

CASE REPORT

Primary Pancreatic Lymphoma Presenting as Gastric Fistula: A Case Report

Chinmay Bera¹, Usha Goenka², Indrajeet Kumar Tiwari¹, Vijay Rai¹, Enam Murshed Khan³, Mahesh Kumar Goenka¹

¹Institute of Gastrosiences, Departments of ²Clinical Imaging and Interventional Radiology and ³Pathology, Apollo Gleneagles hospital, Kolkata, West Bengal, India

ABSTRACT

Non Hodgkin lymphoma primarily originating from the pancreas is a rare condition. Usual presentations include abdominal pain, anorexia and weight loss. Rarely, it may present as acute pancreatitis. Differentiating from the adenocarcinoma is difficult due to similar presentation and imaging features. Histopathological diagnosis is essential considering the impact on the management and prognostication. Presentation as fistulous communication with stomach or colon has not been reported in the literature. Here, we report a 73-year-male with primary pancreatic lymphoma presenting with gastric fistula.

INTRODUCTION

Most primary pancreatic lymphomas (PPL) are non Hodgkin lymphoma (NHL). NHL arising from lymphoreticular organ and involving pancreas secondarily has been seen in 30% of case [1]. Pancreas as the primary site of NHL is very rare. PPL is a rare extra-lymphatic NHL of the B cell type that comprising less than 1 to 2% of all extra-lymphatic lymphomas, and 0.5% of pancreatic tumours [2, 3]. The common presentation of PPL is abdominal pain and associated symptoms like anorexia and weight loss. Acute pancreatitis like presentation is uncommon. PPL may mimic pancreatic adenocarcinoma because of their similar presentation and imaging findings. The key to differentiation among the two is the histology. PPL are much more amenable to treatment compared with pancreatic adenocarcinoma and correct diagnosis is crucial given that PPL has different management and prognosis [4, 5]. It is important to diagnose PPL because of its better prognosis and also a different management strategy in comparison with pancreatic adenocarcinoma. Here we describe a case of PPL with rare presentation as gastric fistula.

CASE REPORT

A Seventy-three-year-old diabetic and hypertensive male presented with history of anorexia, loss of weight

and upper abdominal pain of one month duration with a short duration fever. Past medical history was negative for alcohol consumption or medication use. On examination, he was febrile and there was significant tenderness over the epigastrium without any obvious palpable mass. There was no palpable cervical or axillary lymphadenopathy. On investigation, his haemoglobin was 10.7 gm/dL, total counts: 26410 cells/cu.mm, platelet count: 1, 50,000/cu.mm, urea: 60 mg/dl, creatinine: 1.0 mg/dL (normal range 0.6-1.3 mg/dl), total/direct bilirubin: 1.2/0.6 mg/dl, SGOT/SGPT: 160/167 U/L, Alkaline phosphatase: 246 U/L. His serum amylase level of 101 U/L (normal range 30-110 U/L) and a serum lipase level of 98 U/L (normal range 23-300 U/L). CA 19-9 level was 8.08 U/L. At the time of admission he was in sepsis. He was started on intravenous antibiotics after obtaining blood culture.

Computed tomography (CT) of abdomen was suggestive of necrosis of the tail of pancreas with perigastric collection (**Figure 1**). A drain was placed under CT guidance at the perigastric area (**Figure 2**). There was a large fistula in the greater curvature of the stomach along with perigastric and peripancreatic collection (**Figure 3**). At upper GI endoscopy we found a fistulous opening in proximal body along the greater curvature of the stomach (**Figure 4**). As he was not fit for surgery, CT guided aspiration of the necrotic material was done and biopsy was taken from the pancreatic tissue. A drain was placed in the perigastric area. He was put on nasojejunal tube feeding and IV antibiotics according to blood culture and sensitivity report. Gradually his general condition improved.

Fine needle aspiration cytology from the pancreatic tail region mass has showed blood elements and necrotic

Received May 27th, 2016 - Accepted September 10th, 2016

Keywords Gastric Fistula; Pancreatic Lymphoma, Familial

Correspondence Chinmay Bera

Institute of Gastrosiences

Apollo Gleneagles Hospital

Kolkata 700054

India

Tel 0049-202-896-2243

E-mail chinmay.bera@gmail.com



Figure 1. Enhanced CT abdomen shows necrosis in the region of the tail of pancreas with perigastric collection.



Figure 2. Contrast enhanced CT abdomen shows necrosis in the region of the tail of pancreas with drain in situ.



Figure 3. CT abdomen shows fistulous communication (arrow) between greater curvature of the stomach and perigastric collection.

debris in the smear. Histopathological examination (**Figure 5**) of the core biopsy specimen was showing fibrocollagenous tissue infiltrated by small to medium sized lymphoid cells having high nuclear to cytoplasmic ratio, dense chromatin and scant cytoplasm. The immunohistochemical studies (**Figure 6**) showed diffuse positivity to CD20, CD10 and bcl2 whereas cytokeratin and CD3 were negative in tumor cell. Ki67 was present in 60 % of tumor cells.

After few days, feculent materials were seen coming out of the drain placed at the peripancreatic collection. A repeat CT abdomen showed fistulous communication between colon and greater curvature of the stomach. Chemotherapy was considered after surgical closure of the fistula but patient was not willing to continue further management.

DISCUSSION

Pancreatic tumours have been classified as epithelial and non epithelial tumour. Pancreatic lymphoma is categorized as a non epithelial tumor of the pancreas. Primary pancreatic lymphoma (PPL) is a rare extranodal manifestation of any histopathological subtype of B cell non Hodgkin lymphoma. Secondary involvement of the pancreas by the B cell non Hodgkin lymphoma is also a rare occurrence. Pancreatic lymphoma represents less than 1% to 2% of all pancreatic malignancies, and less than 1% of all extranodal non-Hodgkin's lymphomas [2]. As described by Behrns and colleagues in 1994, certain criteria must be met to suggest that the lymphoma is of pancreatic origin and not a nodal lymphoma extending to the pancreas [1]. According to these criteria, a lymphoma localized to the pancreas is defined as a pancreatic mass with involvement of peripancreatic lymph nodes without distal lymph node involvement, no hepatic or splenic metastases, and a normal white blood cell count. Primary pancreatic lymphoma was identified in 0.2 % of all patients with lymphoma and 0.4 % of all patients with any primary pancreatic tumor. Most



Figure 4. Gastroscopy shows fistulous opening along the greater curvature of stomach.

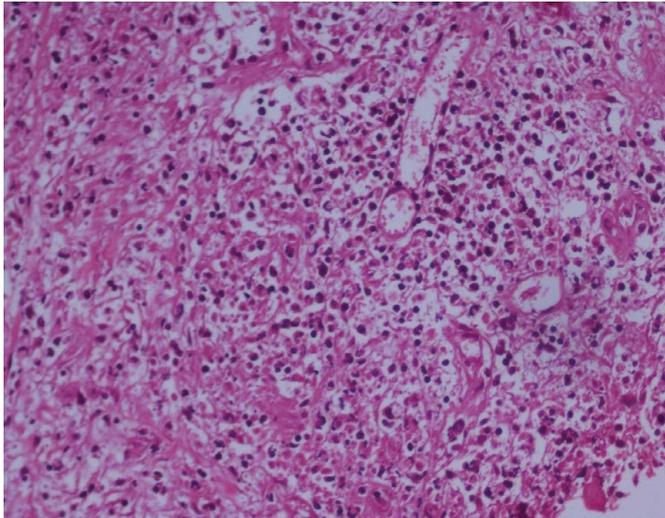


Figure 5. Histopathological study from the pancreatic cell mass shows fibrocollagenous tissue infiltrated by small to medium sized lymphoid cells having high nuclear to cytoplasmic ratio, dense chromatin and scant cytoplasm.

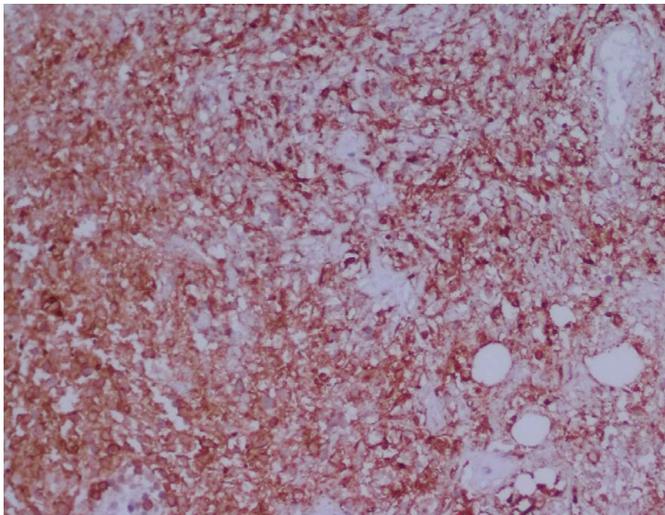


Figure 6. CD-20 immunostaining shows diffuse membranous positivity.

commonly found in elderly male. Common presentation include abdominal pain along with B symptoms [5]. Also mild alteration of the main pancreatic duct and elevation of serum amylase levels may be observed but the clinical manifestation of pancreatitis is infrequent. Involvement of stomach and other organ due to their proximity causing fistulous communication has not been mentioned in the literature.

Cross-sectional imaging may be helpful in differentiating between adenocarcinoma and primary lymphoma; however, it is not specific. Histologic analysis is crucial for differentiating between these two [2]. Accurate diagnosis by histopathological examination is critical for the timely diagnosis and non-surgical management of lymphoma [5]. Percutaneous or endoscopic core biopsy should be performed to establish the diagnosis. Ultrasound and Computed Tomography guided biopsy techniques can easily provide sufficient diagnostic tissue without major complications. In most patients, the diagnosis can be established [6]. Diagnostic criteria as defined by Dawson *et al.* include: 1. neither superficial lymphadenopathy

nor enlargement of mediastinal lymph nodes on chest radiography; 2. a normal leukocyte count in peripheral blood; 3. main mass in the pancreas with lymphnodal involvement confined to peri pancreatic region; and 4. no hepatic or splenic involvement [7]. Presence of leukocytosis in our patient can be explained by sepsis, otherwise he was fulfilling all other criteria's mentioned by Dawson *et al.*

Investigation of histological marker such as CD20, CD3 and Ki67 can be useful [8]. CD20 was strongly positive in our patients and also Ki67. Serum carbohydrate antigen 19-9 (CA 19-9) level in patients with PPL is usually not elevated. This is in contrast with pancreatic adenocarcinoma, in which almost 80% of cases have a high CA19-9 level [9].

EUS has also been used to evaluate pancreatic masses and surrounding structures. EUS can provide histological diagnosis of PPL [10]. Flamenbaum *et al.* described the typical endoscopic sonography findings as a strongly hypoechogenic appearance in the pancreas, hypertrophy in all its segments, a hyperechoic wall in the common pancreatic duct contrasting with the adjacent parenchyma, and multiple isoechogenic peripancreatic lymph nodes [11]. These authors concluded that the endoscopic sonography findings were highly specific and allowed distinction of lymphoma from all other pancreatic tumors.

Treatment options include surgical resection, chemotherapy alone, or combined radiation and chemotherapy. Response rates to chemotherapy alone are very good, with approximately 72% of patients have showed no evidence of disease at 34 months [12].

Conflict of Interest

The authors declare that there is no conflict of interests regarding the publication of this paper.

References

1. Behrns KE, Sarr MG, Strickler JG. Pancreatic lymphoma: is it a surgical disease? *Pancreas* 1994; 9:662-67. [PMID: 7809023]
2. Freeman C, Berg JW, Cutler SJ. Occurrence and prognosis of extranodal lymphomas. *Cancer* 1972; 29:252-60. [PMID: 5007387]
3. Baylor SM, Berg JW. Cross-classification and survival characteristics of 5,000 cases of cancer of the pancreas. *J Surg Oncol* 1973; 5:335-58. [PMID: 4355621]
4. Nayer H, Weir EG, Sheth S, Ali SZ. Primary pancreatic lymphomas: a cytopathologic analysis of a rare malignancy. *Cancer* 2004; 102:315-21. [PMID: 15386314]
5. Grimison PS, Chin MT, Harrison ML, Goldstein D. Primary pancreatic lymphoma--pancreatic tumours that are potentially curable without resection, a retrospective review of four cases. *BMC Cancer* 2006; 6:117. [PMID: 16674812]
6. Mallery JS, Centeno BA, Hahn PF, Chang Y, Warshaw AL, Brugge WR. Pancreatic tissue sampling guided by EUS, CT/US, and surgery: a comparison of sensitivity and specificity. *Gastrointest Endosc* 2002; 56:218-24. [PMID: 12145600]

7. Dawson IM, Cornes JS, Morson BC. Primary malignant lymphoid tumours of the intestinal tract. Report of 37 cases with a study of factors influencing prognosis. *Br J Surg* 1961; 49: 80-89. [PMID: 13884035]
 8. Naito Y, Okabe Y, Kawahara A, Taira T, Yamagushi T, Abe H, Arikawa S, et al. Guide to diagnosing primary pancreatic lymphoma, B-cell type: immunocytochemistry improves the diagnostic accuracy of endoscopic ultrasonography-guided fine needle aspiration cytology. *Diagn Cytopathol* 2012; 40:732-36. [PMID: 21416645]
 9. Lin H, Li SD, Hu XG, Li ZS. Primary pancreatic lymphoma: report of six cases. *World J Gastroenterol* 2006; 12:5064-67. [PMID: 16937508]
 10. Arcidiacono PG, Carrara S. Endoscopic ultrasonography: impact in diagnosis, staging and management of pancreatic tumors. An overview. *J Pancreas* 2004; 5:247-52. [PMID: 15254358]
 11. Flamenbaum M, Pujol B, Souquet JC, Cassan P. Endoscopic ultrasonography of a pancreatic lymphoma. *Endoscopy* 1998; 30:S43. [PMID: 9615896]
 12. Miller TP, Dahlberg S, Cassady JR, Adelstein DJ, Spier CM, Grogan TM, LeBlanc M, et al. Chemotherapy alone compared with chemotherapy plus radiotherapy for localized intermediate and high-grade non-Hodgkin's lymphoma. *N Engl J Med* 1998; 339:21-6. [PMID: 9647875]
-