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# CASE REPORT

## Primary Pancreatic Lymphoma - A Diagnostic Dilemma

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**ABSTRACT**

Primary pancreatic lymphoma (PPL) is a rare form of extranodal lymphoma, mimicking the carcinoma of the head of pancreas clinically [1]. Most PPLs are non-Hodgkin's lymphomas (NHLs), predominantly the diffuse large B-cell type [2]. We present here, a case of primary pancreatic non-Hodgkin's lymphoma, presenting with abdominal pain and jaundice and CT scan features suggestive of cholangiocarcinoma. The patient underwent surgical resection of the tumour and the histopathology revealed the primary involvement of the pancreas by the lymphoma cells. Immunohistochemistry confirmed the diagnosis as diffuse large B-celltype of lymphoma. The patient refused chemotherapy postoperatively and expired after 6 months of follow up.

**Key Words**: pancreas, non-Hodgkin's lymphoma (NHL), primary, B-cell, adenocarcinoma

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### Introduction

Malignant lymphomas arising primarily from the pancreatic parenchyma or in the peripancreatic lymph nodes are extremely rare. These are mostly NHLs, the low grade B-cell types [1], [2]. PPL accounts for 1% of all extranodal NHLs and 0.7% of all pancreatic malignancies [3],[4]. Clinically, most PPLs are misdiagnosed as pancreatic adenocarcinomas. It is important to distinguish between PPL and pancreatic adenocarcinoma due to a marked difference in the treatment and prognosis of the two diseases. We hereby present a case of PPL diagnosed as diffuse large B-cell type of NHL.

### Case Report

A 62 year old male presented with pain in the abdomen, jaundice, anorexia, nausea and vomiting of a duration of 4 months. Per abdomen examination revealed mass and tenderness in the right hypochondrium. No palpable lymphadenopathy was observed. Chest X-ray did not reveal any hilar or mediastinal lymphadenopathy. Routine blood investigations revealed normal WBC count and morphology (TLC: 4,300 cells/$mm^3$, DLC: $P_{74}L_{19}M_6E_1$). The liver function test was deranged with total bilirubin -6.4mg% and direct bilirubin -6.0mg%. Serum SGOT, SGPT and Alkaline phosphatase were 58.0 IU/L, 56.0IU/L and 886.0IU/L respectively, revealing elevated levels. S. Amylase was elevated- 267.0 U/L (reference values: 0-220 IU/L). CA 19-9 was within the normal range -32.8µ/ml. Serology was non-

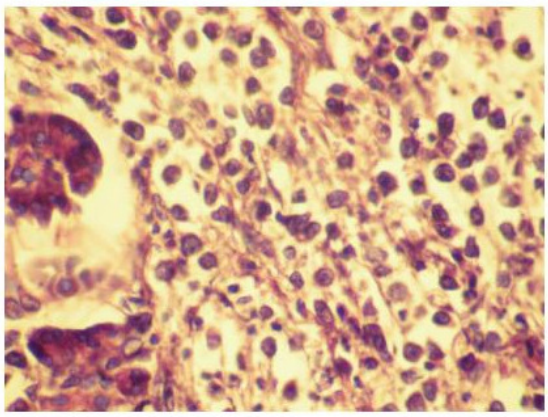
reactive for HIV and the Hepatits B surface antigen.

CT scan revealed an ill-defined irregular high attenuating lesion in the distal common bile duct and the diffuse enlargement of the head of pancreas, with dilatation of the proximal common bile duct and the hepatic duct on contrast enhancement. No obvious peripancreatic lymphadenopathy was observed on CT scan and the diagnosis of cholangiocarcinoma was made, based on the findings of imaging. The patient was posted for surgery and the Classical Whipple's procedure was performed, including antrectomy and pancreatoduodenectomy under general anaesthesia, augmented with epidural analgesia. Peroperatively, no definite lymphadenopathy was observed. The superior mesenteric artery, the superior mesenteric vein and the portal vein appeared unremarkable. The pancreatoduodenectomy specimen, peripancreatic lymph nodes and the gall bladder were sent for histopathological examination.

## Pathological findings

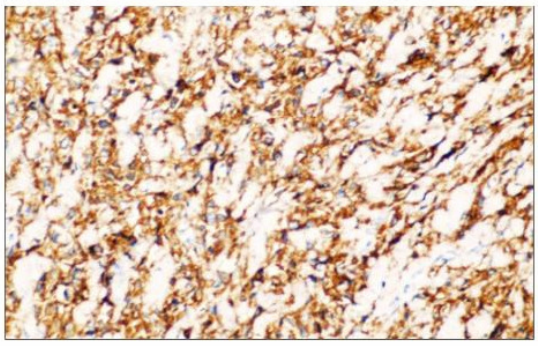
**Gross:** The specimen consisted of the duodenum measuring 8.5cm in length, the pancreas measuring 3.8x3.2x2cm, a part of the stomach measuring 3cm and the gall bladder measuring 8x3cm. Grossly, the head and a part of the body of the pancreas appeared to be diffusely enlarged, with no abnormality in other tissues. On opening the specimen, it was observed that the common bile duct was thickened and dilated. A total of 13 lymph nodes were identified grossly in the peripancreatic tissue, the largest measuring 0.8x0.6x0.5cm.

**Microscopic examination**: Multiple sections from the pancreas revealed a lymphoproliferative disorder [Table/Fig 1]. The individual cells were large, round, having large nuclei with a high N:C ratio and having irregular nuclear membranes and clumped chromatin.



**(Table/Fig 1) 40x (H&E) Microphotograph showing normal pancreatic acini infiltrated by lymphoma cells**

The gall bladder showed features of cholecystitis. Sections from the duodenum and the stomach were unremarkable. The lymph nodes showed features of reactive lymphoid hyperplasia. The tumour cells expressed CD-20 positivity [Table/Fig 2] and were nonreactive for CD-3.



**(Table/Fig 2) Immunohistochemical analysis showing CD20 positive staining of cells, consistent with B-cell origin (× 200).**

A final diagnosis of Primary diffuse large B-cell type of non-Hodgkin's lymphoma was made, based on Behrn's criteria and immunohistopathology reports. The patient was advised chemotherapy but he refused further treatment due to his poor financial status. He expired 6 months after surgery.

## Discussion

Malignant lymphomas infrequently involve the pancreas. The estimated frequency of primary non-Hodgkin's lymphomas of the pancreas is about 1% [3]. The head of the pancreas is the most common location of pancreatic involvement in PPLs [1] and the diffuse large B-cell type is the most common type [3]. T cell lymphomas are very uncommon and carry a poor prognosis [5]. Clinically, they are confused with the more common pancreatic adenocarcinomas. A frequent occurrence of B-cell lymphoma has been reported in association with acquired immunodeficiency syndrome in recent years, though our patient was sero-negative for the Human immunodeficiency virus by the ELISA test.

In the review of literature, about 87% of cases of pancreatic lymphoma were found to be misdiagnosed clinically and radiologically as pancreatic adenocarcinoma, because there was no marked difference between the two, as regards to age, sex or symptoms. However, unlike carcinomas, PPLs are treatable and carry a comparatively better prognosis even if they are not detected at an early stage [3]. Till date, nearly 150 cases of PPLs have been reported in literature, with a strong male predilection (male:female ratio is 13.3:1) and increasing age (mean age of 57.5 years), a profile similar to that of our patient [2],[6].

Behrn's et al [7] suggested the following criteria for the diagnosis of primary pancreatic lymphoma; 1) Tumour confined within the pancreas at the time of diagnosis and investigations failed to reveal any evidence of lymphoma elsewhere; 2) Lymphadenopathy confined to only the peripancreatic group of lymph nodes without any involvement of distant groups or superficial lymph nodes.No hepatic or splenic involvement; 3) Normal leucocyte count with no abnormal cells in peripheral blood and bone marrow

In our case, the tumour tissue was limited to the pancreas without any lymph node involvement or distant metastasis. The total and differential WBC counts were also within normal limits on three separate occasions, thus pointing to primary pancreatic pathology.

Data reveal that the S. carbohydrate 19-9(CA 19-9) levels in PPL patients are normal or slightly elevated, while in cases of pancreatic adenocarcinoma, they are higher in almost 80% patients [2]. In our case, CA 19-9 was within the normal range.

Imaging plays a key role in the diagnosis of PPL. CT scan is the most preferred imaging technique which is used in the detection and the characterization of pancreatic tumours. The image of PPL resembles that of carcinoma on CT scan, like the enlargement of the pancreatic head and density changes [2]. However, the presence of a well circumscribed mass in the pancreas without the involvement of the pancreatic duct and the enlargement of the surrounding lymph nodes rule out the diagnosis of pancreatic adenocarcinoma [8],[9]. Arcari et al [10] insisted that the final diagnosis of PPL should be made, based on histopathological examination, as imaging techniques were not able to distinguish PPL from pancreatic adenocarcinoma. In our case however, there was no definite mass or lesion in the pancreas and no recognizable lymphadenopathy. But there was the presence of the dilatation of the common and the hepatic ducts. The diagnosis of cholangiocarcinoma was suggested on CT scan imaging, thus favouring surgical treatment. Histopathology revealed the features of NHL, which on immunohistochemistry, was confirmed to be of the diffuse large B-cell type. Though the treatment of choice in lymphoma cases is chemotherapy or radiotherapy, the reason for surgery in our case was the lack of definitive diagnosis before the operation. It has been proved that surgery itself does not play a role in improving the patient's survival rate, but in cases of biliary or gastrointestinal obstruction, surgery should be performed to relieve the symptoms [2].

To conclude, primary pancreatic lymphoma is a rare entity presenting with non-specific symptoms, laboratory tests and imaging results. Confirmed diagnosis can be made either by histopathology or immunohistochemistry.

Chemotherapy or radiotherapy is the treatment of choice, while surgical intervention is considered to have no impact on patient survival.

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