Case Report

A Primary Hepatic Lymphoma Treated with Liver Resection and Chemotherapy

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Received 4 May 2014; Revised 12 July 2014; Accepted 13 July 2014; Published 4 August 2014

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Primary hepatic lymphoma (PHL) is a rare malignancy, which is frequently misdiagnosed. Although chemotherapy is the treatment of choice there are reports that a combination of surgery and adjuvant chemotherapy can offer better results. Herein we present an interesting case of a large primary non-Hodgkin lymphoma originating from liver which was treated with a liver resection and chemotherapy.

1. Introduction

Primary hepatic lymphoma (PHL) is an extremely rare malignancy, accounting for less than 0.4% of extranodal non-Hodgkin lymphomas and 0.016% of all non-Hodgkin lymphomas [1]. The majority of PHL cases originate from B cells while T-cell lymphoma is less common. The etiology of PHL is unknown and although the liver contains lymphoid tissue, host factors seem to make the liver a poor environment for the development of malignant lymphomas [2]. Herein we present an interesting case of a large primary non-Hodgkin lymphoma originating from liver which was treated with a liver resection and chemotherapy.

2. Case Report

A 57-year-old woman presented to the outpatient clinic complaining of upper abdominal discomfort, nausea, fatigue, intermittent low-grade fever, and weight loss in the last 5 months. Due to these symptoms a gastroscopy was done 4 months earlier which showed H. pylori infection for which she was given medical treatment. Otherwise, she had no remarkable past medical history. On physical examination a large, tender mass was found at the epigastrum with significant hepatomegaly but there was no splenomegaly or peripheral adenopathy. Peripheral blood cell counts revealed a mild anemia (hemoglobin 9.8 g/dL) and elevated white cell count (13.9 × 10^9/μL). Liver function tests were as follows: aspartate aminotransferase (AST) 47 IU (normal range <31); alanine aminotransferase (ALT) 22 IU (normal range <34); alkaline phosphatase (ALP) 134 IU (normal range <120); gamma-glutamyl transpeptidase (γGT) 147 IU (normal range <38); and lactate dehydrogenase (LDH) 555 IU (normal range <247). Serological tests for hepatitis B and C virus were negative. Serology for HIV was negative too. The levels of tumors markers were cancer antigen 19-9 (Ca19-9) 58.1 IU/mL (normal range <27), carcinoembryonic antigen (CEA) 31.5 IU/mL (normal range <5), and cancer antigen 125 (Ca 125) 88, 9 IU/mL (normal range <35). Alpha-fetoprotein (AFP) and carcinoembryonic antigen (CEA) were within normal values. Contrast-enhanced computed tomography (CT) revealed a large, solid mass (15 × 10 × 9 cm) occupying the left and partially the right hepatic lobe with heterogeneous enhancement. A magnetic resonance imaging (MRI) showed a low signal in
T1 and high signal in T2 sequences mass with heterogeneous density (Figure 1). Our differential diagnosis included (1) primary hepatocellular cancer, (2) focal nodular hyperplasia, and (3) metastasis from gastrointestinal cancer. The latter was ruled out with a gastroscopy and a colonoscopy. The tumor was symptomatic and potentially respectable and thus an exploratory laparotomy was carried out. During the operation a solid and well-circumscribed mass was found on the left and partially on the right hepatic lobe and an extended left heptectomy was performed (Figure 2). No palpable lymph nodes were evident in the mesentery, the hepatoduodenal ligament, or the retroperitoneal space. The gastrointestinal tract was normal too. The histopathological examination showed large lymphoid cells with oval to round vesicular nuclei containing fine chromatin. There were one, two, or multiple nucleoli and numerous mitotic figures. Necrotic cells were also observed (Figure 3). Immunohistochemistry was positive for the pan-B-cell marker CD20 (Figure 4), as well as LCA and CD43, while it was negative for CD3, CD5, CD10, CD138, MUM1, bcl6, and bcl2. The lymphoid cells showed also a high proliferation fraction as detected by Ki67 (MIB-1) immunostaining (Figure 5). The diagnosis of B-cell non-Hodgkin lymphoma of high malignancy was made. The patient had an uneventful recovery with regression of B symptoms and she was discharged on the 10th postoperative day. She was referred to the department of hematology for further treatment. Bone marrow aspiration as well as thoracic and neck CT scans was normal. The patient was given four cycles of chemotherapy including rituximab, cyclophosphamide, hydroxydaunorubicin, Oncovin, and prednisone (R-CHOP) and twelve months after the operation she is disease-free.

3. Discussion

Primary hepatic lymphoma (PHL) is defined according to Caccamo’s criteria as lymphoma confined only to the liver without the involvement of any other organ like spleen, bone marrow, lymph nodes, peripheral blood, or other tissues until at least six months after diagnosis [3]. It can occur at any age but is usually found in the fifth or sixth decade of life, with male/female ratio of 2-3:1 [4]. The pathogenesis of PHL is still unclear and it has been associated with EBV, HCV, HIV, or HTLV infections, liver cirrhosis, systemic lupus erythematosus, and immunosuppressive therapy [5]. However, our patient had none of the above conditions or risk factors for PHL. The clinical manifestations are atypical including abdominal pain, fever, weight loss, and night sweats.

Hepatomegaly is the most frequent finding. Liver function tests are usually abnormal with elevation of ALP and LDH. The latter is increased in 30–80% of the cases [5]. On the contrary, tumor markers are usually within normal ranges with aFP and CEA being normal in almost 100% of the patients. Elevated LDH, with normal aFP and CEA, constitutes a valuable biologic feature [6] but it was not evaluated in our case. Radiological features of PHL are usually nonspecific and the most common presentation on
Primary hepatic lymphoma is a rare malignancy, which is frequently misdiagnosed. However, it should be considered in cases of space occupying liver lesions with elevated LDH and normal AFP and CEA. If the index of suspicion for PHL is high, a liver biopsy should be obtained. PHL is chemosensitive and early aggressive combination of chemotherapy may result in sustained remission. The role of surgery has not yet been clarified but, in patients with localized disease, surgery followed by adjuvant chemotherapy should be considered to prevent recurrence.

Conflict of Interests

The authors declare that there is no conflict of interests regarding the publication of this paper.

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