A Case of Primary Pancreatic non-Hodgkin B-cell Lymphoma Mimicking Autoimmune Pancreatitis

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INTRODUCTION

Non-Hodgkin lymphoma (NHL) frequently involves the gastrointestinal tract, in particular the stomach and the small bowel. Rarely, it can also be a cause of pancreatic masses. Clinical presentation is often non-specific and may overlap with other pancreatic conditions such as carcinoma, neuroendocrine tumours and autoimmune pancreatitis. We report a case of primary pancreatic lymphoma in a young woman with jaundice, fever and abdominal pain mimicking autoimmune pancreatitis. Clinical evaluation included the abdominal Computed Tomography scan, Magnetic Resonance Imaging and an upper gastrointestinal endoscopy that revealed a large duodenal mass. Endoscopic biopsies were performed and eventually histological examination was coherent with a diagnosis of primary pancreatic lymphoma.

Key words: primary pancreatic lymphoma – autoimmune pancreatitis – non-Hodgkin lymphoma.

CASE REPORT

A 33-year-old woman was admitted to the emergency unit for jaundice, fever and a two week history of abdominal pain radiating to the back. Past medical history was unremarkable except for gestational diabetes 8 months previously. At presentation, the laboratory tests showed total bilirubin 7.5 mg/dL (0.3-1.2 mg/dL), conjugated bilirubin 5 mg/dL, alanine aminotransferase 578 IU/L (0-40 IU/L), aspartate aminotransferase 110 IU/L (0-40 IU/L), alkaline phosphatase 1135 IU/L (70-290 IU/L), gamma-glutamyl transpeptidase 376 IU/L (0-50 IU/L), CA 19-9 33.3 U/mL (0.0-37 IU/mL)
with normal blood count. Abdominal ultrasound revealed two pancreatic hypoechoic lesions in the head (3 x 3.5 cm) and in the isthmus (2 cm), with one peripancreatic lymph node of 10 mm. Abdominal Computed Tomography (CT) scan with iodinate contrast medium showed a dilation of the common bile duct (diameter 10 mm) with a pancreatic hypodense mass (4.5 x 4 x 5 cm) involving the head of the pancreas with diffuse enlarged “sausage-shaped” pancreatic gland and irregular narrow pancreatic duct suggestive for autoimmune pancreatitis (Fig. 1). Furthermore, abdominal Magnetic Resonance (MRI) showed a hypovascular enlargement of the whole pancreas with homogeneous enhancement in the tardive phases and multiple bilateral hypovascular renal lesions (Fig. 2). Since serum IgG4 levels were elevated (183 mg/dL, normal values 8-140 mg/dL), a diagnosis of AIP was made and steroid treatment was started (prednisone 1 mg/Kg), followed by progressive jaundice resolution (bilirubin at discharge: 2.4 mg/dL).

After three weeks of therapy the patient was readmitted for jaundice with serum bilirubin 6.5 mg/dL (conjugated bilirubin 5 mg/dL). The laboratory tests revealed a mild leucocytosis (white blood cells count 15,740/mmc). Radiological findings were similar to the previous ones. During hospitalization the patient developed nausea and severe epigastric pain; therefore, an upper gastrointestinal endoscopy (UGIE) was performed, which revealed a duodenal ulcerated sessile lesion proximal to the papilla (diameter 3 cm) (Fig. 3). The histological examination of the prelevated biopsies concluded for a non-Hodgkin B cell lymphoma (Figs 4, 5). The patient was then referred to oncologists and started chemotherapy. One year after the initial diagnosis her clinical condition is stable, with complete resolution of the duodenal and pancreatic lesions.

**DISCUSSION**

Primary pancreatic lymphoma accounts for less than 1% of all malignant lymphomas and for 0.2% of all pancreatic malignancies [1, 6]. It frequently represents a diagnostic
Primary pancreatic non-Hodgkin B-cell lymphoma mimicking autoimmune pancreatitis

Fig. 4 A, B. Lymphoid infiltrate (cells of large size) with prevalent expression of anti CD20 antibody supporting a diagnosis of B cell non-Hodgkin lymphoma (x40).

Fig. 5 A, B. Intense lymphoid infiltrate characterized by large pleomorphic elements with irregular nuclei and scant cytoplasm, surrounding pancreatic glandular structures, without cohesion and organization. Pancreatic glands are entrapped by cells that do not show atypias (H&E x40).

challenge as it shares clinical and radiological findings with other pancreatic disorders such as other neoplasms and AIP, an inflammatory disease of the pancreas that usually presents with painless obstructive jaundice, pancreatic mass/enlargement and response to steroids administration. Diagnostic criteria for AIP include: typical imaging findings on CT/MRI dynamic scans, elevated levels of serum IgG4, other organs involvement (renal masses, tubulointerstitial nephritis, sclerosing cholangitis, retroperitoneal fibrosis, submandibular masses), steroid responsiveness and, when available, histology with immunostaining. In cases with typical CT/MRI features for AIP and serum elevation of IgG4 or other organs involvement, a diagnosis of type 1 AIP can be made without tissue confirmation, reserving histological/cytological examination to patients with non-conclusive results on other examinations [7].

We describe the case of a young woman with a PPL mimicking an AIP. Clinical presentation of the disease and radiological appearance including renal involvement were coherent with a diagnosis of AIP, eventually confirmed by the presence of elevated levels of IgG4; the initial clinical response to administration of steroids gave further strength to the initial hypothesis. When typical clinical, radiological and serological findings are present, diagnosis of AIP does not include histology [7]. In this particular case, the relapse of jaundice after steroids, administration required further evaluations and tissue sampling became mandatory. Endoscopic ultrasound guided needle aspiration or CT guided core biopsies were not conducted since UGIE revealed the presence of a duodenal mass that was easily biopsied allowing the diagnosis of a PPL.

Gastrointestinal involvement by pancreatic lymphoma is reported in the literature as a possible but infrequent presentation of the disease [6]. To our knowledge, the only recent case was reported by Dupre et al. in 2011 as a fatal upper gastrointestinal hemorrhage due to erosion of the duodenal wall by a pancreatic lymphoma [8].

After diagnosis, our patient was successfully treated with chemotherapy. Common protocols consider the use of anti CD20 antibodies or anthracyclines with satisfactory results and the 5 years mortality refers to 45% of the cases [5, 9-11].

CONCLUSION

The present case underlines the fact that conventional diagnostic criteria for AIP are not always adequately sensitive and specific to achieve a clear differential diagnosis with other pancreatic diseases, especially in the presence of overlapping clinical and radiological presentation. The therapeutic approach and prognosis of these two diseases are dramatically different, and given the risk of possible misdiagnosis or delay in diagnosis, further assessment is advisable when doubts are present and novel diagnostic methods should be investigated.

Conflicts of interest: None to declare.

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REFERENCES