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Case Report

Symptomatic Pancreatic Involvement Revealing B Lymphoblastic Lymphoma

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Abstract

Lymphoblastic lymphoma (LBL) is a rare cancer that accounts for less than 2% of all non-Hodgkin malignant lymphomas. The pancreatic localization of LBL is extremely rare and is still little known in current medical practice. It is most often asymptomatic and fortuitous discovery.

We report an original observation of acute pancreatitis revealing type B LBL (B-LBL) in an 8-year-old child, with concomitant multi-organ infiltration (pancreatic, renal, muscular, and retro-peritoneal).

As rare as it is, this digestive complication of B-LBL deserves to be known by the hospital practitioners and it is advisable to think of this diagnosis in front of an acute pancreatitis of the child which is not proven.

Keywords: Acute Pancreatitis; Lymphoblastic Lymphoma; Pancreatic Involvement; Pancreas; Non-Hodgkin Lymphoma

Introduction

Lymphoblastic lymphoma (LBL) is a rare cancer that accounts for less than 2% of all non-Hodgkin malignant lymphomas [1-3]. It is particularly common in male children, but can occur in both sexes and at any age [1-3]. It is 90% of cases of type T (T-LBL), while LBL type B (B-LBL) is much rarer (10% of cases) [3].

Concomitant extra-medullary infiltration of several organs remains uncommon and unusual during the B-LBL; in particular pancreatic and renal involvement [4-8]. These lesions are most often asymptomatic and are fortuitous discovery during radiological investigations; the symptomatic forms of these localizations are exceptional and often represent a real diagnostic challenge [5,9].

We report an original observation of symptomatic pancreatic localization revealing B-LBL with multi organ infiltration in an eight-year-old child.

Case Report

8-year-old boy, with no notable pathological history, was admitted for acute abdominal pain associated with bilious vomiting.

The somatic examination noted a febrile child at 39°C, skin and conjunctival pallor, a sensitive epigastrium, and a hard and sensitive mass in the right flank.

Biology showed a marked inflammatory syndrome with an erythrocyte sedimentation rate at 90 mmH1, a C-reactive protein at 32 mg/l and a fibrinogen at 11 g/l, a normochromic normocytic anemia at 10 g/dl, hyperamylasemia at 180 IU/l (N <45 IU/l) and hyperlipasemia at 220 IU/l (N <60 IU/l) consistent with the diagnosis of acute pancreatitis. The rest of the basic biological tests were within normal limits.

Doppler abdominal ultrasonography showed enlarged heterogeneous pancreas (Figure 1) and two enlarged multi-nodular kidneys, with a large heterogeneous and hypervascularized right retroperitoneal mass (Figure 2). The liver and gallbladder were without abnormalities.

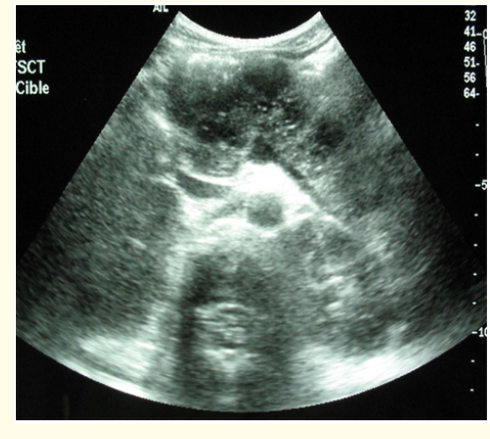

Figure 1: Abdominal ultrasound: enlarged, heterogeneous, and polylobed pancreas.

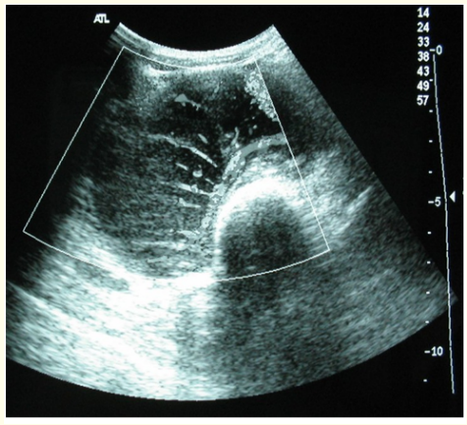

Figure 2: Abdominal Doppler ultrasound: hypervascularized retroperitoneal mass.

The abdominal computed tomography (CT) showed multiorgan involvement with a multi-nodular appearance of both kidneys, pancreas, psoas muscles and the presence of a large right para-vertebral mass (Figure 3).

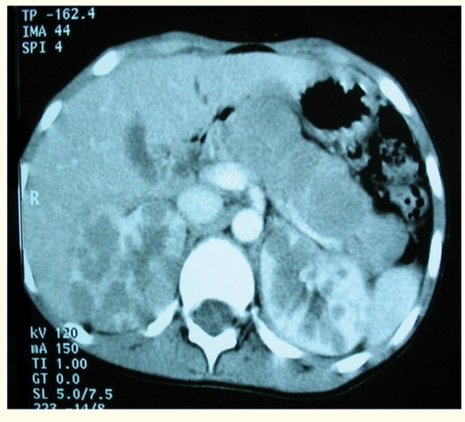

Figure 3: Axial abdominal CT with contrast: multiple and richly vascularized pancreatic and bilateral renal masses.

Abdominal, thoracic and cerebral nuclear magnetic resonance imaging (MRI) confirmed similar findings in the abdomen (Figures 4-6) and did not revealed thoracic or cerebral involvement.

CT-guided biopsy of the retro-peritoneal mass and immunophenotype by flow cytometry led to the diagnosis of B-LBL. The myelogram showed infiltration with 10% blasts.

At the end of these investigations, the diagnosis of acute pancreatitis secondary to a pancreatic localization of an advanced B-LBL, associated with renal, retroperitoneal, and muscular infiltration was retained. The patient was transferred to medical oncology department for adapted chemotherapy.

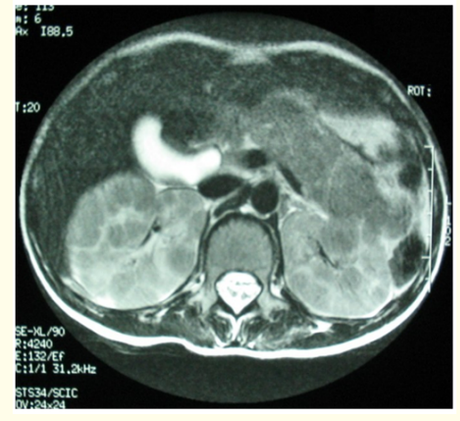

Figure 4: Axial abdominal T2-weighted MRI: Multinodular enlargement of the pancreas and the two kidneys with spontaneous hypersignal.

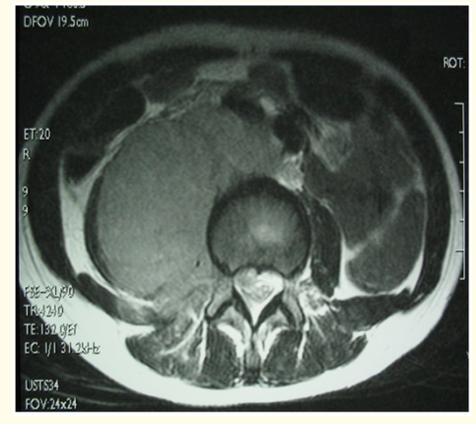

Figure 5: Axial abdominal T1-weighted MRI: large retroperitoneal right para-vertebral mass with hyposignal.

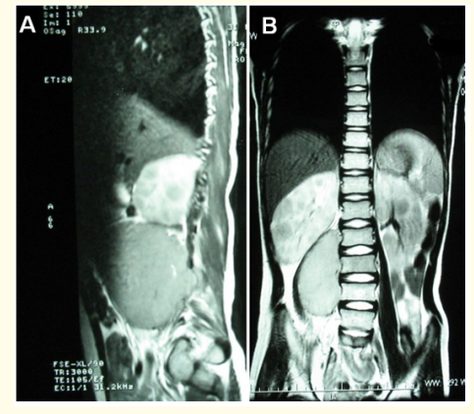

Figure 6: T1/Fatsat-weighted abdominal MRI with gadolinium injection in sagittal (A) and frontal (B) sections: right para-vertebral mass with discretely heterogeneous signal, enlarged and multinodular two kidneys and pancreas.

Discussion

In 80% of cases, the B-LBL is presented in acute form with bone marrow infiltration (> 20% bone marrow blasts according to the 2017 World Health Organization classification) and peripheral blood involvement defining particular form of B lymphoblastic lymphoma leukemia [2,3]. Tumor forms of B-LBL presenting with masses in different tissues and organs are rarer [2]. Lymph nodes are the preferred seat of B-LBL [3]; subsequently come skin, bones, soft tissues, spleen, and liver [3].

The pancreatic localization of B-LBL is extremely rare [10,11], and is still little known in current medical practice [8]. This localization can remain totally asymptomatic and would be accidentally discovered on radiological examinations [5]. Sometimes we can note a purely biological expression (hyperamylasemia/hyperlipasemia) without clinical signs [6], and more rarely the clinic may be of the type of abdominal pain, vomiting, acute pancreatitis, obstructive jaundice, or symptoms of glycemic abnormalities [7,8,10,11].

Pancreatic involvement of B-LBL may be present at the time of diagnosis [8] or may occur secondarily during relapses [7]. It fits most often, as our case, in the context of a diffuse form with multi organ involvement [4,11,12], but can remain isolated and primitive [10].

The renal involvement of B-LBL is also rare [9,13], and often represents a diagnostic challenge [9]. It is classically presented as a bilateral enlargement of the kidneys [9,14]; more rarely there are abnormalities of urinary sediment, tubulopathies, ionic and acid-base balance disorders, and acute renal failure [9,13,14].

The treatment of choice is adapted chemotherapy with a fairly satisfactory response rate. However, relapses remain frequent during these locations [1-3,9,13,14].

Conclusion

Pancreatic infiltration is rare and often unrecognized during B lymphoblastic lymphomas. Acute lymphomatous pancreatitis is an exceptional and unusual clinical manifestation of this localization.

Our observation is further characterized by concomitant renal, muscle and retroperitoneal involvement. It is thus necessary to think of this diagnosis in front of any acute pancreatitis of the child which is not proven.

Conflicts of Interest

None

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