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Case report

Primary pancreatic lymphoma in a young adult male: A challenging diagnosis

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Abstract. Primary pancreatic lymphoma (PPL) is a rare clinical entity most likely to be misdiagnosed as pancreatic adenocarcinoma on presentation. The most common histological type of PPL is non-Hodgkin's lymphoma. We present a case of a 48-year-old man admitted to our hospital for abdominal pain and progressive jaundice. A computed-tomography (CT) and magnetic resonance imaging (MRI) scan were performed and a large mass with slight enhancement after contrast agent administration at the pancreatic head was found. The patient underwent endoscopic-ultrasound (EUS) guided fine needle aspiration of the pancreatic mass. Histopathological diagnosis revealed a primary pancreatic lymphoma. We discuss the typical features of computed tomography (CT) and MRI of this rare clinical condition to help radiologists in the timely diagnosis for faster and more accurate diagnostic framing and to direct the patient to correct and timely treatment. (www.actabiomedica.it)

Key words: primary pancreatic lymphoma, body imaging, computed tomography, magnetic resonance imaging, case report

Introduction

Primary pancreatic lymphoma (PPL) is a very rare condition, accounting for only 0.1% of malignant lymphomas and 0.2% of primary pancreatic tumors (1, 2).

The World Health Organization has provided specific diagnostic criteria for this disease: first, the disease must be located mainly in the pancreas, adjacent lymph nodes involvement and distant spread can co-exist. The primary clinical presentation has to involve primarily the pancreatic gland (3).

PPL can develop at any age, but generally affects patients in the 5th or 6th decade of life and has a slight male prevalence.

Clinical symptoms are usually not specific and include mainly abdominal pain, palpable abdominal mass, weight loss; additional symptoms may be present such as jaundice, nausea, diarrhea, vomiting, intestinal obstruction, and pancreatitis symptoms.

Jaundice is often observed as this pathology primarily involves the pancreatic head in more than 80% of cases (4,5).

Case presentation

A 48-year-old man was admitted to the Emergency Department of our hospital for recurrent

episodes of abdominal pain in the last period associated with progressive jaundice and widespread itching.

The patient also reported asthenia, lack of appetite and a weight loss of about 4 kg in the last two weeks.

The patient denied the use of drugs in the last period, not having had high-risk sexual behavior and not having gone abroad for travel in the last period.

The clinical examination documented the presence of a tense abdomen, widely painful, preferentially at the upper abdominal quadrants.

Laboratory testing identified a high total bilirubin level of 4.9 mg/dL, normal aspartate and alanine transaminases level, high serum amylase level 453 U/L and serum lipase level 300 U/L (normal values: total bilirubin less than 1 mg/dL; serum amylase level 30 to 110 U/L; serum lipase level 0-160 U/L respectively). His complete blood count, renal biochemistries, and coagulation studies were within normal limits as well as tumor markers (AFP, CA125, CA 19-9 and CEA). The patient then underwent a radiological examination with CT and MRI. Positron emission tomography-CT (PET-CT) was not performed.

CT showed a large, low-attenuating mass with slight enhancement after contrast agent administration at the pancreatic head. There was no dilation of the pancreatic duct upstream nor signs of infiltration and encasement of the surrounding arterial and venous vessels (Figure 1 A, B and C).

The MRI examination confirmed the neoformation at the pancreatic head as an unevenly hyperintense area in T2-weighted sequences that presented

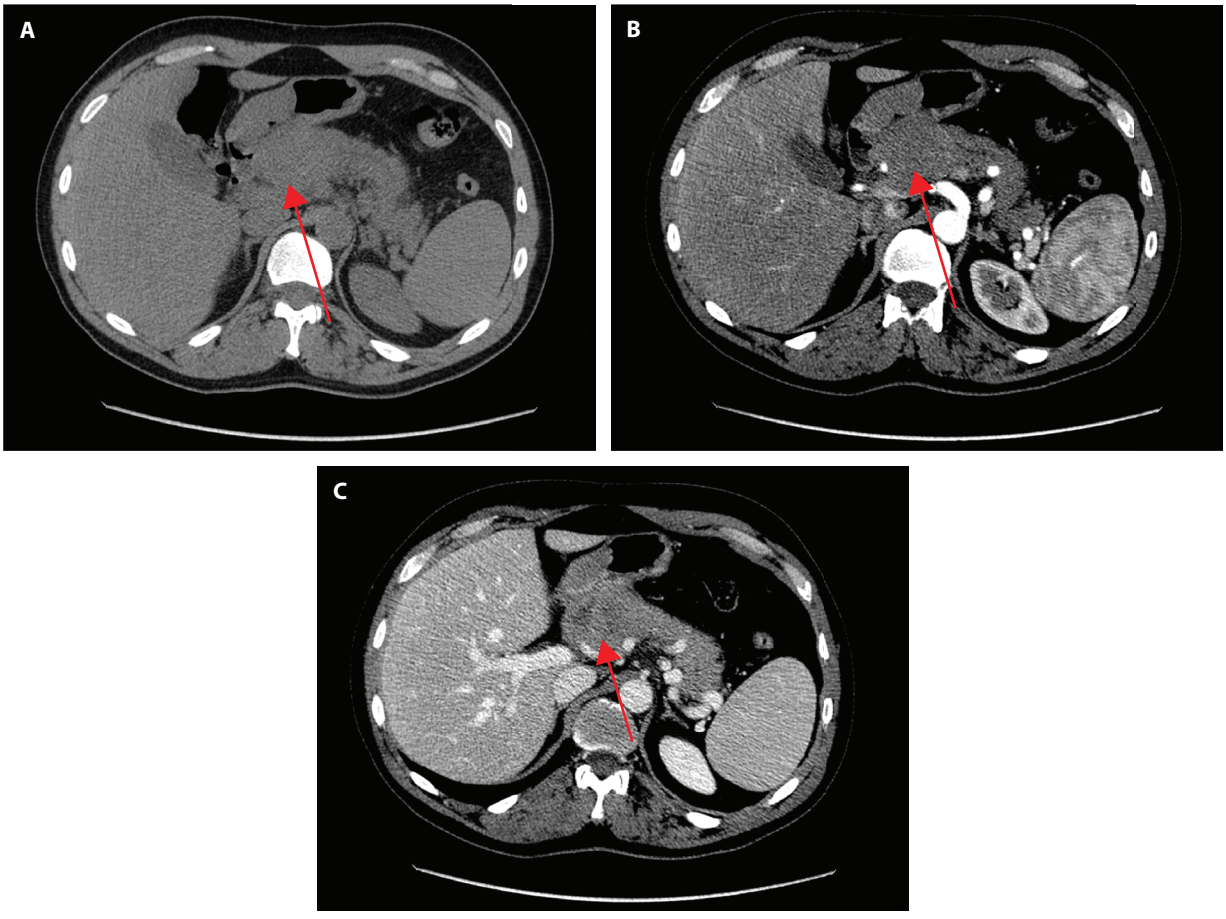

Figure 1. CT study. A) unenhanced CT scan shows a hypodense mass in the pancreatic head. CT images after contrast medium administration in the arterial-pancreatic (B) and portal-venous phase (C) show progressive enhancement of the neoplasm with low attenuation compared to residual pancreatic parenchyma. Some enlarged lymph nodes were documented in the peripancreatic fat.

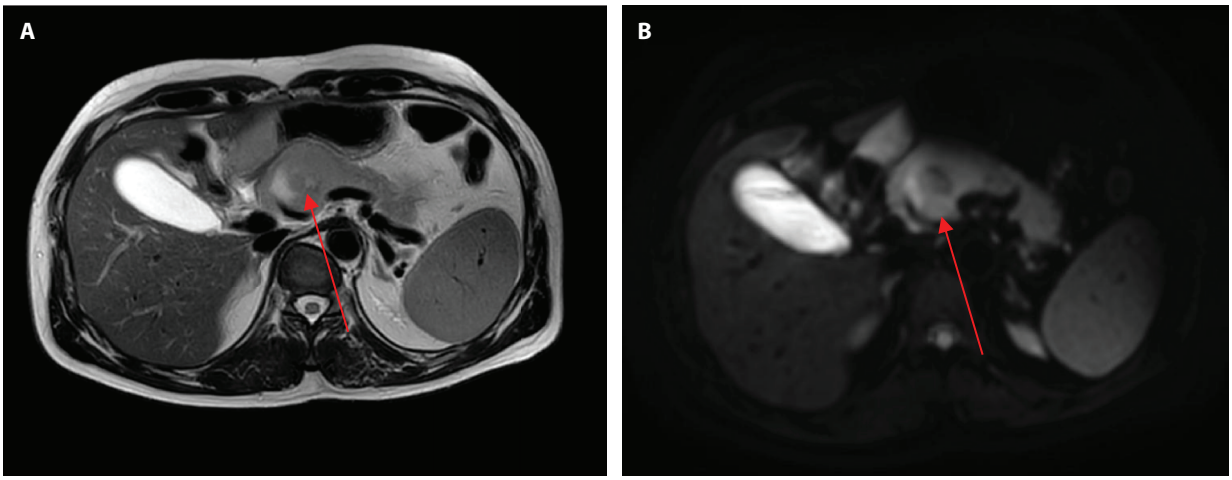

Figure 2. A and B. MRI study. A) Axial T2-weighted image shows a heterogeneously hyperintense masse in the pancreatic head B) axial DWI b800 image shows heterogeneous hyperintensity at the expansive process of the head of the pancreas.

hyperintense signal in the DWI-weighted sequence at high b values (b800) (Figure 2 A and B).

After imaging features the patient underwent an endoscopic-ultrasound (EUS) examination to perform a biopsy of the pancreatic head lesion.

The tissue frustule was made up of atypical lymphoid neoplastic elements positive for the markers CD20, PAX5, Bcl2 and Bcl6 and with a high proliferative index (Ki67: 95%). Histology confirmed the diagnostic hypothesis of primary pancreatic lymphoma.

Therefore, the patient was sent to a specialist oncologist to carry out the appropriate therapies.

Discussion

PPL is a rare clinical entity most likely to be misdiagnosed as pancreatic adenocarcinoma on presentation.

The most common histological type of PPL is non-Hodgkin's lymphoma; diffuse large B cell lymphoma, as in our case, is the most common type of non-Hodgkin's lymphoma and represents 30% of newly diagnosed cases (6).

The diagnostic criteria of PPL include mass located primarily in the pancreas, with involved lymph nodes confined to the peripancreatic area, no hepatic or splenic involvement, no palpable superficial lymphadenopathy, no mediastinal nodal enlargement, and a normal white cell count (7).

PPL occurs mostly between the fifth and sixth decade of life, with a predilection for the male sex (male: female 7:1) and is manifested mostly at the pancreatic head (8).

Clinical features are often non-specific and similar to other pancreatic diseases. Abdominal pain is the most common presenting symptom, followed by abdominal mass, weight loss, jaundice, acute pancreatitis, small bowel obstruction, and diarrhea.

The clinical presentation of patients with PPL of the head of the pancreas can often correspond to that of patients with pancreatic adenocarcinoma, therefore a correct radiological differential diagnosis is difficult. Obstructive jaundice is less frequent than in pancreatic adenocarcinoma (4).

Typical systemic symptoms of non-Hodgkin's nodal lymphoma, such as fever, chills, and night sweats, are infrequent.

Transabdominal ultrasonography, endoscopic ultrasound (EUS), CT and MRI are well-established modalities for evaluating pancreatic lesions.

On CT imaging with intravenous contrast, the majority of lesions are presented as well defined, sometimes voluminous and infiltrating, homogeneous masses with low attenuation compared to pancreatic parenchyma enhanced with only mild enhancement.

Less frequently, pancreatic lymphoma may present as a diffuse enlarged gland with an infiltrating tumor that could mimic acute pancreatitis; typical

features of acute pancreatitis, including peripancreatic fat stranding and peripancreatic inflammation, are usually minimal if even present. While vessels can be stretched due to a mass effect, irregularities, and size changes due to tumor invasion are generally absent (4). This peculiarity is useful to make a differential diagnosis with ductal adenocarcinoma, which causes vascular infiltration already in the initial stages of disease, with associated erosion of the walls of the vessels, stenosis, and possible neoplastic thrombosis (9).

At MRI imaging, pancreatic lymphomas usually appear as homogeneous masses, with low signal intensity on T1WI and variable signal intensity, low or high, on T2WI and generally less enhanced than the surrounding parenchyma on DCE-MRI (4,10).

Diffusion-weighted imaging can recognize areas of pancreatic parenchyma implicated in neoplasm and could play a role in the assessment of treatment (11).

The rarity of PPL makes it difficult to make a proper differential diagnosis with other kinds of pancreatic neoplasms such as ductal adenocarcinoma, pancreatic cancer more frequent or, more rarely, with hypovascular neuroendocrine tumors.

PPL could also be confused with the focal or diffuse forms of autoimmune pancreatitis; all these conditions present different management approaches, with distinct treatment and prognosis (1,12,13).

The definitive diagnosis is established only after histopathological and cytopathological examinations with molecular confirmatory tests since the clinical and radiological characteristics of this pathology are not pathognomonic.

Two possible options for tissue sampling are EUS-guided biopsy and secondly CT-guided biopsy which is minimally invasive.

Management of PPL consists of surgery, chemotherapy, radiotherapy or a combination of both. High-grade pancreatic lymphoma typically responds well to standard chemotherapy, and long-term remission of the disease may be achieved with chemotherapy alone. Surgery is still controversial, and it has been shown that pancreatic resection alone does not improve the survival rate of PPL (14,15).

Conclusion

In summary, we exposed a rare case of primary pancreatic lymphoma (PPL) discovered by CT and MRI investigation and subsequently confirmed by histopathological examination.

If a pancreatic mass is found, primitive pancreatic lymphoma, though rare, should be considered as a possible diagnosis. This is the fundamental reason why it is necessary to know how to recognize the imaging characteristics of such pathologies, differentiate them from other pancreatic pathologies to achieve rapid radiological diagnosis, avoid wrong diagnoses and unnecessary surgery.

Nevertheless, histological diagnosis is often essential to arrive at a definitive diagnosis.

Conflict of Interest: Each author declares that he or she has no commercial associations (e.g. consultancies, stock ownership, equity interest, patent/licensing arrangement etc.) that might pose a conflict of interest in connection with the submitted article.

Ethical Considerations: We hereby certify that we had a written statement of informed consent from the patient and obtained his permission to submit the case report for publication.

Authors Contributions: Concept and design was performed by U.T. and V.T.; provision of study patients was performed by U.T, V.T., N.M., C.S. and M.C.; collection of data and data analysis was performed by U.T, V.T., N.M., C.S., M.C., F.S.G., D.M. and R.B.; all the authors participated in manuscript writing; all the authors approved the final version of the manuscript.

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