

Primary Pancreatic Lymphoma with Active Gastric Bleeding. A Clinical Case and Review Literature.

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Abstract

Introduction: Primary pancreatic lymphoma is a rare clinical entity representing <0.5% of pancreatic cancers and 1% of extranodal lymphomas. Due to the lack of evidence of cases in the literature, their clinicopathological features, differential diagnosis, optimal therapy, and outcomes should be better defined.

We will present a case of a 41-year-old woman who presented symptoms such as nausea, epigastric pain, red blood vomiting, and melena.

At laparotomy, a large necrotic mass was found in the body and tail of the pancreas, infiltrating the stomach and spleen. A biopsy of the mass confirmed large B-cell lymphoma. The patient was diagnosed and started chemotherapy.

Conclusion: Primary pancreatic lymphoma is a rare clinical entity often misdiagnosed as pancreatic adenocarcinoma on presentation. Its clinical manifestations, radiological features, and biochemical signs are usually non-specific. It is based on a precise diagnosis made possible by histologic examination. Combined therapy remains the most optimal treatment approach for PPL but needs further evaluation.

Keywords: PPL, B-cell lymphoma, Melena, laparotomy.

Introduction

Primary pancreatic lymphoma is a rare clinical entity representing <0.5% of pancreatic cancers and 1% of extranodal lymphomas. [1, 2]

Due to the lack of cases in the literature, their clinicopathological features, differential diagnosis, optimal therapy, and outcomes must be better defined. [2](fig. 1)

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Primary pancreas lymphoma (PPL- primary pancreas lymphoma) is rare, while non-Hodgkin's lymphoma is the most common type.[3]

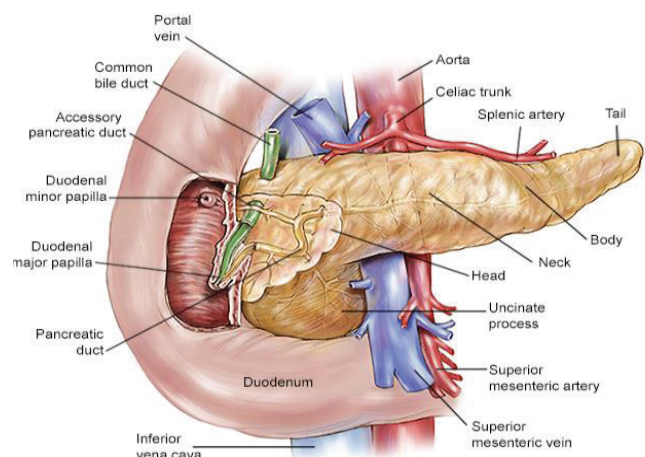


Figure 1. Anatomy of Pancreas

Generally, PPL is found in the pancreatic head, which has a large amount of lymphoid tissue, though it can be found anywhere along the gland [4]. Few case reports have discussed anatomical variation, but only one case report has been published describing PPL localized to the tail [5]. This case report discusses the finding of PPL solely in the pancreatic tail. [6].

Presentation of the Case

A 41-year-old woman complained of a sudden feeling of nausea, followed by several episodes of vomiting red blood. For 2-3 months, the patient referred to fatigue, physical weakness, and abdominal discomfort limited to the upper left quadrants.

She presented to the emergency department of the University Hospital Center "Mother Teresa," Tirana, Albania, a tertiary center, for further evaluation.

The patient was being treated at the time with benzodiazepine-class medications for anxiety and mood disorders. She denied using AIJS, alcohol, medicinal plants, illegal drugs, recent travel, trauma, and contact with sick people.

In the emergency, vital signs, hemodynamics, and respiratory conditions were unstable.

Physical examination showed pale skin and mucous membranes, with abdominal pain in the epigastrium and left hypochondrium. Rectal examination revealed normal stools.

Admission labs showed hemoglobin 6.0 mg/dL, mean cell volume (MCV) 80 FL, blood urea (BUN) 50 mg/dL, and creatinine 1.3 mg/dL. Liver function tests (LFTs), amylase, and lipase were regular.

In the gastroduodenal endoscopy (EGD) for anemia and hematemesis, a considerable deformity was observed in the significant curvature, with the appearance of a gastric ulcer.

This ulcer was closed after an attempt at sclerotherapy of the active hemorrhagic focus, but only after the patient underwent a further radiological diagnostic study as an Abdominal CT - Scanner. (fig. 2)

1.5-2 hours after EGD, the patient's general condition worsened significantly, with a profound drop in hemogram

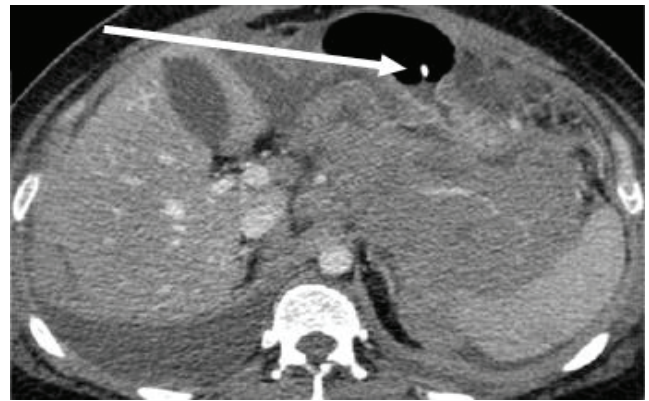


Figure 2. Abdominal CT--Scanner

parameters, hemoglobin 4 mg/dl, RBC 1.1 M/ml, MCV 72.

She underwent an exploratory laparotomy, where a 7 cm corpocaudal pancreatic mass was found, which perforated the greater curvature of the stomach, with compromise of the structure of the superior pancreatic artery, without compromise of the biliary system and the rest of the digestive tract.

The patient underwent corpocaudal pancreatectomy, splenectomy (fig. 3), and subtotal gastrectomy.

The mass sent for biopsy resulted in pancreatic large B-cell lymphoma.



Figure 3. Corpocaudal pancreatectomy and splenectomy

Microscopic Findings

Examination of the spleen and pancreas fragments shows a neoplastic lymphoid infiltration with large pleomorphic cells with thin nuclei and irregular contours. Some cells

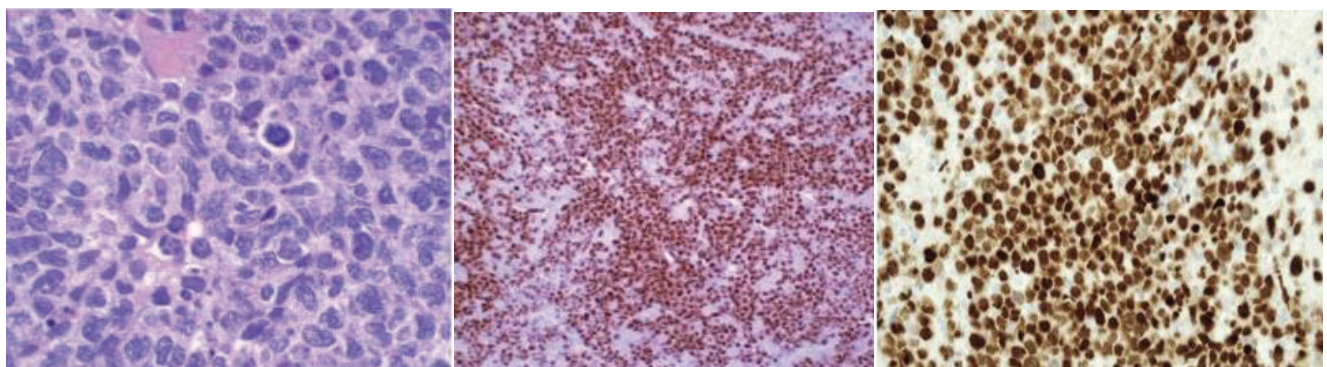


Figure 4. Pancreatic large B-cell lymphoma.

have distinct nucleoli, and the cytoplasm is relatively wide.

In the immunohistochemical study, neoplastic cells appear to have positive expression of CD20(+) while becoming CD3(-), CD10(-), MPO (-), CD30(+), CK (-), EMA (-), Ki-67 (+). The findings are consistent with B-cell lymphoma. Furthermore, the extent and exclusion of lymph nodes were assessed based on histology with hematoxylin and eosin. (fig. 4)

The patient underwent 13 chemotherapeutic sessions up to 2 years after the intervention, and according to cross-sectional images and scans, she has not had any relapse.

Discussion

Primary pancreatic lymphoma (PPL) is infrequent, accounting for only 1% of extranodal lymphomas and more than 0.5% of pancreatic tumors. [1]

From the literature, it was found that the most common variant of pancreatic lymphoma is B-cell lymphoma with giant cells, which accounts for 80% of all cases of PPL. [6].

Clinical signs such as epigastric pain, acute pancreatitis, small bowel obstruction, and diarrhea are the most common but non-specific symptoms. [7]

Common non-Hodgkin's lymphoma symptoms, such as rashes, chills, and night fever, are rare in PPL. [8]

Widely accepted criteria for diagnosing PPL were established by Dawson and include a mass in the pancreas, the absence of superficial or mediastinal lymphadenopathy on chest imaging, an average peripheral blood leukocyte count, and no involvement of the hepatic system or spleen. [9]

Most PPLs occur in the head of the pancreas, although they can also be found in other parts of the pancreas.[10] In our case, the patient presented with pain on the left side of the abdomen, and the mass was located in the body and tail of the pancreas.

Non-specific biochemical markers help to make an accurate diagnosis of PPL.

CA 19-9, a serum tumor marker, is usually elevated in patients with pancreatic adenocarcinoma but is unchanged mainly in patients with PPL.[11]

80% of patients with pancreatic adenocarcinoma have a high level of the marker CA 19-9, while in patients with PPL, the level of CA 19-9 in the serum is normal or shows only a slight increase when a biliary obstruction appears.[12]

High levels of LDH and β 2-microglobulin are considered high in lymphoproliferative cases but not necessarily high in PPL.[13]

However, a higher level of β 2-microglobulin and LDH are indicators of a poor prognosis for patients with PPL. [13]

Various imaging examinations are used in the evaluation of pancreatic lesions.

CT classifies PPL into two morphological groups: the pancreas has a structure similar to what appears in pancreatitis (with densification of the fatty tissue around the pancreas); a well-circumscribed tumor occurs in the pancreas.[14]

Merkle *et al.* has described the imaging findings in pancreatic lymphoma as a bulky, localized pancreatic head tumor without significant Wirsung duct dilatation, enlarged lymph nodes below the level of the renal veins, and invasive and infiltrating growth through to the retroperitoneal or upper abdominal organs and the gastrointestinal tract.[14] The presence of calcification or necrosis can exclude PPL.[15]

Regarding endoscopic retrograde cholangiopancreatography (ERCP) results, ductal dilatation often seen in pancreatic adenocarcinoma has not been reported in patients with PPL, which may help exclude the diagnosis of PPL. [16] Magnetic resonance imaging is entirely non-specific. [17]

Clinical data, biochemical markers, and radiological examinations help to diagnose PPL but are non-specific. Therefore, HISTOPATHOLOGY is necessary.[18]

Initially, the biopsy can be taken by fine-needle aspiration. However, it was shown that aspiration performed with the help of EUS increases the diagnostic accuracy rate for diagnosing PPL. CT-assisted biopsy, which is less invasive, is another sampling option.[19]

Biopsy can be taken during laparoscopic or laparotomic exploration. In our case, the diagnosis was based on the biopsy obtained during laparotomy.[20]

Treatment

The strategy for treating PPL includes surgery, chemotherapy, radiotherapy, or a combination of these. [21] Most chemotherapy protocols include CHOP (cyclophosphamide, hydroxydaunorubicin, oncovine, prednisone) and sometimes Rituxan along with CHOP (R-CHOP).[22]

Most patients with PPL who receive only chemotherapy can recover from the pathology in the long term. The 5-year survival rate of PPL patients treated with chemotherapy is lower compared to patients treated with a combination of surgery and chemotherapy.[22]

Surgical intervention, however, is controversial, as removal of the pancreas alone has been proven not to increase survival rates in PPL. For patients with mechanical jaundice, choledochostomy is an effective treatment option. [23, 24]

The role of radiotherapy has yet to be defined except as an adjuvant to chemotherapy. A study by Behrns showed some evidence that an initial surgical removal, combined with chemotherapy and radiotherapy, increases the survival of patients with PPL.[20]

The choice of the appropriate treatment should be made depending on the general condition of the patient as well as the evolution of the disease. [19]

Conclusion

We present a rare case of PPL, some clinical and radiological features, and biochemical signs that are not specific to PPL. Patients with PPL have a much better prognosis than those with adenocarcinoma, and treatment differs widely from that of other pancreatic tumors. It is based on a precise diagnosis made possible by histologic examination. Combined therapy remains the most optimal treatment approach for PPL but needs further evaluation.

COI Statement: This paper has yet to be submitted in parallel, presented fully or partially at a meeting, podium, or congress, published, or submitted for consideration beforehand.

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Patient consent: The patient's consent was obtained for the publication of the case.

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