ISSN 2689-8268 | Volume 8

American Journal of Surgery and Clinical Case Reports

Case Report Open Access

Rare Case of Primary Pancreatic Lymphoma Following Complex Environmental Exposure

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Received: 28 May 2025

Accepted: 09 Jun 2025

Published: 12 Jun 2025

J Short Name: AJSCCR

Citation: W Sebastian. Rare Case of Primary Pancreatic Lymphoma Following Complex Environmental Exposure. Ame J Surg Clin Case Rep. 2025; 8(16): 1-3

1. Abstract

Primary pancreatic lymphoma (PPL) is an exceptionally rare malignancy, accounting for only 0.1% of lymphomas and 0.2% of pancreatic tumors. Risk factors include male gender, smoking, alcohol use, family history, and environmental exposures such as heavy metals or industrial toxins. We present the case of a 52-year-old male with diabetes, hypertension, and prior prostate cancer, who developed back pain, worsening hyperglycemia, and elevated liver enzymes. He had significant environmental exposure as a first responder during the September 11th attacks. Imaging revealed a pancreatic head mass, initially concerning for adenocarcinoma. Endoscopic ultrasound with fine-needle aspiration identified low-grade follicular lymphoma. He was treated with Rituximab, later transitioning to Revlimid and Obinutuzumab after disease recurrence. This case highlights the diagnostic challenges in differentiating PPL from other pancreatic neoplasms and emphasizes the importance of thorough evaluation, appropriate tissue sampling, and long-term surveillance, particularly in patients with complex medical histories and known environmental exposures.

2. Introduction

Primary pancreatic lymphoma (PPL) is an exceedingly rare disease that makes up only 0.5% of all pancreatic masses [1]. Diagnosis requires an accurate fine needle aspiration (FNA) or core needle biopsy. Flow cytometry can be used to confirm the diagnosis, as in our case, however there are instances where multiple biopsies may be necessary. Despite often having similar presentations to other pancreatic malignancies, distinguishing PPL is critical as treatment and prognosis are vastly different. While pancreatic adenocarcinoma often results in decreased life expectancy and is treated with a combination of surgical resection, chemotherapy, and/or radiation, surgical resection is rarely indicated in PPL and is generally treated with systemic therapy [2-3].

3. Case Study

We present the case of a 52 year old male with a past medical history of diabetes, hypertension, and prostate cancer status post cyber knife treatment, and exposure to hazardous substances while working as a first responder in the September 11th, 2001 World Trade Center attacks who presented to his primary care physician's office with complaints of occasional back pain, worsening hyperglycaemia and elevated transaminases on routine blood work. Social history is significant for daily alcohol use and 20 pack year smoking history. Our patient was found to have a 10-pound weight loss over two weeks on review of systems but no findings of urine or stool color changes, early satiety, nausea, vomiting, or constitutional symptoms. Blood work was notable for a total bilirubin of 1.1, alkaline phosphatase 330, AST 80, ALT 313, CA 19-9 72, and negative hepatitis panel.Abdominal ultrasound revealed a head of pancreas mass as well as gallbladder sludge. MRI confirmed a 3.5cm x 4.4cm mass at the pancreatic head with abrupt narrowing at the common bile duct (CBD). The main pancreatic duct was of normal caliber. Due to the constellation of symptoms and objective data, there was concern for underlying malignancy and our patient underwent endoscopic ultrasound and FNA. This revealed a head of pancreas lesion causing biliary strictures, peripancreatic lymphadenopathy, loss of fat plane at the superior mesenteric vein. The distal pancreas appeared unremarkable and was not concerning for chronic pancreatitis. He subsequently developed post-procedure pancreatitis and biliary obstruction (total bilirubin 2.2) and underwent stenting of his CBD, at which point the endoscopists also obtained CBD brushings.Initial FNA results returned as negative for malignant cells but demonstrated acini and lymphocytes. Our patient had elevated IgG4 levels (166, upper limit of normal 96) and the sample was sent for IgG4 staining to rule out focal autoimmune pancreatitis. Staining returned as negative for IgG4. An outside review of the FNA returned consistent with chronic pancreatitis however re-review was concerning for low-grade follicular lymphoma. Fluorescence in-situ hybridization (FISH) analysis detected BCL2 rearrangement. A second facility reviewed the FNA slides and concluded that the sample was benign pancreatic parenchyma mixed with abundant small lymphocytes and recommended further testing. CBD brushing returned without malignancy (Table 1). Positron emission tomography (PET) scan was done which demonstrated intense avidity at the pancreatic head with enlarged periportal, gastrohepatic, paraaortic, and aortocaval nodes (Figure 1).

Following discussion with our patient for further testing in the form of repeat FNA versus surgical biopsy to confirm diagnosis and initiate treatment, he elected to undergo repeat FNA. More focused stains and testing confirmed atypical lymphocytes in specimen, compatible with low grade lymphoma. Positive flow cytometry with kappa light chain restricted CD10 positive mature B-cells. Given confirmation

Table 1: Summarizing FNA, immunologic, and staining results.

	JSUMC	Mt. Sinai	MSKCC
ThinPrep with CellBlock (10/13/2020)	Acini, numerous lymphocytes seen. **Negative for malignant cells**.	Report 1: **Chronic pancreatitis.** While there are no prominent plasma cells, storiform fibrosis or obliterative phlebitis. **No tumor is seen.**	Atypical. Benign pancreatic parenchyma admixed with abundant small lymphocytes.
Pancreas EUS/ FNA cell block		**Addendum 2:<u>follicular lymphoma, low grade.</u>**	
Microscopic description and immunostains		**atypical interstitial lymphoid infiltrate** with no definite formation of follicles. The lymphocytes are small in size with condensed chromatin, inconspicuous nucleoli and wild nuclear irregularity. Immunostains highlight numerous B-cells (CD20+, PAX6+) with admixed T-cells (CD3+). The B-cells are CD10+, BOLB+, BCL2+, and BCL1-. MIB1 proliferation rate is low ~10%, CD138 and MUM1 positive cells are rare. Kappa and lamba stains are non-contributory.	
FISH	• BCL6 rearrangement-not detected • MYC rearrangement-not detected • MYC amplification: not detected • BCL2 rearrangement: **DETECTED**		
IgG4		Immunohistochemical stains performed on submitted tissue block show a few <u>CD138-postive cells</u> which are however **<u>negative</u> with an immunostain for IgG4.**	

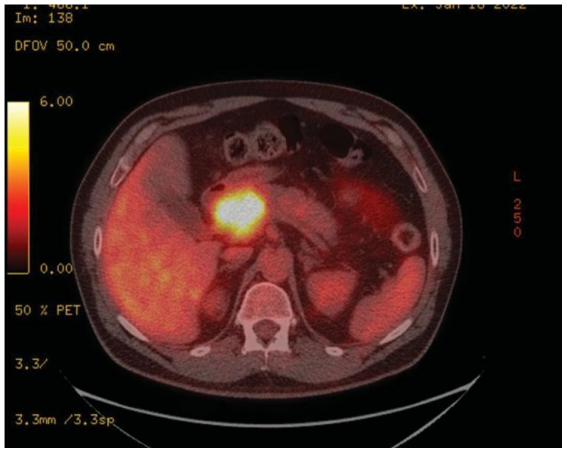

Figure 1: PET/CT Scan showing intense avidity at the pancreatic head, with periportal lymphadenopathy. SUV of pancreas head mass 10.1 compatible with malignancy.

of low grade, follicular lymphoma, the patient was referred to medical oncology for initiation of systemic therapy. The patient underwent 4 cycles of Rituxin monotherapy. The patient tolerated systemic chemotherapy well although difficult glycemic control. The patient continued an active surveillance strategy. Initial PET/CT three months after initiating systemic therapy without abnormal FDG uptake, resolution of head of pancreas mass and surrounding adenopathy. Repeat PET/CT 7 months later demonstrated recurrence of head of pancreas mass with max SUV 10. Repeat biopsy confirmed recurrence with low grade follicular lymphoma, demonstrated by CD20 positive B-cells forming nodules that expressed BCL2 and focal weak BCL6. Follicular lymphoma had a low proliferate rate by Ki67%, approximately 5%. Flow cytometry results positive for CD positive monoclonal B-cell population. Systemic therapy was initiated with revlimid/obinutuzumab. PET/CT with resolution of head of pancreas mass and surrounding lymphadenopathy. Surveillance interval increased to every 3 months while on therapy with no evidence of disease. The patient has now completed revlimid/obinutuzumab. He continues on PET/CT surveillance every three to six months with no further recurrence at 3.5 years after diagnosis.

4. Discussion

Our case presented here reflects the diagnostic difficulty with primary pancreatic lymphoma (PPL). PPL is rare and represents 0.1% of all malignant lymphomas and 0.2% of all pancreas tumors [4]. Establishing the diagnosis of PPL is vital for the management of patients as it can help to avoid unnecessary surgical procedures or mismanagement. Diagnosis of PPL can pose diagnostic difficulty as the symptoms can be generalized and often mimic other disease processes such as pancreatic adenocarcinomas [5]. As in our

case, the presentation of this patient was initially concerned for pancreatic adenocarcinoma and the patient required multiple endoscopic procedures and biopsies to confirm PPL diagnosis. Patients with PPL often undergo multiple tissue sampling attempts to get optimal tissue for diagnosis. Tissue acquisition may be obtained through endoscopic, percutaneous or surgical modalities. Accurate diagnosis is paramount for optimizing patient outcomes. A comprehensive retrospective study reviewing 107 articles on PPL from 2000 to 2020 revealed that PPL was diagnosed following extensive surgeries in 21.9% of cases, while 4.0% were identified post-mortem [6]. There has been considerable data on the utilization of FNA in aiding the diagnosis of pancreatic carcinomas, however, for PPL the literature is limited. A study that looked at 14 cases of PPL (6 DLBCL and 4 follicular) that were diagnosed with FNA found that DLBCL subtype of PPL required an average of 3.9 passes to obtain sufficient diagnostic material vs 4.3 in those with follicular subtype7. Although it is important to consider the addition of immunophenotyping as lymphoma is difficult to diagnose on fine needle aspiration alone [7]. A retrospective case-control of 11 patients found that FNA alone only had a 28% accuracy in diagnosing PPL without the use of flow cytometry, while those with adenocarcinoma had a 91% accuracy [8]. Immunophenotyping allows for identification of PPL, but can further characterize PPL into the subtypes, which further helps to customize treatment. Most of the PPL pathology is consistent with diffuse large B-cell lymphoma (DLBCL) (53.6%) followed by follicular lymphoma (9.8%) [6].

Although no standardized guidelines exist, evidence indicates that (PPL) generally responds well to systemic therapy, reducing the need for extensive and potentially debilitating surgical procedures. Therapies should be tailored based on identified histological subtype. Endoscopic procedures can be utilized in conjunction with systemic therapy to address complications of pancreas head mass such as biliary obstruction.Environmental factors should be considered in the development of cancer following exposure to the ash, debris, and toxins associated with the 9/11 attacks. In this case, the patient, a first responder during 9/11, experienced prolonged exposure to harmful toxins in the aftermath of the event. Studies have indicated that such exposure is associated with an increased risk of both leukemia and lymphoma [9-10].Finally, this case highlights the need for continued surveillance in patients with PPL. From the limited data on PPL, recurrence rates have been estimated to be around 20% and often involving distant sites such as CNS [11].

4. Conclusion

This case highlights the importance of increased awareness regarding atypical presentations of extranodal lymphomas, such as primary pancreatic lymphoma (PPL). This is the first known reported case of PPL in a patient with a previous direct link to the hazardous exposure at the devastation sites of the September 11th attacks. It also underscores the need for comprehensive diagnostic strategies, ongoing surveillance, and tailored treatment approaches in managing these complex hematologic malignancies. Future research should prioritize the long-term outcomes of similar patients, particularly those with prior exposure to hazardous substances, to further elucidate the relationship between environmental factors and the development of extranodal lymphomas like PPL.

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