

Management of Hepatic Burkitt Lymphoma in Children

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Case Report

Volume 3 Issue 1

Received Date: June 02, 2018

Published Date: June 15, 2018

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Abstract

Hepatic Burkitt lymphoma is extremely rare in childhood and can be overlooked in differential diagnosis of liver masses.

Patient: A8-year-old girl presented with a 1 month history of abdominal pain and weight loss and jaundice.

Results: Physical examination revealed hepatomegaly and no palpable lymph node. Laboratory finding showed mild anemia (hemoglobin, 10,8 g/dL), elevated transaminase (ALT, 305 IU/L; ASAT, 755 IU/L), elevated bilitubin (Bilirubin total, 179mg/L, Bilirubin direct, 143mg/l). Abdominal ultrasound showed a multiple hepatic lesions. Liver biopsy examination confirmed Burkitt's lymphoma. No metastasis was detected in the thoracic cavity, bone marrow, and spinal fluid. The patient was treated with the combination regimen of cyclophosphamide, doxorubicin, vincristine, prednisone and high dose methotrexate. Cytosine arabinoside and methotrexate were added for CNS prophylaxis by intrathecal installation. Serial follow-up ultrasound showed a marked decrease in the size of hepatic lesions but residual hilar lymph nodes at 2cm and the control showed stable size of lymph node after 28 months of chemotherapy.

Conclusion: The clinical feature of primary hepatic lymphoma varies from no symptom to fulminant hepatic failure. There are no specific imaging criteria for diagnosing primary hepatic Burkitt's lymphoma. Thus, histology by biopsy is necessary for diagnosis.

Keywords: Liver; Burkitt's lymphoma

Introduction

Burkitt lymphoma (BL) is a highly aggressive B-cell non-Hodgkin lymphoma. Its primary hepatic localization is rare entity (PH-BL) [1]. We describe a case of PH-BL presenting with a jaundice and abdominal pain.

Case Report

An 8 years old girl, presented with abdominal pain and jaundice and weight loss for 1 month before her admission. There was also a 3 days of history of moelena without fever or diarrhea. Physical examination revealed

hepatomegaly measured 26cm below the subcostal margin, no palpable lymphnode or spleen.

Laboratory findings were Hemoglobin 10,8gr/dL, Platelet 406000/mm³, WBC 13460/mm³, alanin aminotransferase 3005(ALT) 23 IU/L, aspartate aminotransferase (AST) 755 IU/L, total bilirubin 179,3mg/L, direct bilirubin 143 mg/L and lactate dehydrogenase (LDH) 500/L. Alpha foetoprotein was normal at 8,9UI/ml. Coagulopathy test (Prothrombin time and partial thromboplastin time) was normal.

Abdominal ultra sound showed multiple large low-density mass lesions in both lobes of liver with no enhancement, with hilary lymphnodes but no thrombosis was evident. No bile duct dilatation was noted (Figure 1). Chest x-ray was normal.

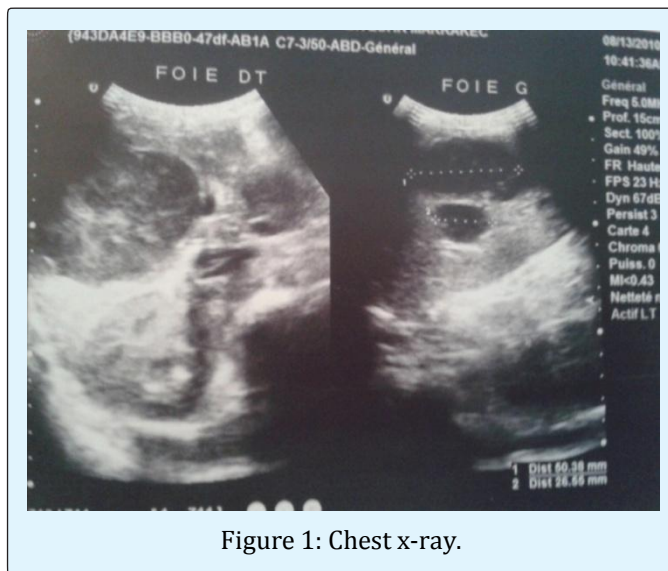


Figure 1: Chest x-ray.

Ultra sonography- guided liver biopsy was done. Microscopic evaluation showed diffuse infiltration of monotonous small to medium sized cells with vesicular nuclei, dense chromatin pattern, prominent nucleoli. Mitotic figures were frequent, and many scattered tangible macrophages creating starry-sky pattern. Immuno histochemical study was not available. Spinal fluid analysis and bone marrow biopsy results were negative. Endoscopy, to explore hermeloena, was not done because patient didn't present any episode at her admission.

She received chemotherapy according to MAT IV (MoroccoAlgeria and Tunisia)/ GFAOP (Franco-africain

pediatriconcology group) protocol. The patient had full regression of jundice at first and hepatomegaly since the first course. Her ALAT and ASAT levels normalized.

Abdominal US control showed a regression of hepatic lesions and lymphnodes, but appearance of coeliomesenteric adenopathy at 22mm which persisted since 28 months after the end of treatment and disappear after. Last control of patient were at 4 years after treatment and abdominal US was normal (Figure 2).

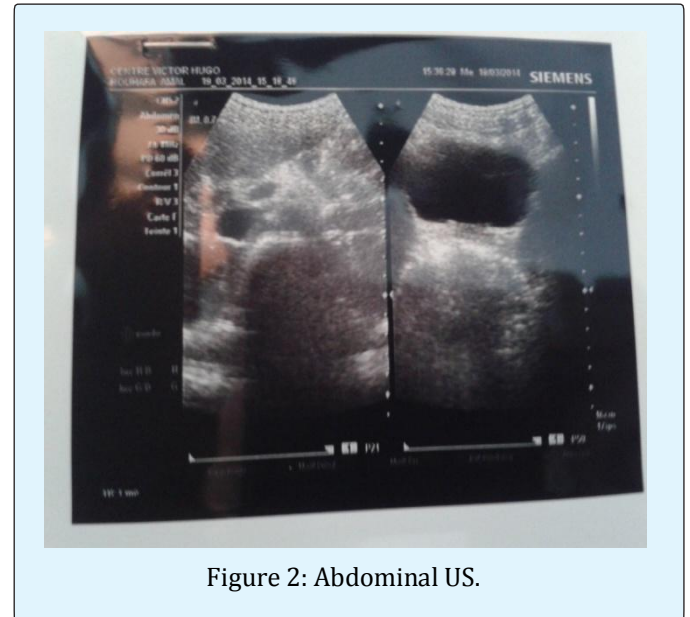


Figure 2: Abdominal US.

Discussion

According to Lei criteria in primary hepatic lymphoma symptom expression mainly originate from liver infiltration with no distant lymphadenopathy or leukemoid reaction in peripheral blood smear [2,3]. PH-BL, a highly aggressive subset of NHL, is a very rare entity [2]. There have been no more than 15 cases of primary hepatic Burkitt's lymphoma reported. Most of the reported patients were young, aged about 32 years old on average, and male, with a large proportion being children [4].

It is accounting for only 0.4% of all extra nodal lymphomas and make up to 3% of primary hepatic lymphomas [3]. It was a first case in our unit in Morocco. On histopathologic examination, Burkitt's lymphomais defined as infiltration of neoplastic cells, which are marked lyuniform in size and shape. The nuclei are approximately the same size as the nuclei of the admixed

histiocytes and therefore are smaller than the centroblasts and contain two to four basophilic nucleoli. The nuclear contours are generally round without deep indentation. The cytoplasm is strongly basophilic with small round cytoplasmic vacuoles best observed in air-dried touch imprints [5].

The presence of tangible body macrophages, phagocytosing abundant apoptotic debris creating starry-sky appearance is characteristic finding [6]. On immunohistochemical examination the neoplastic cells are positive for surface IgM and Ig light chain ($\kappa > \lambda$), pan-B-cell antigens (CD19, CD20, CD22, CD79 α) and the germinal center associated markers such as CD10 and BCL6.

The clinical feature of primary hepatic lymphoma varies from no symptom to fulminant hepatic failure. Fever, weight loss, night sweating (known as B symptoms), right upper quadrant pain, hepatomegaly, fatigue, jaundice, nausea, vomiting, and splenomegaly are common symptoms and rarely, bleeding tendency, ascites, pleural effusion, hepatic encephalopathy can occur [2].

But the common clinical manifestations in the reported cases were abdominal pain, pyrexia and the finding of hepatomegaly on physical examination [3]. There are no specific imaging criteria for diagnosing of PHBL. Sociological or histological exams are necessary [3].

Early administration of multiple chemotherapeutic agents increases the curative potential of Burkitt's lymphoma and overall survival make up 87% at 5 years after oncological treatment [2,7]. Prognosis of patients with primary hepatic Burkitt's lymphoma does not necessarily seem to be unfavorable [4]. Very high cure rates and long term survival are possible with intensive chemotherapy regimens [8].

Conclusion

The best method for the diagnosis of hepatic lymphoma is needle biopsy of the liver guided by radiological exam. Surgery in the management of PHBL is

not well defined but aggressive combination chemotherapy is necessary.

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