Intraductal Mucin-Hypersecreting Neoplasm of the Pancreas: US, CT and ERP Findings

Ki Yeol Lee, M.D.

Purpose: To evaluate US, CT and endoscopic retrograde pancreatography (ERP) findings of intraductal mucin-hypersecreting neoplasm (IMHN).

Materials and Methods: We categorized an IMHN as belonging to a group of disease that can be clinically detected by the pooling of mucus produced by tumors inside the main duct or branch ducts of the pancreas, thus causing dilatation of these ducts. We retrospectively reviewed the US, CT and ERP findings of five patients (hyperplasia, 4; adenocarcinoma, 1).

Results: On US and/or CT, diffuse dilatation of the main pancreatic duct was demonstrated in five patients. In two cystic lesions corresponding to cystic dilatation of branch ducts were noted at the pancreatic head (n=1) and neck (n=1). In one patient, multiple cystic lesions were seen on CT along the entire pancreas, but these were not detected on US. In all patients, dilatation of the main duct and/or its branch ducts was seen on ERP. Filling defects resulting from mucin in the main duct or branch ducts were noted in all cases. Lymphadenopathy was not seen on US or CT, but carcinoma peritonei was found in the adenocarcinoma patient at the time of operation.

Conclusion: IMHN usually shows smooth, diffuse dilatation of the main duct, and on US or CT, dilated branch ducts are occasionally noted. ERP findings are the most characteristic and conclusive; dilatation of the main duct and/or branch ducts is noted, with a filling defect corresponding to mucin. If overlooked during diagnosis, an IMHN might be interpreted simply as chronic pancreatitis, and so the recognition of this neoplasm is important.

Index Words: Pancreas, radiography
Pancreas, CT
Pancreas, neoplasms

In 1982, Ohhashi et al. (1) described a patient with hypersecretion of mucin from the pancreas caused by a ductal neoplasm and since then, similar tumors have been reported in the literature, particularly by Japanese authors. They were given various names: mucin-producing tumors, mucin-hypersecreting carcinoma, ducctectatic mucinous cystadenoma, cystadenocarcinoma, and intraductal papillary mucinous neoplasms of the pancreas associated with mucinous duct ectasia (2–7). These entities are characterized by marked dilatation of the entire or segmental main duct associated with filling defects during endoscopic retrograde pancreatography (hereafter, ERP), and by the excretion of mucin through the patulous orifice of the enlarged papilla of Vater (major duodenal papilla) during endoscopy (2–4, 8). The disease was found to include not only malignant but also benign cases including hyperplasia and adenoma (3). It was also found that, in some cases, the tumors did not develop only in the main duct, but also in branch ducts (4).

Increasing attention is being paid to the morphology of this tumor, its prognosis, and its relationship with intraductal papillary neoplasms. This study aims to
describe in detail the imaging findings of five IMHN and to discuss their differential diagnosis.

**Materials and Methods**

We categorized IMHN as belonging to a group of diseases that can be clinically detected by the pooling of mucus produced by tumors inside the main or branch ducts of the pancreas, thus causing dilatation of these ducts, and between 1989 and 1996, studied five such cases. A review of ultrasound (US), computed tomography (CT) and ERP files disclosed five patients with diffuse dilatation and filling defects of the main duct and/or cystic dilatation and filling defects of branch ducts.

Two of five patients had undergone partial pancreatectomy, and the remaining three had undergone pancreaticoduodenectomy, opening and closure (due to carcinoma peritonei), and brush cytology respectively. On histologic examination, four cases of hyperplasia and one of adenocarcinoma of the pancreas were identified. Three of these patients were men, and two were women, and they were aged between 40 and 70 (mean, 56) years. Radiologic findings of this IMHN, including US, CT, and ERP were reviewed, as was the clinical data. The radiologic and clinical data relating to four histologically-proven cases of classic mucinous cystadenoma (n=3) or cystadenocarcinoma (n=1) of the pancreas were also reviewed.

**Results**

On US, dilatation of the main duct (Fig. 1) was detected in five patients, and hypoechoic round or ovoid lesions were seen in two; one of these was located at the pancreatic head, and one at its neck.

On CT scans, diffuse dilatation of the main pancreatic duct was seen in five patients (Fig. 1, 2). In one, multiple cystic lesions were seen at the head, body and tail, and the intrahepatic and extrahepatic bile ducts were dilated by invasion of the adenocarcinoma (Fig. 1).

Septum-like structures were detected in two patients, and were due to clustering of the cysts, but mural nodules, a thickened cyst wall, and calcification were not present (Fig. 2). The rest of the pancreas was normal in three patients and atrophied in two patients (Fig. 2); there were no enlarged lymph nodes or obvious metastases in the peripancreatic tissue, but during surgery, carcinoma peritonei was found in the cystadenocarcinoma patient. On CT or US scans, the mucin was not detected.

In four patients, dilatation of the main and branch ducts was seen on ERP and in one, the main duct was dilated with multiple filling defects, without dilatation of the branch ducts (Fig. 2). The maximum diameter of the main duct ranged from 6 to 25 (mean, 9.5 mm); in three cases, it had a smooth contour (Fig. 2) and in two, its appearance was tortuous (Fig. 1). In all patients, filling defects of varying sizes were noted in the main pancreatic duct (Figs. 1, 2) or cystic lesions, the filling defects corresponded to a mucoid substance that was observed endoscopically at the patulous orifice of the papilla of Vater.

The level of serum amylase was not elevated in all patients and diabetes mellitus was detected in two patients. The radiologic and clinical findings of these five cases are presented in Table 1.

**Discussion**

A primary characteristic of a mucin-producing tumor is that it is present in the dilated main or branch ducts of the pancreas. Histologically, the cystic pancreatic duct that contains viscous mucin is lined by tall columnar cells, which often form papillary projections and encroach on secondary ducts. According to the classification of atypicality in the epithelium, this tumor is classified as either hyperplasia without atypia, atypical hyperplasia, adenoma, or adenocarcinoma. Although gross inspection revealed no polypoid or elevated tumor in the main duct or cystic lesions, histologic examination showed neoplastic columnar mucinous epithelium with prominent papillary growth (2).

Itai et al. (9) described ductectatic cystadenoma or adenocarcinoma of the pancreas which is characterized by localized dilatation of branch ducts and the presence of tumor cells, and spreads along the main duct and produces excessive amounts of mucin. According to that report, US and CT demonstrated small cystic lesions, with lobulated or irregular margins located preferentially in the uncinate process, and the main pancreatic duct was dilated. On ERP, localized, prominent cystic dilatation of a branch duct of the main pancreatic duct was seen, and there were grape-like clusters of pear-shaped pools of contrast material associated with filling defects of various sizes. In three of patients, cystic lesions were located at the head, neck and entire pancreas, respectively. Itai et al. also reported five cases of "mucin-producing" carcinoma of the pancreas, which was characterized by smooth, diffuse dilatation of the main duct associated with filling defects on ERP. In contrast to their findings, our
case of mucinous cystadenocarcinoma showed, on ERP, irregular dilatation of the main duct with multiple cysts communicating with this duct along the entire pancreas, and filling defects.

Mucinous obstruction of the biliary tree may develop. A fistulous communication from the cystic lesion to the common bile duct continued to drain copious amounts of mucinous material and invasion of the bile duct and obstruction by a tumor, rather than by the mucinous material may cause biliary obstruction(2, 10). In addition, one of our patients developed biliary dilatation because of carcinomatous invasion of the distal common bile duct. Pancreatic insufficiency may ensue when the lesion further expands in the duct sys-

![Fig. 1. A. Ultrasonogram of oblique view of the pancreas reveals dilatation of the main duct (arrows). B. and (C) CT scan demonstrates multiple ovoid cysts in the entire pancreas (open arrows) and the distal common bile duct is dilated (arrowhead). D. ERP shows marked dilatation of the entire main duct communicating with cystic lesions seen on CT scan with filling defects (double arrows). E. Photomicrograph (H&E; original magnification, ×100) of biopsy specimen shows infiltrative area in the pancreatic parenchyma by gland-forming cancer cells (curved arrows).]
Ki Yeol Lee: Intraductal Mucin-Hypersecreting Neoplasm of Pancreas

Fig. 2. A and B. CT scan shows dilatation of the main duct (small black arrows) and clustered cysts-like lesions in the body of the pancreas (large black arrows).

C. ERP shows extreme dilatation of the main duct with filling defects (open arrows) correspond to the mucin plugs.

D. Pancreaticoduodenectomy specimen with IMHN. Cut surface shows dilated main pancreatic duct (asterisks). D; duodenal mucosa.

E. Photomicrograph (H&E; original magnification, ×400) of the histologic specimen shows papillary hyperplasia featuring papillary projections of tall columnar epithelium. Arrowheads indicate atypical hyperplasia showing pseudostratification of epithelium with pelomorph and dense nuclei.

F. Follow-up CT scan, 2 months after pancreaticoduodenectomy shows remained pancreatic body and tail with still dilated pancreatic duct.
Two of our patients developed pancreatic insufficiency with diabetes, but the level of serum amylase was not elevated. This then increases the amount of mucin to such an extent that it cannot be washed out by normal pancreatic secretion, resulting in permanent duct occlusion with its sequelae, acinar atrophy, pancreatic fibrosis, and exocrine as well as endocrine insufficiency(2).

Because of the well-known malignant potential of the classic type of mucinous cystic neoplasm(mucinous cystadenoma-cystadenocarcinoma), this and IMHN must be clearly distinguished. In contrast to IMHN, which shows no sex predilection, a mucinous cystic neoplasm mainly affects middle aged women(11). It presents as a cystic abdominal mass, preferably located in the tail of the pancreas, and does not usually communicate with the duct system. There have been sporadic case reports of classic cystadenoma or adenocarcinoma in which the lesion communicates with the pancreatic duct, common bile duct, or duodenum(12). In these rare cases, the size of the communicating orifice is, however, small and a solid portion of the cystadenocarcinoma is detected by means of CT or US (2, 3, 5, 9, 10, 13, 14). An IMHN and a mucinous cystic neoplasm may be difficult to distinguished by ERP, and a resected specimen may be required(2). In the four cases of classic cystadenoma or cystadenocarcinoma, there was no communication between the cystic lesion and the duct system.

Chronic pancreatitis can mimic this entity both radiologically and clinically. Common findings include diffuse, corkscrew dilatation of the main duct and branch ducts, with or without filling defects resulting from stones(15, 16). Figure 2 shows a case of hyperplasia with atypia where dilatation of the main duct was smooth on ERP with filling defect; this was different from the finding in chronic pancreatitis. Dilatation in the adenocarcinoma case was seen on US to be beaded, however, and this appearance is also seen in cases of chronic pancreatitis.

Another condition in which findings include marked dilatation of the entire main pancreatic duct is carcinoma of the papilla of Vater, in which the common bile duct is also markedly dilated. Hypotonic duodenography and/or ERP is necessary for differential diagnosis.

It is known that an IMHN has a low potential for malignancy(2, 4, 8, 9); these tumors are clearly distinguished from malignant lesions of the pancreas, such as ductal cell adenocarcinoma, by the absence of invasive tumors or metastasis, medical histories that cover many years, and the fact that patients who undergone an operation live for many years after surgery without evidence of tumor recurrence(2, 3). Yamada et al. (4), however, reported 12 malignant cases in a series of 22 mucin-producing tumors of the pancreas, and our case of mucinous cystadenocarcinoma showed tumoral infiltration of the parenchyma; the bile duct was invaded, and peritoneal seeding occurred. Because the reported number of these tumors is so far very small, however, a larger series of cases is needed if the risk of malignancy is to be indicated more precisely.
In conclusion, US and/or CT scans of IMHN usually demonstrate smooth, diffuse dilatation of the main duct, and dilated branch ducts are occasionally noted. Endoscopic observation is useful for diagnosis; enlargement of the papilla and its patulous orifice, and excretion of mucin are seen in about 90% of patients. ERP findings are the most characteristic and conclusive; dilatation of the main duct and/or branch ducts is noted. Filling defects on ERP are another feature of this entity, and they correspond to mucin or, occasionally, to polypoid tumors. ERP is therefore valuable in differentiating cystic lesions of the pancreas and leads to enables a definitive diagnosis of IMHN.

References

췌장의 관내 점액 과분비성 종양: US, CT와 ERP 소견

이 기 열

목 적: 췌장의 관내 점액 과분비 종양(Intraductal Mucin Hypersecreting Neoplasm [IMHN])의 초음파, 전산화단층촬영과 내시경적 역행성 췌장관 조영술의 소견을 알아보고자 하였다.

대상 및 방법: 저자들은 IMHN을 주췌장관(main pancreatic duct) 혹은 분지관(branch duct)내의 종양에 의하여 점액 저류(mucin pool)가 형성되고 이것에 의하여 췌장관 확장된 질환으로 분류하고 이러한 환자 5명 (hyperplasia : 4, adenocarcinoma : 1)을 대상으로 하여 초음파, 전산화단층촬영과 내시경적 역행성 췌장관 조영술의 소견을 후향적으로 분석하였다.

결 과: 초음파나 전산화단층촬영에서 주췌장관의 확장이 5명에서 관찰되었고, 초음파에서 분지관의 남성확장에 해당하는 저액 저류(mucin pool)가 형성되고 이것에 의하여 주췌장관이 확장된 질환으로 분류하고 이러한 환자 5명 (hyperplasia : 4, adenocarcinoma : 1)을 대상으로 하여 초음파, 전산화단층촬영과 내시경적 역행성 췌장관 조영술의 소견을 후향적으로 분석하였다.

결 론: IMHN은 초음파나 전산화단층촬영에서 주췌장관의 확장 혹은 국한성 남성병변을 보이며, 역행성 췌장관 조영술에서는 주췌장관 혹은 분지관의 확장과 함께 점액에 해당하는 중만 결손을 보인다. IMHN은 진단 당시 간과하여 단순한 만성췌장염으로 진단될 가능성이 많기 때문에 방사선학적으로 인지하는 것이 중요하다고 사료된다.
새로운 국제질서가 만들어지고 급변하는 국제관계속에 아시아 태평양시대의 구조적 역할을 하는 현재의 우리 나라는 경제, 사회 등 모든 분야에 그 위상이 크게 달라져 있습니다. 그중 의학 학술 분야에서는 어느 선진국 못지 않은 세계적인 수준이라 자부할 수 있으며 이제는 배우는 나라에서 배움을 주는 나라로 발전하였습니다.

이에 대한방사선의학회에서는 중국을 포함한 아시아 각국 방사선과 의학도들을 선발하여 학문적 수준과 시설이 나은 우리 나라 연수기관에서 6개월에서 1년 동안 수련시켜 우리 나라 방사선의학의 수준을 세계 여러 나라에 과시하고 나아가 아시아 국가간 협력과 우호증진 및 학술교류에도 기여하고자 합니다. 이에 소요되는 제반 비용은 삼성 GE의료기기의 후원으로 이루어집니다.

여러 수련병원에서는 학회에서 주관하는 이 프로젝트에 적극 참여하시어 대한방사선의학회의 국제화 노력에 협조하여 주시기 바랍니다.

• 참여 희망 신청:
  본학회 사무국 또는 대한방사선의학회 국제협력위원회

• 문의사항:
  대한방사선의학회 국제협력위원회 (위원장: 오용호, 간사: 윤종현)
  전화: (02)224-4362, FAX: (02)476-4719, E-mail: Chyoon@amc.ulsan.ac.kr