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Primary Pancreatic Lymphoma, Report of a Rare Case

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Abstract Primary pancreatic lymphoma (PPL) is a very rare malignancy. The diagnosis of PPL is challenging because of similar manifestations to pancreatic adenocarcinoma or other diseases of the pancreas. It is mainly seen in men and the 5th or 6th decades of life. Here we report a 60-year-old lady with pancreatic mass, presented with abdominal pain, weight loss, and GI disturbance. An abdominal CT scan showed a homogenous mass, measured 60mm, in the lesser sac with the probable origin of the pancreatic body and a mass-like lesion on the head of the pancreas. Endoscopic ultrasound-assisted fine needle aspiration (EUS-FNA) was done and cell block was prepared for histopathology and immunohistochemistry evaluation. The final diagnosis of Large B cell lymphoma was made. The patient received an R-CHOP chemotherapy regimen with a good therapeutic response. Accurate diagnosis is important, due to response to chemotherapy and avoidance of unnecessary radical surgery.

Keywords: *primary pancreatic lymphoma, case report*

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1. Introduction

Primary pancreatic lymphoma (PPL) is a very rare malignancy [1]. About 10-15% of all Non-Hodgkin Lymphomas occur in the gastrointestinal tract that often invades the small bowel and the stomach. Seldom appears in the pancreas [2]. The diagnosis and therapy of PPL are challenging because it is a rare disease with similar manifestation to pancreatic adenocarcinoma or other diseases of the pancreas [3,4]. PPL usually occurs in the 5th or 6th decade of life. The clinical presentation is non-specific and includes jaundice, nausea, weight loss, abdominal mass, abdominal pain, vomiting, diarrhea, and bowel obstruction that leads to pancreatic adenocarcinoma misdiagnosis [1,5,6]. Strong male predominance with a male/female ratio of 7/1 is reported in the cases [7]. However, others reported male involvement in about 65% of cases [8]. Diagnostic criteria of PPL are the mass-involved lymph nodes surrounded by the pancreas without any splenic and hepatic involvement and no palpable superficial lymphadenopathy, normal chest x-ray, and white blood cell count [9].

2. Case Report

A 60-year-old lady was presented, as an outpatient, with nausea, vomiting, weight loss, and abdominal pain for 5 months. In past medical history, ischemic heart disease and hyperlipidemia were positive. Drug and family history were unremarkable. For the patient lab tests and ultrasound, exams were requested. Complete blood count showed, WBC: 8.07 $x10^3$ /µl with neutrophil: 69.8%, RBC: 4.4$x10^6$ /µl, Hemoglobin: 10.4 gr/dl, MCV: 76.2 femtoliter and Platelet count: 474 $x10^3$ /µl. ESR was 88 mm/hour and CRP was 3+. Ultrasound exam of abdomen and pelvis showed a mild fatty change in the liver, microlithiasis in the left kidney (3mm), and duplicated pelvicalyceal system in the left side. Hypo echoic lobulated mass in the pancreas was also noted with a diameter of 60x50mm with a recommendation for a CT scan for better evaluation. An abdominal CT scan showed a homogenous mass, measured 60mm, in the lesser sac with the probable origin of the pancreatic body with no evidence of peripheral invasion and a mass-like lesion on the head of the pancreas. Venous contrast CT scan was recommended for better evaluation of later mass. Also sliding hiatal hernia was noted. The patient was admitted for an ultrasound-guided biopsy. Biopsy was done and the pathologist reported on cylindrical gray tissue (2x0.2cm), atypical glands in the fibrotic stroma (Figure 1), and noted that the findings are in favor of malignancy (Figure 2), but better characterization requires immunohistochemistry or re-biopsy. Due to non-diagnostic pathology reports and normal tumor markers, the patient was referred to a more equipped center for endoscopic ultrasound-assisted fine needle aspiration (EUS-FNA). Description of procedure was: "There was a large heterogeneous mass lesion in the region of pancreas head, body and tail with encasement of adjacent vessels without direct involvement. There were few regional lymph nodes. A small amount of ascites was noted. EUS-FNA of mass was done with needle Gauge 22 Boston. The common bile duct diameter was 8mm, with some sludge. The gallbladder contained sludge. The celiac

area was unremarkable. Final diagnosis: Pancreatic mass, highly suspicious for pancreatic cancer". Cellblock was provided and the pathologist reported: Atypical lymphoid cell population with high proliferation capacity and noted: "The possibility of high-grade B cell lymphoma should be seriously considered in this patient". Immunohistochemistry was done and showed positivity for CD20 and a high Ki-67 index. CD3, CD10, CD138, Bcl-6, MUM1, CD5, CyclinD1 and IgG4 were negative in atypical cells. The pathologist reported that these findings are in favor of Large B cell lymphoma. Chemotherapy was initiated for the patient with the diagnosis of pancreatic lymphoma. The patient received R-CHOP (Rituximab, Cyclophosphamide, Doxorubicin, Vincristine, and Prednisolone). After 5 courses of treatment, the patient was in good condition. CT scan of abdomen and pelvis after initiation of chemotherapy showed: "unremarkable visceral organs with atrophic pancreas and no retroperitoneal lymphadenopathy". Informed consent was obtained from the patient.

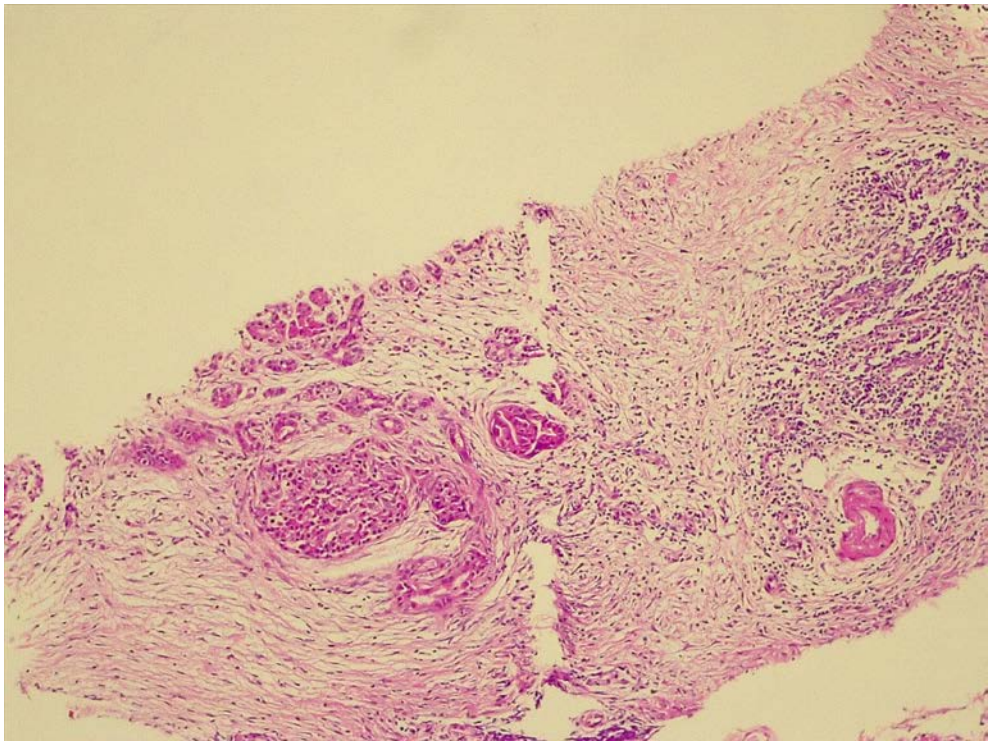

Figure 1. Core -needle biopsy, pancreas. Distort glands and lymphoid infiltrate in fibrotic stroma. Hematoxylin-Eosin staining. Magnification x100

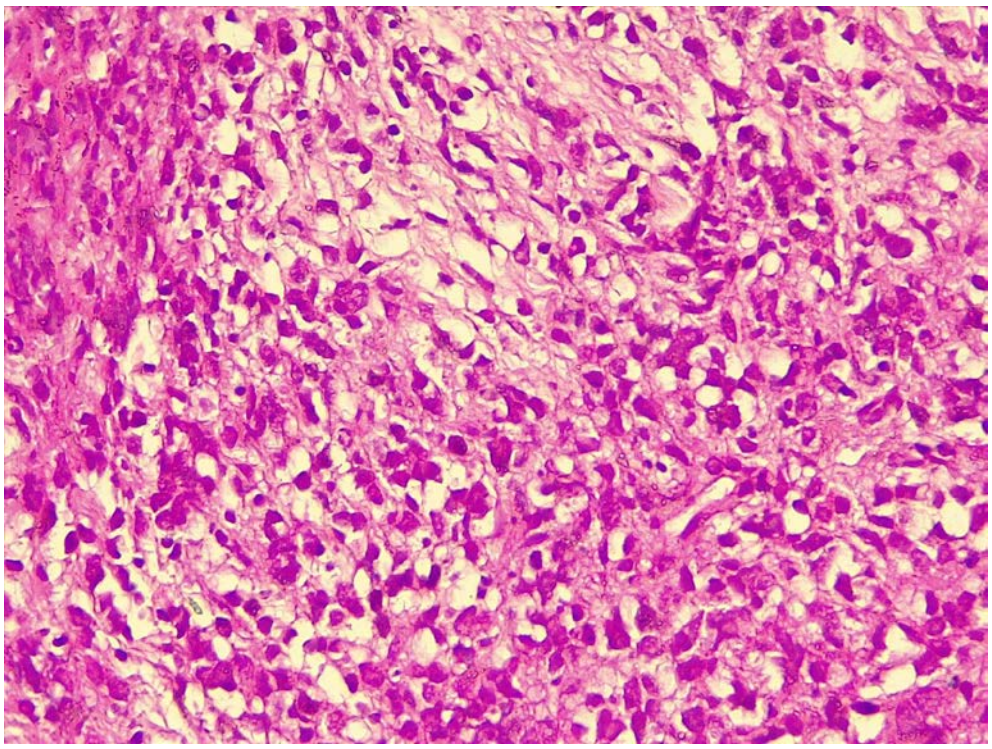

Figure 2. Atypical cells in favor of malignancy. Hematoxylin-Eosin staining. Magnification x400

3. Discussion

PPL accounts for about 5% of all pancreatic masses and 1% of all extranodal lymphomas. PPL usually occurs in males and the 5th or 6th decades of life. The diffuse large B cell lymphoma is the most common histopathological subtype in approximately 80% of cases. However, follicular lymphoma and other subtypes may also be seen [1,2]. The most common manifestation is weight loss and abdominal mass, while the other clinical manifestations of PPL, such as fever, dyspepsia, pain, nausea, night sweats, and chills, are less common [1,10]. In a recent review, the median age was reported, 53.1 years, and the most common symptoms as abdominal pain and jaundice [8]. The present patient had abdominal pain, weight loss, and GI disturbance, but no jaundice. Medium size of 60.6mm is reported, but cases as large as 200mm are on the report [8]. The maximum diameter of mass, in our patient, was 60mm with the bulk in the body, but also with a mass-like lesion in the head of the pancreas. Dawson et al. established criteria for diagnosing PPL as a normal leukocyte count with absent lymphadenopathy on imaging and involvement of spleen and liver. The head of the pancreas is the dominant part of involvement with the tumor [10,11]. The diagnosis and treatment of this disease are difficult. The site of involvement could mimic the presentation of pancreatic adenocarcinoma and other diseases [3]. The biochemical markers, such as CA 19-9, are often nonspecific for diagnosis. However, it usually increases in pancreatic adenocarcinoma while seldom elevated in patients with PPL [11]. Elevated levels of lactate dehydrogenase (LDH) are seen in 50.4% of cases in a review [8]. Criteria more in favor of PPL are younger age, greater tumor size, presence of B symptoms, absence of jaundice or diabetes mellitus [2]. Tumors greater than 60mm and the finding of distant lymph nodes in radiologic evaluation are clues in favor of lymphoma [8]. Distinguishing between PPL and other malignancies may be possible by EUS, or CT-guided FNA biopsy. When the FNA biopsy is not helpful for diagnosis, laparotomy or laparoscopy may be altered to conduct a biopsy of the lymph nodes or pancreatic mass [4]. PPL is presented in two forms: a focal form in the pancreatic head and a seldom diffuse/infiltrative form [12]. Due to different therapeutic modalities, preoperative diagnosis of PPL is of great importance to avoid unnecessary radical surgery [2].

4. Conclusion

Primary pancreatic lymphoma is a very rare type of pancreatic malignancy with non-specific symptoms and signs, should be considered in the clinical, radiology, and histopathology differential diagnosis of pancreatic lesions. Accurate diagnosis is important, due to response to chemotherapy and avoidance of unnecessary radical surgery.

Acknowledgments

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Conflict of Interest

The authors declare that there is no conflict of interest in this work.

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