopic views.

Case Report

A 73-year old man was admitted to hospital owing to abdominal pain which is localized in right upper quadrant and epigastric regions, jaundice and itching lasting for 1 month. Vital signs are normal. His skin and sclera were icteric. Among laboratory tests, AST 204(0-35) U/l, ALT 231(0-35) U/l, ALP 575(30-120) U/l, GGT 200(0-55) U/l, total bilirubin 28 mg/dL(0,2-1,2), direct bilirubin 22,1 mg/dL(0-0,2). Amylase and lipase values were normal. Ca19-9 level is 1397 U/ml (0-35).

Abdominal ultrasonography revealed a mass approximately 7 cm in diameter in head of pancreas. The subsequent contrast-enhanced computed tomography revealed 7x6 cm lesion that contains cystic areas in the head of pancreas (Figure 1). In Endoscopic Ultrasonography (EUS) approximately 6 cm sized, heterogeneous cystic areas-containing lesion that involves head and body parts of pancreas was observed. Pancreatic duct dilatation in the proximal part of the mass and lymphadenopathies around the mass were identified (Figure 2). IPMN- characteristic 'fish-mouth' appearance of papilla with a huge amount of mucus exudation is seen in Endoscopic Retrograde Cholangiopancreatography (ERCP) (Figure 3). Upon pancreatography, pancreatic duct is observed with the heterogeneous cystic appearance, short-segment stenosis and proximal dilatation (Figure 4). Following taking fluid from pancreatic duct for cytological examination, 10 F plastic stent was placed. Cholangiography revealed a stenosis at the same level with pancreatic duct. Endoscopic sphincterotomy was performed and 10 F plastic stent was placed also in biliary system.

Cytological examination of the material that sampled during ERCP showed highly degenerated mucinous cell groups with no obvious sign of dysplasia or malignancy. This case was referred to general surgery department for operation.

In the operation, 7x6 cm in dimension, solid, tumoral mass was seen in the head of pancreas (Figure 5). Multiple lymph nodes existed around the coeliac trunk, hepatic artery, and paraaortic region; however vessel invasion or intra-abdominal organ metastasis was not observed. Pathological examination of the specimen was compatible with pancreatic mucinous carcinoma and a large number of dissected lymph nodes were metastatic. This case was diagnosed as IPMN-based pancreatic mucinous cancer and referred to oncology department.

Discussion

It is reported that IPMN consisted of the 5% of all cystic pancreatic lesions and 20-25% of cystic neoplasm's. It is more frequent in males and mean age of onset is 65 years. Frequently, they are located in the head region and tuncate process of pancreas. They secrete a large volume of mucin [4].

Patients generally become symptomatic with abdominal pain that spreads to back and increasing with meal. Acute, recurrent or chronic pancreatitis findings may be found. First-line diagnostic scanning methods are MRCP and thin-section pancreatic protocol CT and they can not only detect tumor but also show relations with adjacent tissues, lymph node spread or metastasis. Both methods are quite sensitive for detecting mural nodules [5,6]. Larger nodules than 3 mm that seen in CT or MRCP means chance of malignancy is high [7].

Endoscopic Ultrasonography signs of main-duct IPMN are diffuse or segmental dilatations on pancreatic duct and intraductal (mural) nodules. Pathological diagnosis can be made by taking sample with EUS-guided fine-needle aspiration biopsy, yet absence of malignant cells does not exclude malignancy [8]. Mucin accumulation or nodular areas in pancreatic duct on ERCP or MRCP, can show filling defects or dilatation zones. Characteristic sign on ERCP is diffuse or segmentally dilated pancreatic duct without stricture, orifice of papilla is opened referred as "fish-mouth" [8]. During ERCP, mucin exudation from pancreatic duct may be seen [9]. The diagnosis is frequently made by intraductal biopsy and pancreatic duct lavage cytology in course of ERCP in addition to other scanning methods.



Figure 1: Contrast-enhanced computed tomography, 7x6 cm lesion that contains cystic areas in the head of pancreas (white arrow).



Figure 2: EUS, approximately 6 cm sized, heterogeneous cystic areas-containing lesion that involves head and body parts of pancreas.

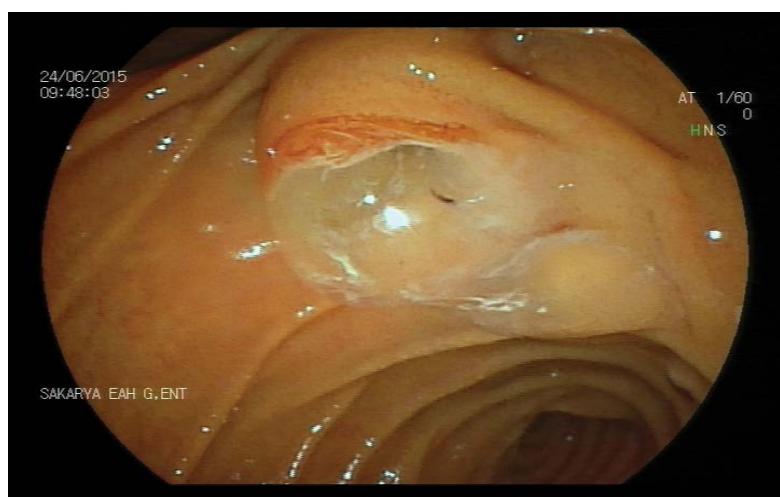


Figure 3: Endoscopic Wiew, IPMN- characteristic 'fish-mouth" appearance of papilla with a huge amount of mucus exudation is seen.

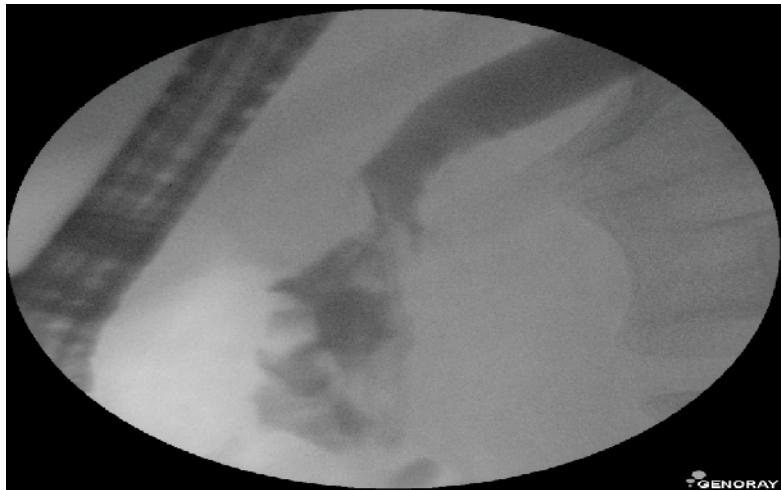


Figure 4: ERCP, Upon pancreatography, pancreatic duct is observed with the heterogeneous cystic appearance, short-segment stenosis and proximal dilatation in the head region.

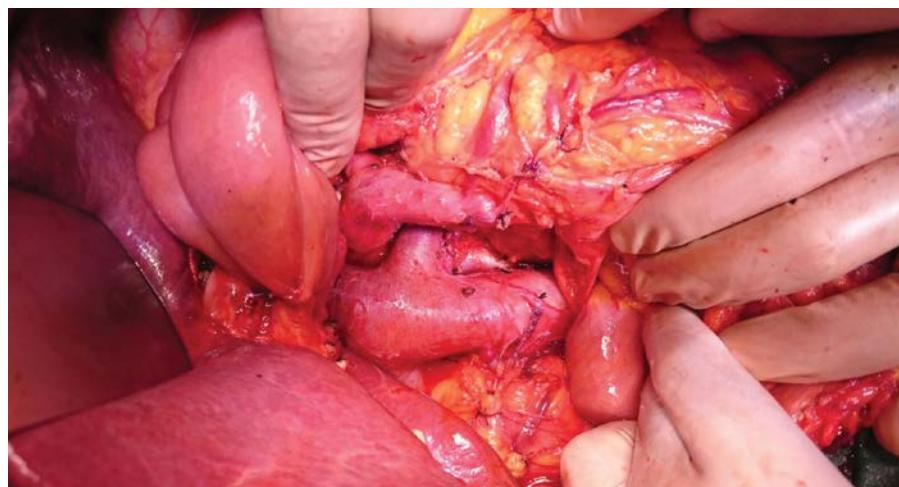


Figure 5: Whipple procedure. In the operation, 7x6 cm dimension, solid, tumoral mass was seen in the head of pancreas.

Larger cyst than 3 cm, presence of mural nodules larger than 1 cm and wider pancreatic duct diameter than 7 mm are in support of malignancy [10]. Positron Emission Tomography (PET) is another method that can be used in detection of malignancy and judgment of operation in matter of IPMN [11]. Levels of CEA and Ca 19-9 can be helpful to differentiate IPMNs as non-invasive one sand invasive carcinomas [12].

Malignancy probability of intraductal papillary mucinous neoplasms is 36-53 % whereas this ratio is 8-36 % in all mucinous cystic neoplasms [13]. Upon the features of the lesion, different surgical procedures that ranges from local excision to pancreaticoduodenostomy are available in the treatment of IPMN [14]. Post-operative adjuvant chemotherapy is suggested to patients who have invasive IPMN, and it is associated with better surveillance in who have positive surgical margins and lymph node involvements [15].

Conclusions

Intraductal Papillary Mucinous Neoplasia is a relatively rare condition that sometimes there is difficulty in diagnosis. Herein we presented an IPMN case with all classical appearances in imaging

modalities. Especially endoscopic view of the “fish-mouth” sign in ERCP of the patient was quite characteristic.

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