Intraductal Papillary Mucinous Tumors of the Pancreas: A Case Report

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Intraductal papillary mucinous tumor of the pancreas is a rare pancreatic cystic tumor which produces and accumulates massive mucin in pancreatic duct. We present a case of a 70-year-old man who suffers from repeated abdominal pain and is initially diagnosed as pancreatitis with pseudocysts. CT and ERCP show a severely dilated pancreatic duct with enhancing mural nodules. Thick mucin is aspirated out under CT guidance, which raises the suspicion of intraductal papillary mucinous tumor of the pancreas.

Key words: pancreas, pancreatic duct, pancreatic cystic neoplasm

Intraductal papillary mucinous tumor (IPMT) of the pancreas is a mucin-producing tumor which arises from epithelial lining of main pancreatic duct and/or the branches of pancreatic ducts [1]. Other terms used to describe this condition are mucin-producing tumor, intraductal mucin-hypersecreting tumor, intraductal papillary neoplasm, mucinous ductal ectasia, etc., but IPMT is most frequently used in recent reports [2]. IPMT can be benign or malignant according to its pathological features, including benign hyperplasia adenoma; borderline low grade dysplasia adenoma; high grade dysplasia adenoma (carcinoma in situ); and malignant adenocarcinoma [3]. We present a patient who suffers from recurrent pancreatitis and does not have history of alcoholism and diabetes. Before surgery, thick mucin is aspirated out from the dilated pancreatic duct under computed tomography (CT) guidance. This finding raise the suspicion of intraductal papillary mucinous tumor of the pancreas.

CASE REPORT

A 70-year-old male suffered from repeated abdominal pain for six months. In each episode, his serum amylase and serum lipase were highly elevated, but he had no jaundice. This patient did not have alcoholism or diabetes. Abdominal CT (fig. 1) showed a diffusely dilated pancreatic duct at pancreatic body and tail with a diameter of 5cm and mural enhanced nodules. The pancreas parenchyma appeared atrophic without significant calcification. The pancreatic duct was so dilated that resembled pseudocysts or cystic neoplasm. The peripancreatic fat in lesser sac and bilateral anterior pararenal spaces appeared clean without infiltration or effusion, that was quite different from typical appearance of pancreatitis. Endoscopic retrograde cholangiopancreatography (ERCP) (fig. 2) showed a severely dilated main pancreatic duct with some filling defects in the lumen. CT-guided drainage of the pancreatic duct was performed. Thick mucin of 800-1000ml/day was drained out. Subtotal pancreatectomy was done. Microscopic findings (fig. 3) proved

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intraductal papillary mucinous tumor of the pancreas with mild to moderate dysplastic change of papillary epithelial cells which is borderline malignant potential. The resected lymph nodes were free of tumor. After surgery, the patient recovered well and was discharged.

**DISCUSSION**

Intraductal papillary mucinous tumor (IPMT) of the pancreas is a mucin-producing pancreatic tumor which originates from epithelial lining of main pancreatic duct or its branches. The term “intraductal papillary mucinous tumor” is used according to the World Health Organization classification system [4] which was first reported by Ohhashi et al in 1982 [1]. The average age at diagnosis is 65 years (range 30-94 years), with a male: female ratio of 1: 2.4. The etiology and risk factors are unknown [5]. Clinical symptoms include pancreatitis-like abdominal pain, diarrhea/steatorrhea and obstructive jaundice which are caused by thick mucinous ductal obstruction and diabetes mellitus [6]. Main pancreatic duct dilatation may be segmental or diffuse. Diameter of the dilated duct usually ranges from 3 to 5cm and may be up to 10cm [7]. Severe ductal dilatation is easily mistaken as cystic neoplasm. Typical CT findings of IPMT are main pancreatic duct dilatation over 2cm in diameter, branches of pancreatic ducts dilatation with cystic-like lesions, pancreatic atrophy with diffuse main pancreatic duct dilatation, enhanced mural nodules and thick mucin in the dilated ducts [8]. ERCP shows segmental or diffuse main pancreatic duct dilatation and/or cystic dilatation of branches of pancreatic ducts. Mucin or tumor is difficult to be differentiated [9]. Since ERCP is an invasive procedure with a risk of sepsis or technical failure, magnetic resonance cholangiopancreatography (MRCP) will be a better choice instead. MRCP can be performed using half-Fourier single shot fast spin echo technique (HASTE) with a high sensitivity up to 93-100% to detect main pancreatic duct and its branches dilatation. MRCP can also detect mural nodules which are hypointense and are different from mucin which are similar with pancreatic juice in signal intensity [5]. Predictive signs of malignancy of IPMT are the presence of diabetes mellitus, gender of male, marked and diffuse dilatation of main pancreatic duct (>10mm), presence of large mural nodules...
(>10mm) and tumor arising from pancreatic head, but none of them are confirmed [10]. Differential diagnosis of IPMT of pancreas include chronic pancreatitis, mucinous cystadenoma, cystadenocarcinoma, serous cystadenoma, pancreatic pseudocyst [5]. Chronic pancreatitis usually occurs in younger patients with alcoholism. CT images show slight main pancreatic duct dilatation combined with calcification and pseudocysts. Mucinous cystadenoma and cystadenocarcinoma are commonly found in middle-aged female. These tumors are located in the body and/or tail of pancreas in 75% of cases. Serous cystadenoma is often located in the head of pancreas with microcystic appearance. However, the diagnosis is still very difficult. The overall prognosis of IPMT is good with a 5 year survival rate of 82% after resection [11],[12].

In conclusion, CT, ERCP and MRCP are very useful tools for diagnose of IPMT, by the presence of severe pancreatic duct dilatation with mural nodules and parenchyma atrophy. CT-guided aspiration of the mucin-like fluid from dilated pancreatic duct would be an important clue to IPMT of pancreas.

REFERENCES

胰臟管內乳頭狀黏液性腫瘤 - 病例報告

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胰臟管內乳頭狀黏液性腫瘤是相當罕見的病例。它能分泌及堆積大量黏液於胰臟內管。本文報告一位70歲男性病患因多次腹痛來求診。臨床初步診斷為胰臟炎及假性囊腫。但經過電腦斷層和經內視鏡逆行性胰管造影術發現胰管明顯擴張，並有管壁結構。電腦斷層為導引引流術抽出膿稠黏液，因此我們高度懷疑此病灶為胰臟管內乳頭狀黏液性腫瘤。

關鍵詞：胰臟，胰管，胰臟囊狀腫瘤