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Primary Hepatic Extranodal Marginal Zone B-Cell Lymphoma of Mucosa-Associated Lymphoid Tissue at the Maradi Reference Hospital, Niger: A Case Report and Literature Review

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Abstract

Primary Hepatic Lymphoma (PHL) is a rare malignant tumor. We report a case of primary hepatic lymphoma diagnosed in a 55-year-old patient who presented with right hypochondrium pain. Clinical examination revealed isolated hepatomegaly. Abdominal ultrasound showed heterogeneous macronodular hepatomegaly. Histological and immunohistochemical examination of biopsy fragments from the hepatic nodules led to a diagnosis of marginal zone lymphoma of mucosa-associated lymphoid tissue (MALT). In summary, this was a primary hepatic non-Hodgkin lymphoma of the MALT type, classified as Ann Arbor stage IV AB.

Keywords

Hepatic Lymphoma, Primary, Maradi/Niger

1. Introduction

Non-Hodgkin lymphoma constitutes a heterogeneous group of malignant hema-

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tologic disorders characterized by abnormal clonal proliferation of T cells, B cells, or both. The majority of non-Hodgkin lymphomas in adults are of the B-cell type [1]. Primary hepatic lymphoma (PHL) is a rare malignant tumor, accounting for 0.016% of all non-Hodgkin lymphomas and 0.4% of extranodal lymphomas [2]. It is characterized by primary lymphomatous involvement of the liver—liver involvement without involvement of the spleen, bone marrow, or other lymphatic organs. They are of B-cell phenotype, large cell type, and are associated with a poor prognosis [3]. The most common site of MALT lymphoma is the stomach. Primary hepatic extranodal marginal zone lymphoma of MALT is classified as a type of non-gastric MALT lymphoma and is considered extremely rare [4].

2. Observation

Mr. H. M., a 55-year-old mason with no known medical history, non-alcoholic, non-smoker, and with no history of herbal medicine use, was admitted to the internal medicine department of the Référence Hospital of Maradi for right hypochondrium pain occurring in the context of asthenia and weight loss, without jaundice, fever, or other associated digestive or extra-digestive signs, all evolving for about a month before his admission. On physical examination, the patient was well oriented in time and space. He had a firm, irregular, painful hepatomegaly, with a liver span of 17 cm. There were no noted profuse night sweats, jaundice, collateral venous circulation, splenomegaly, ascites, spider angioma, digital clubbing, superficial lymphadenopathy, or bone pain. There was also no anemia, bleeding, or infectious syndrome. Otherwise, the clinical examination was unremarkable. From a morphological standpoint, the abdominal ultrasound showed heterogeneous hepatomegaly with solid and mixed nodules, including two large ones measuring 102 and 127 mm, mild ascites; absence of splenomegaly and deep adenopathies (**Figure 1**). An upper digestive endoscopy was performed, without evidence of a gastric, esophago-gastro-duodenal tumor or signs of portal hypertension. Complete colonoscopy and thoraco-abdomino-pelvic CT scan could not be performed due to financial limitation.

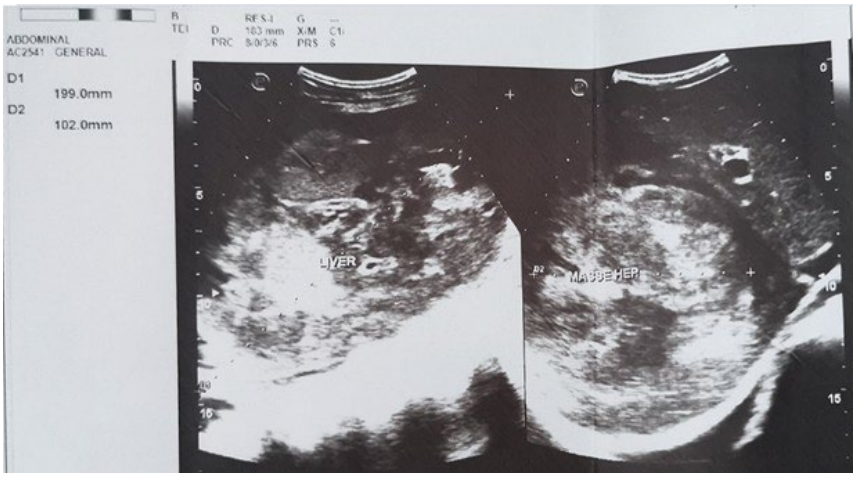

Figure 1. Hepatic mass on abdominal ultrasound.

Serologies for viral hepatitis B, hepatitis C, and HIV were negative. Transaminases, alpha-fetoprotein (AFP), lactate dehydrogenase (LDH), and complete blood count were normal.

As the incompleteness of the evaluation and the financial limitation, an ultrasound-guided liver biopsy was recommended and performed without incident on the 7th day of hospitalization. The results of the histological and immunohistochemical examinations (CD20-, CD19+, and CD5-) received 15 days after the biopsy suggested a mucosa-associated lymphoid tissue (MALT) marginal zone lymphoma (Figure 2).

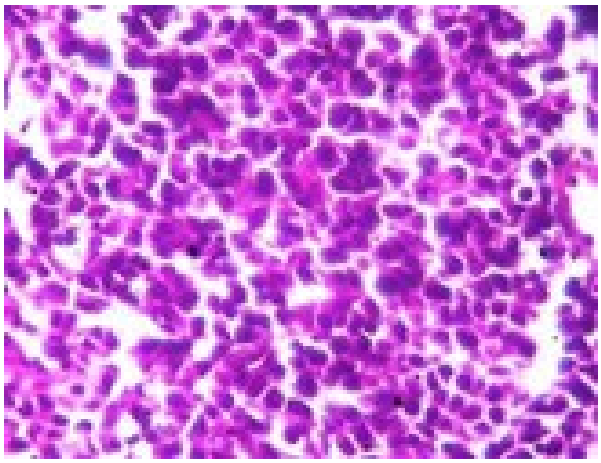

Figure 2. Histology of biopsy fragments from hepatic nodules showing diffuse infiltration by malignant lymphoid cells: Lymphoma.

Given the initial liver involvement, the absence of tumor syndrome, bone marrow failure syndrome, clinical signs suggesting involvement of other organs, or signs of clinical and biological progression; and despite the incomplete workup, we established the diagnosis of a hepatic marginal zone MALT-type lymphoma, classified as Ann Arbor IV AB IPI1.

An extension assessment including a clinical reevaluation of the patient, a thoracoabdomino-pelvic CT scan, a myelogram, a bone marrow biopsy, and a biological assessment of disease progression should have been performed; however, the patient died before the diagnosis.

A polychemotherapy combining cyclophosphamide, vincristine, and prednisone (CVP) or cyclophosphamide, Adriamycin, vincristine, and prednisone (CHOP) would allow obtaining a partial or even complete remission.

After 11 days of hospitalization, the patient was discharged on symptomatic treatment with second-level analgesics. He died at home a few days after his discharge from the hospital in an unspecified context, before the results of the histological examination.

3. Discussion

Non-Hodgkin lymphomas constitute a heterogeneous group of malignant prolifer-

ations of lymphoid cells of extramedullary origin. They present clinically with the development of tumors within lymphoid tissue, particularly lymph nodes [5]. Secondary involvement of the liver is common and occurs in more than 50% of cases of systemic non-Hodgkin lymphoma [6]. Primary lymphoma of the liver accounts for 0.4% of extranodal non-Hodgkin lymphomas and only 0.016% of all cases of non-Hodgkin lymphoma [7]. The etiology of primary lymphoma of the liver is unknown, but several risk factors have been described: behavioral risk factors (alcohol, tobacco, infection with EBV, HIV, hepatitis C and B, Helicobacter pylori, etc.); environmental factors (pesticides, solvents, wood dust, etc.); genetic factors; and certain autoimmune diseases have also been implicated: Sjögren's syndrome, Hashimoto's thyroiditis, rheumatoid arthritis, systemic lupus erythematosus, etc. [5] [6]. The diagnosis of primary lymphoma of the liver should be considered in any hepatic mass, guided by imaging, and confirmed by histopathological examination with immunohistochemical study of a specimen obtained by ultrasound-guided or laparoscopic liver biopsy, or even by exploratory laparotomy. The vast majority of primary hepatic lymphomas correspond to diffuse large B-cell lymphomas [8] [9]. Other histological types described (less than 5% of cases) include immunoblastic, lymphoblastic, Burkitt lymphomas, MALT (mucosa-associated lymphoid tissue) lymphomas, anaplastic large cell lymphomas, and rare cases of T-cell lymphomas [7]. The clinical case we report concerns primary hepatic extranodal marginal zone lymphoma of the MALT type. A similar case was described by Yuki Yamashita *et al.*, a primary hepatic extranodal marginal zone lymphoma of MALT associated with underlying hepatitis B virus (HBV) infection [4], and D. Chatelain *et al.*, who also reported a case of primary hepatic MALT lymphoma [10]. Several cases of primary hepatic lymphoma have been reported in the international literature. Authors such as Hicham Eddou *et al.* [2], R. Makhmari *et al.* [3], C. Nasr Ben Ammar *et al.* [11], and M. Mlika *et al.* [12] have reported a case of diffuse large B-cell non-Hodgkin lymphoma expressing CD20. B. Mouna *et al.* also reported a case of primary large B cell lymphoma with reactivity for CD20, CD5, CD45 and Bcl6 [13]. Our clinical case expands the number of the few cases reported in the international literature.

4. Conclusion

Primary hepatic extranodal marginal zone lymphoma of the MALT type is a rare condition. The diagnosis should be considered in any hepatic mass, especially when the alpha-fetoprotein level is normal. The diagnosis is confirmed by histological and immunohistochemical examination of the liver biopsy specimen. This clinical case has limitations due to the non-exhaustive nature of the assessment. Nevertheless, based on the clinical data and the assessment performed, this diagnosis seems very likely.

Conflicts of Interest

The authors declare no conflict of interest in relation to this article.

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