Primary pancreatic lymphoma (PPL) is a rare extra-lymphatic lymphoma. It is usually found as a large pancreatic mass with no dilatation of the pancreatic duct. Severe dilatations of the pancreatic duct and common bile duct in PPL have not been described. Moreover, internal necrosis in PPL is extremely rare in the literature. Here, we report CT and MRI findings of PPL with severe dilatation of the pancreatic duct, as well as internal necrosis in a 27-year-old man.

Case Report

A 27-year-old man presented with a history of fatigue, jaundice, and weight loss for 20 days. His past medical history revealed that he was a hepatitis B carrier. However, his physical examination was normal without evidence of abdominal tenderness or masses. His serum total albumin, direct albumin, aspartate amino transferase (AST), and alanine amino transferase (ALT) were elevated; however, his serum amylase and lipase levels were normal, as were other laboratory tests including leukocyte counts. Contrast enhanced multi-detector row CT (MDCT) showed a 5 cm long diameter and poorly enhancing mass containing a low attenuated area in the pancreas head, with moderate to severe dilatation of the pancreatic duct, common bile duct, and both intrahepatic bile ducts (Fig. 1A). Lymphadenopathy was not demonstrated; however, displacement of portal vein, IVC, and duodenum was depicted. Abdominal MR imaging was performed for the purpose of planning surgery on a 3T MRI unit using a phased array coil. T1-weighted MR images showed a hypointense homogeneous mass with an internal hyperintense area (Fig. 1A). Lymphadenopathy was not demonstrated; however, displacement of portal vein, IVC, and duodenum was depicted. Abdominal MR imaging was performed for the purpose of planning surgery on a 3T MRI unit using a phased array coil. T1-weighted MR images showed a hypointense homogeneous mass with an internal hyperintense area (Fig. 1B). T2-weighted MR images demonstrated a hyperintense mass with a hyperintense area, as well as severe dilatation of the pancreatic duct (Fig. 1C). A hyperintense area on T1- and T2-weighted MR images was considered as hemorrhage within the mass. However, gadolinium-enhanced T1-weighted MR images showed homogeneous
enhancement (Fig. 1D). ERCP demonstrated severe dilatation of the distal pancreatic duct, both intrahepatic bile ducts, and the common bile duct (Fig. 1E). The preoperative diagnosis was a neuroendocrine tumor or pancreatic ductal adenocarcinoma due to severe dilatation of the pancreatic duct and common bile duct as well as the internal heterogeneous area. An endoscopic US-guided biopsy was performed, but failed. We decided to

Fig. 1. Primary pancreatic lymphoma in a 27-year-old man. 
A. Contrast enhanced MDCT shows poorly enhancing mass (arrowheads) of 5 cm in diameter with low internal attenuation (curved arrow) in the pancreas head, with moderate dilatation of the pancreatic duct (arrow) and both intrahepatic bile ducts. 
B. T1-weighted MR image shows a hypointense homogeneous mass (arrowheads) with an internal hyperintense area (curved arrow) and severe dilatation of the pancreatic duct (arrow). 
C. T2-weighted MR image demonstrates a hyperintense mass with an internal hyperintense area (curved arrow) and severe dilatation of the pancreatic duct (arrow). 
D. Gadolinium-enhanced T1-weighted coronal MR image shows a homogeneous enhancing mass (arrowheads) with a non-enhancing area (asterisk) demonstrating severe dilatation of the distal pancreatic duct (curved arrow) and common bile duct (arrow). 
E. ERCP demonstrates moderate to severe dilatation of the upstream pancreatic duct (arrowhead), both intrahepatic bile ducts, and the common bile duct (arrow).
perform surgery because of the patient's young age and high probability of malignancy on the imaging study. A pylorus preserving pancreaticoduodenectomy was performed, without preoperative chemotherapy.

Upon gross inspection of the surgical specimen, a well circumscribed whitish mass was identified with the narrowing of the downstream pancreatic duct and common bile duct. The tumor invaded the pancreatic duct and duodenal wall (Fig. 2A). A microscopic examination showed focal, well-circumscribed coagulative necrosis within the tumor, considered as hemorrhage on a preoperative MRI (Fig. 2B). The tumor cells displayed a large, round to oval shaped nucleus, scanty cytoplasm, and prominent nucleoli. Immunostaining results indicated that the tumor cells showed positive immunoreactivity for CD20 and CD79a, and negative immunoreactivity for cytokeratin, CD45RO, and CD3 (Fig. 2C). The tumor was pathologically confirmed as malignant diffuse large B-cell lymphoma. The patient had received postoperative adjuvant CHOP (cyclophosphamide, doxorubicin, vincristine, and prednisone) chemotherapy and has been living without evidence of tumor recurrence over 17 months after surgery.

Discussion

PPL is an extremely rare disease which occurs in pancreatic situ, with or without involvement of the peripancreatic lymph nodes [1]. The diagnostic criteria for PPL include: 1) no palpable superficial lymphadenopathy, no mediastinal lymphadenopathy on chest radiography, 2) a normal leukocyte count in peripheral blood, 3) a main mass in the pancreas with lymph nodal involvement confined to the peripancreatic region, and 4) no hepatic
or splenic involvement (2). Most cases are actually pri-
mary low-grade non-Hodgkin’s lymphoma of the B-cell
type. Our case was pathologically confirmed as mali-
gnant diffuse large B-cell lymphoma.

On CT, there are two different morphologic patterns
of pancreatic involvement in patients with PPL. The for-
er is a localized, well circumscribed tumoral form, and
the latter is a diffusely enlarged infiltrating or replacing
form occupying most of the pancreatic gland (3). In a
well circumscribed tumoral type, an MRI shows a ho-
monogeneous hypointense mass within the pancreas on
T1-weighted images with subtle enhancement after
gadolinium enhancement, as well as a heterogeneous
mass with low to intermediate signal intensity on T2-
weighted images. As for a diffuse infiltrating tumor
type, T1- and T2-weighted images showed hypointense
enlarged pancreas, with mild to moderate enhancement
after gadolinium enhancement (4). In our case, the mass
was also homogeneous on CT and MRI with mild homo-
genous enhancement, but the mass contained a necrot-
ic area as a hypopattenuation on CT and a hyperintense
area on T1- and T2-weighted MR images.

Internal hypopattenuation on CT and a hyperintense
area on T1- and T2-weighted MR images were consid-
ered as hemorrhage within the mass. The presences of
calcification or necrosis are reliable findings for ruling
out non-Hodgkin’s lymphoma (5). However, a small het-
erogeneous area which was necrosis within a tumor can
be seen in an isolated case (6). In our case, focal necrosis
within the mass was shown on CT and MRI. This find-
ing was very unusual in untreated non-Hodgkin’s lymph-
oma and made the diagnosis of PPL difficult.

Unlike pancreatic ductal adenocarcinoma, moderate
to severe dilatation of the pancreatic duct is appar-
etly extremely rare in PPL because the pancreatic duct is ei-
ther normal, displaced, or simply narrowed in patients
with PPL (4). Van Beers B et al. (3) reported that dilata-
tion of the pancreatic duct can be usually mild, with a
ratio of duct diameter to distal gland width invariably
less than 0.5 in PPL. Severe dilatation of the pancreatic
duct and common bile duct in our case was demonstrat-
ed on CT and MRI. A dilated pancreatic duct and com-
non bile duct on CT and MRI led us to preoperatively
diagnose a neuroendocrine tumor or pancreatic ductal
adenocarcinoma.

During the surgical procedure, the hard mass with in-
ternal necrosis was seen at the pancreatic head along
with moderate to severe dilatation of common bile duct
and pancreatic duct. The hardness of the tumor and the
presence of internal necrosis suggested a rapid growth
rate and were considered as cause of the dilatation of
the common bile duct and pancreatic duct, which was
different to usual pancreatic lymphoma.

The differential diagnosis of localized PPL in our case
included pancreatic ductal adenocarcinoma and neu-oendocrine tumors such as a large islet cell tumor. The
differential diagnosis of PPL from the pancreatic ductal
adenocarcinoma is very important. For the pancreatic
ductal adenocarcinoma, the primary treatment is con-
sidered to be surgical excision; but in PPL, the primary
treatment is nonsurgical, based on chemotherapy alone
or a combination of chemotherapy and radiation ther-
apy (1). Ductal adenocarcinoma commonly infiltrates in-
to pancreatic parenchyma, peripancreatic fat, as well as
adjacent structures, and dilates the more distal pancreat-
ic duct when the more proximal ductal invasion has tak-
en place. Also CT findings of ductal adenocarcinoma
typically manifest themselves as a lesion that is some-
what inhomogeneous and hypopattenuating relative to
the normally enhancing pancreatic parenchyma.
Neuroendocrine tumors such as the large non-function-
ing islet cell tumor demonstrate hypervascular solid
components, usually in the periphery, and central non-
enhancing areas on CT, which may represent necrosis,
fibrosis, or cystic degeneration (7). Also, islet cell tumors
usually show early strong enhancement on early-dy-
namic contrast CT or MRI as well as frequent hepatic
metastasis (8). However PPL usually demonstrates poor
but homogeneous enhancement (9).

In summary, we present a rare case of PPL with un-
usual imaging findings. Although a bulky homogeneous
mass with moderate to severe dilatation of the pancreat-
ic duct and focal necrosis within tumor is more com-
monly seen in pancreatic ductal adenocarcinoma or a
neuroendocrine tumor, PPL must be included in the dif-
ferential diagnosis.

References
1. Lin H, Li SD, Hu XG, Li ZS. Primary pancreatic lymphoma:
2. Dawson IM, Cornes JS, Morson BC. Primary malignant lymphoid
tumours of the intestinal tract. Report of 37 cases with a study of
Assist Tomogr 1993;17:94-97
4. Merkle EM, Bender GN, Brambs HJ. Imaging findings in pancreat-
ic lymphoma: differential aspects. AJR Am J Roentgenol 2000;174:
671-675
심한 췌관확장을 동반한 원발췌장림프암: 증례 보고

1 전남대학교 의과대학 영상의학교실, 전남대학교병원 영상의학과
2 전남대학교 의과대학 영상의학교실, 화순전남대학교병원 영상의학과
3 전남대학교 의과대학 병리학교실, 전남대학교병원 병리과

허태욱 ∙ 김진웅2 ∙ 하숙희2 ∙ 신상수 ∙ 정용연2 ∙ 강형근2 ∙ 최유덕3

췌장의 원발림프암은 매우 드물어 특징적인 영상소견은 잘 알려져 있지 않으나 췌관염중과는 달리 췌관의 확장
이 드물고 종괴 내부에 괴사를 잘 동반하지 않는다고 알려져 있다. 우리는 심한 췌관 및 총담관의 확장과 종괴 내 괴
사를 동반하였던 27세 남자의 원발췌장림프암의 전산화단층촬영(CT) 및 자기공명영상(MR)소견을 보고하고자 한
다.