CASE REPORT
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A Case of Primary Pancreatic Lymphoma Presenting with Obstructive Jaundice

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A 55-year-old man was admitted to the hospital for jaundice. Computed tomography (CT) scans showed a diffuse mass in the pancreas and peripancreatic area, with infiltration to of the whole pancreas, and overall reduced enhancement compared to normal pancreas. Esophagogastroduodenoscopy revealed elevated mucosal lesion covered hyperemic mucosa at duodenal bulb and ulcerative lesion at body of stomach. Endoscopic ultrasonography revealed an irregular mass with unclear boundaries was observed within the pancreas. Abrupt narrowing of mid to distal common bile duct was seen and the stricture was caused by compression of pancreatic mass. Plastic stent was inserted and clinical improvement was achieved including resolution of jaundice. The patient is currently being treated with combination of rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisolone. We report a case of primary pancreatic lymphoma presenting with obstructive jaundice.

Key Words: Pancreatic mass, Primary pancreatic lymphoma, Obstructive jaundice

INTRODUCTION

Pancreatic lymphoma is a rare disease that accounts for about 1-2% of all pancreatic malignancies and less than 1% of extranodal non-Hodgkin's lymphomas.[1,2] It usually presents as a secondary lymphoma, and is the result of the direct extension of peripancreatic lymphadenopathy, whereas primary pancreatic lymphoma (PPL) of pancreatic origin is extremely rare, and occurs in less than 0.5% of cases.[3] Pancreatic lymphomas occur commonly in immunocompromised hosts, such as patients infected with the human immunodeficiency virus or transplant recipients.[2] Unlike pancreatic cancer, PPL is not usually accompanied by obstructive jaundice.[4] Nevertheless, the authors encountered a case of PPL accompanied by obstructive jaundice in a non-immunocompromised host, and thus report it together with a related literature review.

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CASE REPORT

A 55-year-old man was admitted to the hospital for jaundice. The patient presented with a non-specific medical history and reported intermittent abdominal discomfort one month before admission. He was placed on medical observation, and special examinations were not performed. Approximately 7 days before admission, he observed the onset of red-colored urine, generalized itching, and jaundice. Although he complained of weakness on admission, fever and chills were not observed. He reported overall abdominal discomfort without local and rebound tenderness. A firm mass was palpated in the epigastric area. The patient reported a weight loss of about 7 kg in the last 3 months.

A blood test performed at admission revealed a white blood cell (WBC) count of 6,100 (neutrophil 14.5%, lymphocyte 72%), hemoglobin of 12.6 g/dL, platelet count of 331,000/mm^3, total bilirubin of 37.2 mg/dL, direct bilirubin of 34.1 mg/dL, aspartate aminotransferase (AST) of 203 IU/L, alanine aminotransaminase (ALT) of 237 IU/L, alkaline phosphatase (ALP) of 1,197 IU/L, γ-glutamyltranspeptidase of 1,113 IU/L, lactate dehydrogenanse of 730 mg/dL, amylase of 110 U/L, lipase of 201 U/L, carbohydrate antigen 19-9 (CA 19-9) of 236 U/mL, as well as the absence of HBs Ag, HCV Ab, HIV Ab, and the presence of anti-HBs Ab.

Computed tomography (CT) scans showed a diffuse mass in the pancreas and peripancreatic area, with infiltration to of the whole pancreas, and overall reduced enhancement compared to normal pancreas. Furthermore, since the bile duct was compressed by the mass, the intrahepatic bile duct was dilated and some peripancreatic lymph nodes were enlarged. However, distant lymph node enlargement was not observed, and there were no specific findings in the intraperitoneal organs such as the liver and spleen (Fig. 1 A and B).

Esophagogastroduodenoscopy revealed elevated lesions in the duodenal bulb with relatively clear surrounding boundaries, which were covered with hyperemic mucosa. Ulcerative lesions were found in the greater curvature of the body of the stomach along with some hematins in the ulcer base. Bleeding was not observed (Fig. 2 A and B).

Endoscopic ultrasonography revealed overall thickening of the muscularis mucosa layer in the ulcer lesion of stomach and invasion of the submucosa and proper muscle layers. An irregular mass with unclear boundaries was observed within the pancreas and the main pancreatic duct of the body was measured up to 2.4 mm (Fig. 3 A and B).

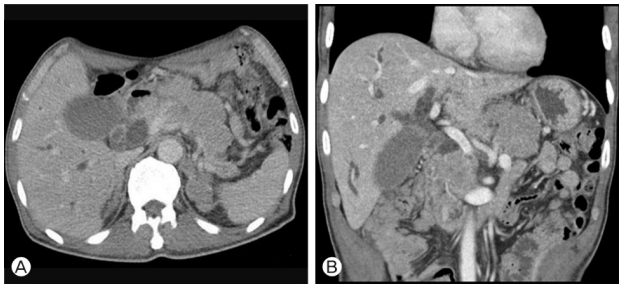

Fig. 1. Abdominal computed tomography (**A**) diffuse mass in the pancreas and peripancreatic area, with infiltration to the whole pancreas (**B**) the bile duct was compressed by the mass, the intrahepatic bile duct was dilated.

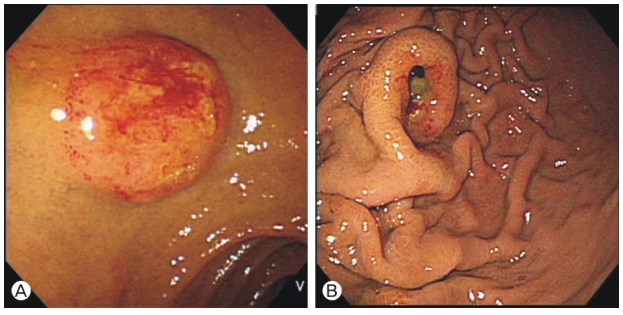

Fig. 2. Esophagogastroduodenoscopy (**A**) elevated mucosal lesion covered with hyperemic and friable surface (**B**) ulcerative lesion was found at body of stomach and some hematin in the base of ulcer.

The patient underwent endoscopic retrograde cholangiopancreatography in order to relieve the obstructive jaundice. Since the cholangiogram revealed abrupt narrowing and upstream dilatation of the mid to distal common bile duct, a plastic stent (Cotton-Leung® 10 French 7 cm) was inserted (Fig. 4 A and B). The liver function subsequently improved, resulting in a reduction in bilirubin levels.

Biopsy samples of the lesions in the stomach and duodenum showed relatively abundant cytoplasm and multiple lymphocytes with 2-4 distinct nuclei. Strong positive staining for CD 20 was observed on immunohistochemistry, and the patient was diagnosed with diffuse large B cell lymphoma (Fig. 5 A and B).

The patient is currently being treated with the R-CHOP

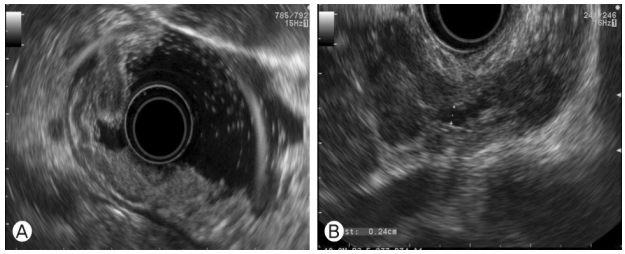

Fig. 3. Endoscopic ultrasonography (**A**) overall thickening of the muscularis mucosa layer in the ulcer lesion of stomach and invasion of the submucosa and proper muscle layers (**B**) An irregular mass with unclear boundaries was observed within the pancreas and the main pancreatic duct of the body was measured up to 2.4 mm.

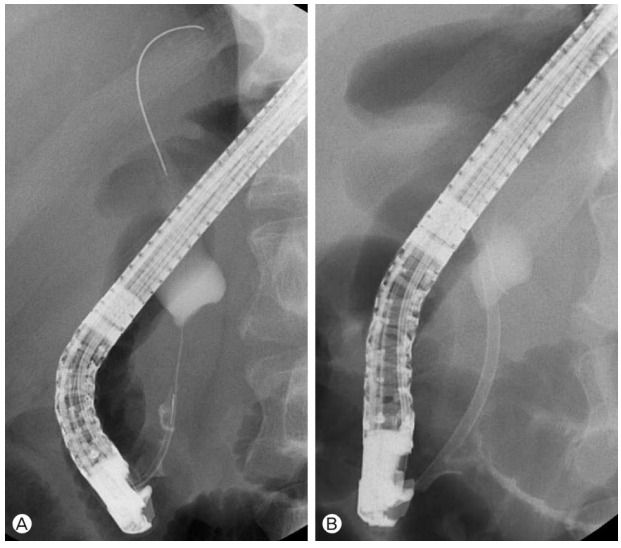

Fig. 4. Endoscopic retrograde cholangiopancreatography (**A**) abrupt narrowing and upstream dilatation of the mid to distal common bile duct (**B**) a plastic stent was inserted across the ampulla.

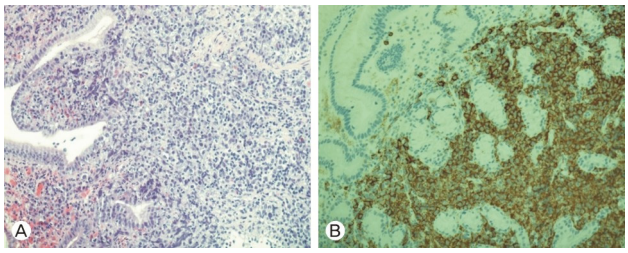

Fig. 5. Microscopic findings (**A**) showed relatively abundant cytoplasm and multiple lymphocytes with 2-4 distinct nuclei (**B**) strong positive staining for CD 20 was observed on immunohistochemistry.

regimen (combination of rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisolone) and regular follow-up visits have been scheduled.

DISCUSSION

PPL is an extremely rare disease, which is known to occur in the pancreas in situ. The clinical manifestation and imaging findings are similar to those of other pancreatic mass lesions such as pancreatic cancer. However, in contrast to patients with pancreatic cancer, the mean age of patients with PPL was 57.5 years. Additionally, PPL is more common in men (male to female ratio of 13:3) and potentially treatable, even when detected at a later stage.[6] In this case, the patient was a 55-year-old man.

Patients with PPL may have a variety of clinical presentations. Although approximately 83% and 58% of all patients with PPL complain of abdominal discomfort and a palpable mass, respectively, some present with other symptoms including jaundice, weight loss, bowel obstruction, and diarrhea.[6] In this case, the mass was palpated in the epigastric area, and symptoms such as abdominal discomfort, jaundice, and weight loss were reported. In contrast, fever, chills, and night sweats, which are commonly seen in patients with non-Hodgkin's lymphoma, are not common in patients with PPL. This patient did not also complain of such symptoms.[6]

Blood tests alone have limited value in the diagnosis of PPL. As demonstrated by this case, if symptoms are accompanied by bile duct obstruction, both CA 19-9 and total bilirubin can be simultaneously elevated in cases of PPL and difficult to differentiate from malignant obstruction resulting from cancer of the pancreatic head. However, imaging findings may show marked differences between them. While pancreatic cancer commonly invades surrounding major vessels as it increases in size, PPL predominantly shows peripancreatic lymph node enlargement rather than the invasion of the surrounding vessels. In addition, unlike pancreatic cancer, metastasis of the liver or spleen is rarely observed in cases of PPL.[4]

Similar to the other carcinomas, the final diagnosis of PPL is established by a histopathological examination. In most cases, the probability of a definite diagnosis is highest when tissues are obtained surgically. However, the relatively less invasive endoscopic ultrasonography guided fine needle aspiration (EUS-FNA) procedure has been commonly used in recent years. Nevertheless, EUS-FNA is limited by the false-negatives that are frequently generated because of the lesser amount of tissues obtained by EUS-FNA than those from surgical procedures.[7] Since synchronous lesions were found in the stomach and duodenum in this case, endoscopic forcep biopsy was performed, and histopathological confirmation was obtained without additional EUS-FNA.

Although surgery is usually performed when the definite diagnosis of PPL is not established, chemotherapy using a com bination of rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisolone known as R-CHOP regimen is performed in most cases. Chemotherapy is effective for the treatment of PPL and approximately 78% of patients treated with chemotherapy did not have any evidence of disease 34 months after treatment.[8] Since this patient is being treated with R-CHOP, we will evaluate the response to chemotherapy in the future.

In general, the prognosis of non-Hodgkin's lymphoma is evaluated based on the international prognostic index (IPI).[9] This patient presented with three clinical risk factors including LDH level elevation, Ann Arbor stage IV, and one1 more site of extranodal involvement, which identifies him as having a high intermediate risk and a 43% 5-year survival rate. Therefore, careful follow-up visits are required in the future.

SUMMARY

PPL is a rare non-epithelial tumor, which occurs in the pancreas. Although imaging findings are helpful in the diagnosis of PPL, they are often difficult to differentiate from pancreatic cancer. Pancreatic cancer is commonly associated with obstructive jaundice, while PPL occurs in the head of pancreas, and is rarely accompanied with jaundice. However, in some cases, they can present with obstructive jaundice and CA 19-9 elevation. Therefore, EUS-FNA has been widely used for histopathological confirmation in recent years. However, because of the lesser amount of tissues that are obtained with this method, establishing a definite diagnosis is difficult. In these cases, confirming the presence of synchronous lesions in the stom-

ach and duodenum, where endoscopic forcep biopsy can be performed, is important. The authors encountered a case of primary pancreatic lymphoma accompanied by obstructive jaundice, and now reported it along with review of the related literatures review.

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