CASE REPORT
Gastroenterology & Hepatology

JKMS

DOI: 10.3346/jkms.2010.25.12.1814 • J Korean Med Sci 2010; 25: 1814-1817

Xanthogranulomatous Pancreatitis Combined with Intraductal Papillary Mucinous Carcinoma In Situ

Yo Na Kim[1], Shin Young Park[1], Young Kon Kim[2,3], and Woo Sung Moon[1,3,4]

Departments of Pathology[1], Radiology[2], Institute for Medical Sciences[3], Research Institute of Clinical Medicine and Diabetes Research Center[4], Chonbuk National University Medical School, Jeonju, Korea

Received: 29 April 2010
Accepted: 7 June 2010

Address for Correspondence:
Woo Sung Moon M.D.
Department of Pathology, Chonbuk National University Medical School, 20 Geongi-ro, Geumam-dong, Deokjin-gu, Jeonju 561-180, Korea
Tel: +82.63-270-3086, Fax: +82.63-270-3135
E-mail: mws@chonbuk.ac.kr

This work was supported by the Ministry of Science & Technology (MoST)/Korea Science & Engineering Foundation (KOSEF) through the Diabetes Research Center at Chonbuk National University (R13-2008-005-0000-0).

Xanthogranulomatous lesion is a rare condition in which lipid-laden histiocytes are deposited at various locations in the body. Xanthogranulomatous pancreatitis (XGP) associated with an intraductal papillary mucinous tumor (IPMT) is extremely rare. In this study, we described a case of XGP associated with IPMT and include a review of the literature. A pancreatic cystic mass was detected in a 72-yr-old woman by abdominal computed tomography. Pylorus-preserving pancreaticoduodenectomy was performed and diagnosis of XGP combined with intraductal papillary mucinous carcinoma in situ was made. After 13 months of follow-up, the patient is in good health without any evidence of tumor recurrence. Although XGP associated with IPMT is rare, we suggest that such cases should be brought to the attention of clinical investigators, as it may produce clinical features that mimic pancreatic cancer.

Key Words: Pancreas; Xanthogranulomatous Inflammation; Cystic Tumor

INTRODUCTION

Xanthogranulomatous lesion is a rare condition in which lipid-laden histiocytes are deposited at various locations in the body. Xanthogranulomatous pancreatitis (XGP) associated with an intraductal papillary mucinous tumor (IPMT) is extremely rare. To our knowledge only two cases have been reported in the English-language literature and this is the first reported case in Korea (1, 2).

CASE REPORT

A 72-yr-old woman was referred to our hospital on April 26, 2009 with incidentally detected pancreatic cystic mass. She visited local clinic due to uncontrolled diabetes mellitus which had been treated since 2 yr ago. She had lost 7-kg of body weight over the last 3 months. The physical examination of the patient was unremarkable. The blood count and serum chemistry, including amylase, were within normal limits. Carcinoembryonic antigen (CEA), carbohydrate antigen 19-9 (CA19-9) were also normal. Abdominal computed tomography (CT) scan demonstrated diffuse dilatation of main pancreatic duct with multifocal cysts in pancreatic parenchyma. There was no evidence of calcifying chronic pancreatitis such as pancreatic calcification or multifocal stricture of pancreatic duct. Positron emission tomography (PET) CT showed no fluorodeoxyglucose (FDG) uptaked lesion in pancreas. On magnetic resonance cholangiopancreatography (MRCP), main pancreatic duct was tortuously dilated and multifocal variable sized cysts were accompanied in adjacent pancreatic parenchyma, which was indicative of mixed type of intraductal papillary mucinous neoplasm involving both main and branch duct with retention cysts (Fig. 1A). On fat suppressed T1-weighted image and contrast-enhanced arterial phase MRI, the signal intensity of pancreatic neck portion was focally decreased compared to that of normal pancreas, suggesting inflammatory reaction due to ductal obstruction (Fig. 1B). However, any definite pancreatic mass was not identified. At laparotomy, intraoperative ultrasound (IOUS) was performed, and showed the cystic mass extended to the tail of the pancreas. Pylorus-preserving pancreaticoduodenectomy was performed, and on gross examination, the pancreatic duct showed multilocular cystic formations filled with mucus in the head and uncinate process. Very close to the dilated cystic lesion, a yellow nodular lesion was found to be 1.5×1 cm in size (Fig. 2A). Histologically, these dilated ducts were lined by papillary and micropapillary columnar mucous cells with severe dysplastic changes in the absence of stromal invasion, suggesting the presence of papillary mucinous carcinoma in situ (Fig. 2B). The nodular

pISSN 1011-8934
eISSN 1598-6357

lesion was composed of an aggregation of many foam cells, lymphocytes, and plasma cells (Fig. 2C). Immunohistochemistry for CD68 confirmed that the majority of foamy cells were lipid laden macrophages (Fig. 2D). Final diagnosis of xanthogranulomatous pancreatitis combined with intraductal papillary mucinous carcinoma in situ was made. After 13 months of follow-

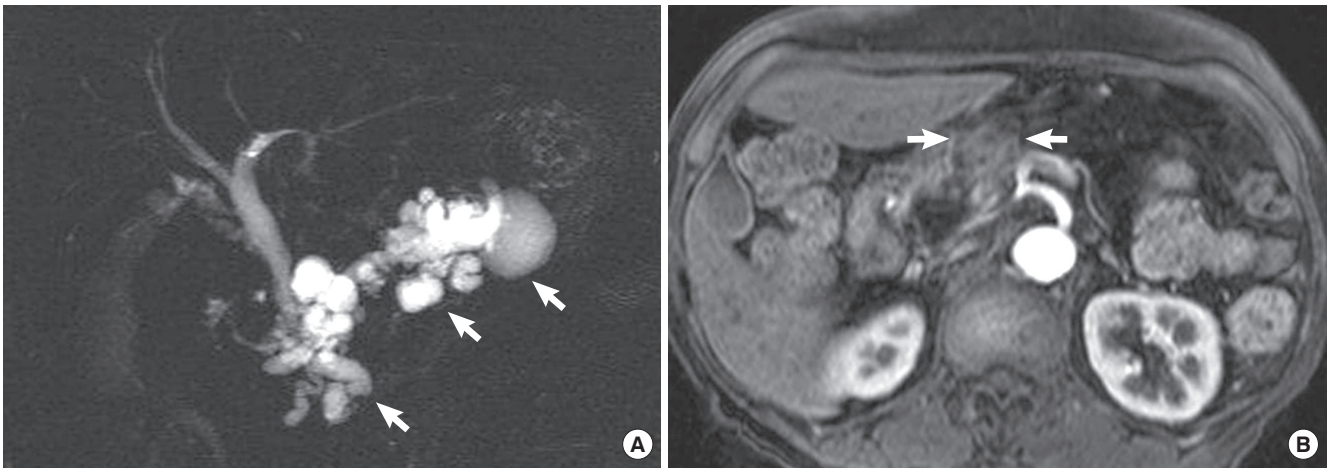

Fig. 1. Images of the pancreatic lesions. (**A**) The coronal single-projection thick-section rapid acquisition with relaxation enhancement MR cholangiography showed a tortuously dilated main pancreatic duct with adjacent variable sized multiple cysts (arrows). (**B**) An axial, pancreatic phase, three-dimensional MRI after the administration of gadopentetate dimeglumine showed an area of ill-defined, focally decreased signal intensity in the pancreatic neck portion (arrows).

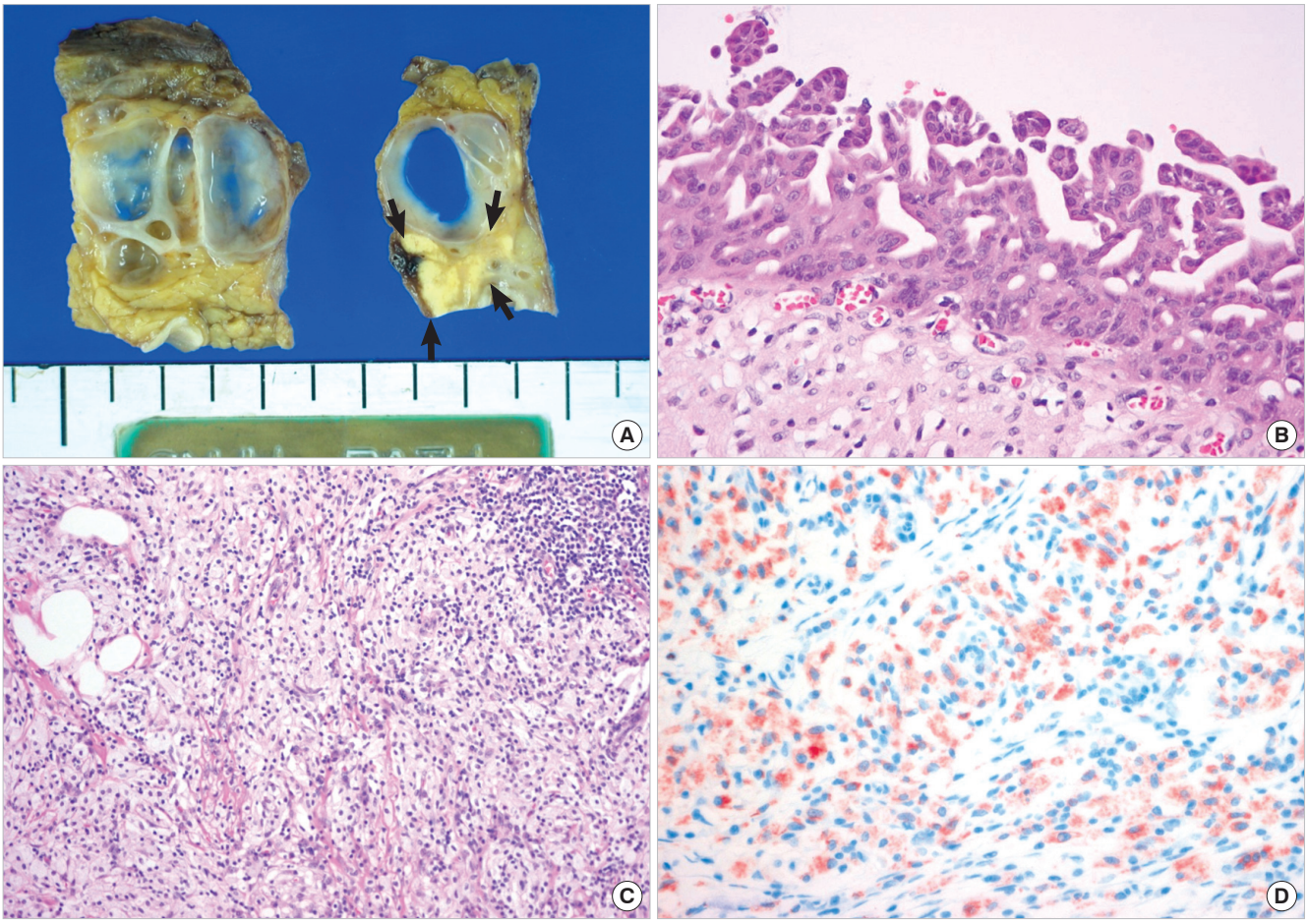

Fig. 2. Pathologic findings of the pancreatic lesions. (**A**) Macroscopically, the pancreatic main duct was dilated, and variable sized-mucin containing cysts and parenchymal yellow nodular lesions were observed (arrows). (**B**) Microscopically, dilated ducts that were epithelized by papillary and micropapillary columnar mucous cells with severe dysplastic change were seen (H&E, ×200). (**C**) The surrounding pancreatic tissue was atrophic and infiltrated with many foamy macrophages, lymphocytes and plasma cells (H&E, ×100). (**D**) Positive CD 68 immunohistochemical staining of the foam cells (×200).

Table 1. Clinicopatholgic features of xanthogranulomatous pancreatitis associated with intraductal papillary mucinous tumor

Cases	Age/Gender	Symptom	Procedure	Associated lesion	XGP lesion
Iso et al. (1)	82/M	Weight loss	Distal pancreatectomy with splenectomy	Intraductal papillary mucinous adenoma	Pancreas, spleen
Kamitani et al. (2)	82/M	Left epigastric pain Pancreatic cystic mass	Distal pancreatectomy with splenectomy Partial gastrectomy	Intraductal papillary mucinous adenoma without malignant change	Pancreas, extended into gastric wall
Present case	72/F	Weight loss Pancreatic cystic mass	Pylorus preserving pancreaticoduodenectomy	Intraductal papillary mucinous carcinoma in situ	Pancreas

XGP, xanthogranulomatous pancreatitis.

up, the patient is in good health without any evidence of tumor recurrence.

DISCUSSION

Xanthogranulomatous lesion is characterized by an aggregation of foamy histiocytes and inflammatory cells. This lesion is considered as a rare variant of chronic inflammation but has been well documented in the gallbladder (3). Xanthogranulomatous pancreatitis associated with IPMT is an extremely rare inflammatory condition. To our knowledge, this is the third case that has been reported of xanthogranulomatous pancreatitis combined with intraductal papillary mucinous tumor being treated with operation (Table 1) (1, 2). Although xanthogranulomatous lesion in the pancreas is a benign condition, excessive surgical resections used to be done in most cases, because of difficulty in distinguishing their condition from pancreatic cancer (1). These characteristic features were described better in the literature about xanthogranulomatous cholecystitis and pyelonephritis (3, 4). In most cases, XGP are clinically and radiologically confused with the carcinoma of the pancreas, because the xanthogranulomatous lesion simulates a malignant tumor, appearing as a poorly defined, yellow, nodular mass and has neoplasm-like properties being capable of local tissue invasion and destruction (1, 2). However, in our case, there was no evidence of any definite mass formation in pancreas based on CT and PET/CT as well as MRI, which was not consistent with macroscopic findings showing nodular mass in surgically resected specimen. We found ill-defined focal decreased signal intensity in pancreas neck on unenhanced- and enhanced T1-weighted image. This area corresponded to the xanthogranulomatous lesion on pathologic correlation. We considered this lesion as early inflammatory reaction of XGP caused by ductal obstruction with mucin leakage. The disagreement between imaging and pathologic findings could be explained by relatively long interval between time of imaging acquisition and operation (35 days). We speculated that xanthogranulomatous inflammation in our case fully developed after imaging study. Clinically, as XGP may resemble pancreatic carcinoma, differentiation is essential by means of intraoperative histological examination to ensure optimal surgical treatment.

Although the mechanism leading to this condition remains unclear, obstructive conditions and infection is commonly considered to be an important factor in xanthogranulomatous inflammation (3-5). In the xanthogranulomatous cholecystitis and xanthogranulomatous pyelonephritis, previous reports have suggested that obstructed lesion, including stone or tumor, induce extravasation of bile juice or urine. It starts as an initial acute inflammatory process followed by a granulomatous reaction and then a cellular-type immulogic response (3). Iyer et al. (6) have reported that obstruction of pancreatic ducts by stone followed by secondary bacterial infection had initiated XGP. Intraductal papillary mucinous tumor of the pancreas is a rare pancreatic cystic neoplasm that arises from the epithelial lining of the main pancreatic duct and/or the branch pancreatic ducts and secrets a thick mucin, which leads to ductal dilatation and obstruction. We speculated that mucin produced by IPMT increased the intraductal and intracystic pressure and that a leakage of mucin into the pancreatic parenchyma produced the xanthogranulomatous changes. Kamitani et al. also reported that increased the intraductal pressure by the mucin produced by IPMT and a subsequent leak of mucin induced the XGP in association with IPMT (2).

In the fact of pancreatic lesions, including pancreatic tumors, pancreatitis, and other lesions that produced ductal obstruction are common, it is unclear why xanthogranulomatous pancreatitis is so rare. Although it is difficult to elucidate the exact underlying mechanisms, accumulation of cases and associated findings should enhance the understanding of the XGP. Although XGP associated with IPMT is rare, we suggest that such cases should be brought to the attention of clinical investigators, as it may produce clinical features that mimic pancreatic cancer.

REFERENCES

1. Iso Y, Tagaya N, Kita J, Sawada T, Kubota K. *Xanthogranulomatous lesion of the pancreas mimicking pancreatic cancer. Med Sci Monit 2008; 14: CS130-3.*
2. Kamitani T, Nishimiya M, Takahashi N, Shida Y, Hasuo K, Koizuka H. *Xanthogranulomatous pancreatitis associated with intraductal papillary mucinous tumor. AJR Am J Roentgenol 2005; 185: 704-7.*
3. Guzmán-Valdivia G. *Xanthogranulomatous cholecystitis: 15 years' ex-*

 DOI: 10.3346/jkms.2010.25.12.1814

perience. World J Surg 2004; 28: 254-7.
4. Eastham J, Ahlering T, Skinner E. *Xanthogranulomatous pyelonephritis: clinical findings and surgical considerations. Urology 1994; 43: 295-9.*
5. Shima Y, Saisaka Y, Furukita Y, Nishimura T, Horimi T, Nakamura T, Tanaka K, Shibuya Y, Ozaki K, Fukui Y, Hamada M, Nishioka Y, Okabayashi T, Taniki T, Morita S, Iwata J. *Resected xanthogranulomatous pancreatitis. J Hepatobiliary Pancreat Surg 2008; 15: 240-2.*
6. Iyer VK, Aggarwal S, Mathur M. *Xanthogranulomatous pancreatitis: mass lesion of the pancreas simulating pancreatic carcinoma-a report of two cases. Indian J Pathol Microbiol 2004; 47: 36-8.*