

# Problems in Diagnosing Lymphoma of the Pancreas with Computed Tomography. A Case Report

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## Abstract

Primary lymphoma of the pancreas is a rare form of extranodal lymphoma accounting for less than 0.5% of pancreatic tumors. Percutaneous fine-needle aspiration of the pancreas with histopathological examination and immunohistochemical assay confirm the diagnosis. A 73 year old male presented with recurrent pancreatic type abdominal pain with significant weight loss over 1 year. He was pale with ill defined epigastric mass. Contrast enhanced CT showed an ill defined poorly marginated non enhancing hypodense mass lesion involving the body of the pancreas. CA 19-9 was normal. CT guided aspiration cytology was suggestive of hemolymphoid malignancy. Immunohistochemistry was positive for Leukocyte Common Antigen and CD 34. It was negative for CD3 and CD 20 indicating an undifferentiated lymphoma. Patient received two sessions of chemotherapy and was followed-up.

## Key words

Pancreatic lymphoma - contrast enhanced CT - fine needle aspiration cytology - immunohistochemistry

## Introduction

Primary lymphoma of pancreas (PLP) is a rare form of extranodal lymphoma accounting for less than 0.5% of pancreatic tumors. The prognosis of lymphoma is more favourable compared to adenocarcinoma. The radiological features of lymphoma are similar to pancreatic adenocarcinoma. Percutaneous fine-needle aspiration of the tumor with histopathological examination and immunohistochemical assay confirms the diagnosis. Here we report

a case of primary pancreatic lymphoma mimicking pancreatic adenocarcinoma.

## Case presentation

A 73 year-old male, a diabetic for two years, presented with recurrent attacks of acute abdominal pain localized to the epigastrium with radiation to the back. There was no history of fever, vomiting or jaundice. He had significant anorexia and a weight loss of 9 kg within one year of illness. The patient had hypertrophic obstructive cardiomyopathy.

On examination he was pale, peripheral nodes were not palpable. He had mild hepatomegaly and an ill-defined mass in the epigastrium. Clinically, pancreatic malignancy masquerading as recurrent acute pancreatitis was considered. Blood sugar was elevated. Serum amylase and lipase levels were normal. Ultrasonogram showed a diffuse enlargement of the pancreas with an ill-defined hypoechoic mass of 3.4 x 3.8 cm in the region of distal head, neck and proximal body. The main pancreatic duct was dilated. Tumour marker CA 19-9 was 5.6 IU / mL (N < 40 IU).

Contrast enhanced CT (CECT) (Fig.1) showed an ill defined poorly marginated non enhancing hypodense mass

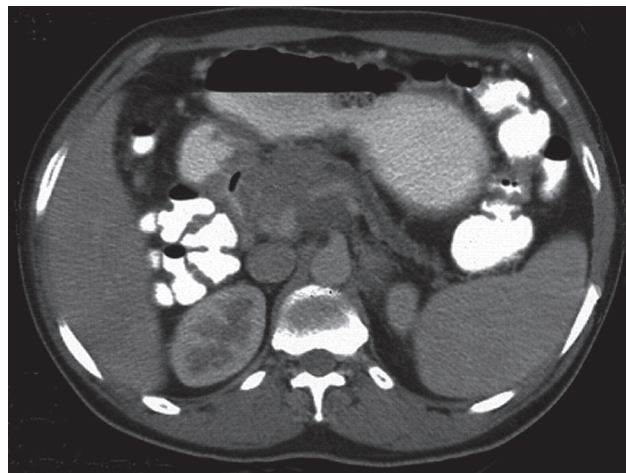


Fig.1 CT aspect. Hypodense mass in the body of the pancreas.

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lesion involving the body of the pancreas, encasing the splenic artery, superior mesenteric vein, splenic vein and splenoportal confluence with loss of fat plane. There was left sided hydronephrosis. CT guided aspiration cytology showed a malignant small round cell tumor with necrosis and fibroblastic reaction suggestive of hemolymphoid malignancy. Immunohistochemistry was positive for Leukocyte Common Antigen and CD 34. It was negative for CD3 and CD 20 indicating an undifferentiated lymphoma. Bone marrow aspiration showed non-Hodgkin's lymphoma stage IV.

The patient received two sessions of chemo-therapy: VP-16-Etoposide, Cytosine Arabinoside and Mitoxantrone. Patient is on follow up.

## Discussion

Primary lymphoma of pancreas (Stage IV) was the most likely diagnosis in the present case. The typical computed tomography of the pancreas was that of loss of fat plane, encasement of major vessels and dilatation of the main pancreatic duct. The immunohistochemistry confirmed the lymphoid lineage. There was no other lymphadenopathy.

Primary lymphoma of pancreas is a rare form of extranodal lymphoma (less than 0.5% of pancreatic tumors) originating from the pancreatic parenchyma (1). Histopathological examination is often mandatory in order to obtain a definitive diagnosis since symptoms and radiological features are quite similar to those of other pancreatic masses. In comparison to adenocarcinoma of pancreas, the prognosis of lymphoma is more favourable. Percutaneous fine-needle aspiration of the pancreas with advanced immunohistochemical assay confirms the diagnosis (1).

Several case reports are available in the world literature where difficulty has been encountered in distinguishing a primary lymphoma of the pancreas from non-Hodgkin's lymphoma involving the regional lymph nodes of pancreas on CT.

Levy et al (2) describe a patient who had an initial diagnosis of adenocarcinoma of the pancreatic head and subsequently was confirmed to have non-Hodgkin's large-cell lymphoma. The patient responded to chemotherapy. Schiefke et al (3) reported a 74 year-old woman with clinical features of pancreatitis. Ultrasound and endoscopic ultrasound revealed a lesion of low echogenicity in the pancreatic head. There were no hepatic metastases, enlarged lymph nodes or vascular infiltration. Exploratory laparotomy with biopsy of the pancreas revealed a diffuse large cell non-Hodgkin's lymphoma.

Kawakami et al (4) reported a 71-year-old Japanese male whose CT scan, angiogram and ERCP indicated pancreatic head adenocarcinoma. CA 19-9 levels were elevated. The patient underwent choledoch-jejunostomy and gastro-jejunostomy. The mesenteric lymph node was positive for lymphoplasmacytic lymphoma. The patient died. Retrospective analysis of the CT scan was suggestive of primary lymphoma of the pancreas rather than adeno-

carcinoma based on the presence of a well defined homogeneous low density mass lesion.

Marked lymphadenopathy around the pancreas due to lymphoma (abdominal lymphoma) occasionally can mimic pancreatic carcinoma on ultrasonography as was seen in the present case. Color Doppler study can differentiate the two pathological entities. In a comparative study of 12 cases of abdominal peripancreatic lymphoma and 21 cases of total pancreatic carcinoma (5), turbulent flow in the involved vessels was considered as diagnostic of pancreatic carcinoma rather than abdominal lymphoma (5). The shape, echogenicity of the lesion, and mode of vascular involvement are not specific and cannot differentiate the two pathologies. Differences in the maximal velocities and resistive indices of the involved vessels in Doppler are nonspecific. Ultrasound and CT with guided biopsy often give a clue to the diagnosis.

Several pitfalls have been reported with use of US and CT study. On dual-phase helical CT (6) lymphomas appear as hypo-attenuating focal lesions or as a diffuse infiltrate of the gland. Absence of biliary tree dilatation, despite the presence of a bulky tumor, or associated extensive retroperitoneal adenopathy offers clue to the diagnosis (7). An autopsy in a patient reported by Matsubayashi et al (7) revealed diffuse, mixed cell-type, non-Hodgkin's lymphoma of T-cell subtype (7).

Abdominal ultrasonography and CECT scanning in a 62-year-old Japanese male revealed several enlarged lymph nodes around the pancreas tail and the hilus of the spleen. Confirmation of stage-I mantle-cell lymphoma (8) was possible only at exploratory laparotomy. In the case reported by Boni et al (1) abdominal CT scan showed a diffuse enlargement of the head and body of the pancreas associated with lymphadenopathy along the lesser gastric curvature. Multiple biopsies of the pancreatic head and regional lymph nodes revealed a primary low-grade non-Hodgkin B cell pancreatic lymphoma.

Schiefke et al (3) reported a low echogenicity mass in the pancreatic head in the absence of hepatic lesions, enlarged lymph nodes or vascular infiltration. These were missed at CT. Exploration laparotomy confirmed the diagnosis. The biopsy specimen revealed a diffuse large cell non-Hodgkin-lymphoma. The case highlighted that imaging techniques like CT could also miss lesions picked up on US.

Other investigations which may help to distinguish between primary lymphoma of pancreas and adenocarcinoma are high levels of serum soluble interleukin-2 receptor (17- 751 U/ml) (7).

The differentiation of primary lymphoma of pancreas, adenocarcinoma and nodal masses in the region of pancreas is important because the management strategy in all these situations differ. In most series, the response to chemotherapy in individuals with non-Hodgkin's lymphoma has been favorable. CHOP therapy is recommended (2,3,9). Yoneda et al (8) combined chemo with radiotherapy.

It is clear that imaging techniques alone cannot distinguish adenocarcinoma and tumors of less common

cellular origin, such as primary pancreatic lymphoma, especially in those with atypical imaging findings. Clinicians should consider this in their differential diagnosis. Though rare, the treatment is non surgical and has a better prognosis than adenocarcinoma. Histological examination of focal masses in the pancreas is therefore mandatory today to confirm the diagnosis. One can thereby avoid major surgery.

## Conclusions

Treatable primary lymphoma of pancreas is an important differential diagnosis of CT reported mass lesion pancreas.

CT guided biopsy is a necessity for mass lesions in the pancreas.

Immunohistochemistry identifies the type of lymphoma and predicts the outcome.

## References

1. Boni L, Benevento A, Dionigi G, Cabrini L, Dionigi R. Primary pancreatic lymphoma. *Surg Endosc* 2002;16:1107-1108.
2. Levy MJ, Wiersema MJ. Endoscopic removal of a biliary Wallstent with a suture-cutting device in a patient with primary pancreatic lymphoma. *Endoscopy* 2002;34: 835-837
3. Schiefke I, Troltzsch M, Keim V. [Pitfalls in diagnosis of non-Hodgkin-lymphoma of the pancreas] *Ultraschall Med* 2002; 23:407-410.
4. Kawakami K, Nomura H, Watanabe Y, Momma F. [Primary pancreatic lymphoma with elevated serum CA19-9 level]. *Rinsho Ketsueki* 2002; 43:292-297.
5. Ishida H, Konno K, Ishida J et al. Abdominal lymphoma: differentiation from pancreatic carcinoma with Doppler US. *Abdom Imaging* 2002;27:461-464.
6. Sheth S, Fishman EK. Imaging of uncommon tumors of the pancreas. *Radiol Clin North Am* 2002;40:1273-1287.
7. Matsubayashi H, Takagaki S, Otsubo T et al. Pancreatic T-cell lymphoma with high level of soluble interleukin-2 receptor. *J Gastroenterol* 2002;37:863-867.
8. Yoneda S, Yoshiji H, Kuriyama S et al. Stage I mantle-cell lymphoma that was difficult to differentiate from abdominal tuberculous lymphadenitis and metastatic pancreatic cancer. *J Gastroenterol* 2002;37:859-862.
9. Cohen Y, Libster D, Amir G et al. Primary ALK positive anaplastic large cell lymphoma of the pancreas. *Leuk Lymphoma* 2003;44:205-207.

## Quiz HQ - 35, pag.76. Answer

The diagnosis is: Waldenström's macroglobulinemia

The presentation supports the critical role of intestinal biopsy for patients with chronic diarrhea and malabsorption. Differential diagnosis involves celiac sprue, Crohn's disease, microscopic colitis, Whipple disease, amyloidosis and malignancy, especially lymphoproliferative disorders. Endoscopic examination was unusual, requiring biopsies from the intestinal mucosa. Histopathological examination revealed acellular deposition of a PAS-positive material in the villi, simulating the microscopic appearance of amyloidosis or Whipple disease. The sample was negative

for Congo red stain and the bone marrow biopsy revealed the typical malignant proliferation of IgM-producing lymphocytes.

Waldenström's macroglobulinemia is a neoplasia characterized by malignant proliferation of the IgM-producing lymphocytes. The disease presents with hepatosplenomegaly in one third of cases. Gastrointestinal involvement may occur in an infiltrative pattern characterized by diffuse infiltration of the bowel wall with neoplastic cells similar to the pattern seen in other immunoproliferative disorders or, more commonly, acellular macroglobulin is deposited predominantly at the tips of the villi and interstitium, leading to malabsorption.