Splenic Marginal Zone Lymphoma Associated With Hepatitis B Virus Infection: A Case Report

J Mathew, I Aldean

Citation

J Mathew, I Aldean. *Splenic Marginal Zone Lymphoma Associated With Hepatitis B Virus Infection: A Case Report*. The Internet Journal of Surgery. 2002 Volume 5 Number 1.

Abstract

Hepatitis B virus infection has been implicated in the development of hepatocellular and haematopoietic malignancies. We present a case of splenic marginal zone lymphoma associated with hepatitis B virus infection and discuss its possible role in the aetiology of such lymphomas.

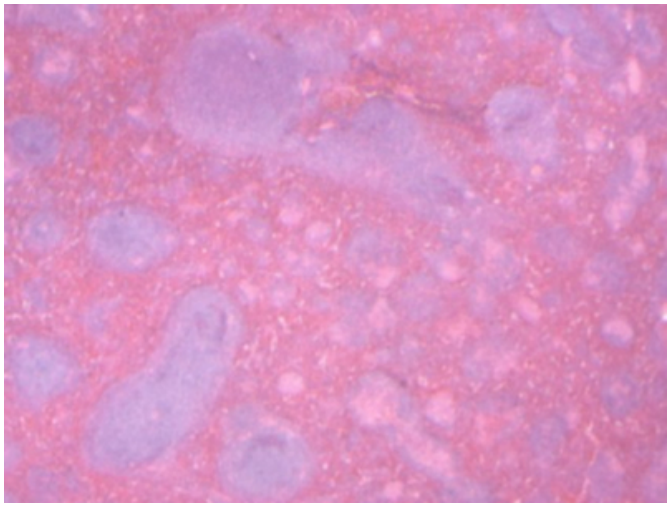
CASE REPORT

A 67-year-old man was seen in clinic with a two-year history of tiredness and night sweats. Of late, he had been complaining of abdominal discomfort and a swelling involving the left side of the abdomen. He had lost one and a half stone in weight over the past two years. He has been under the care of physicians for chronic liver disease caused with Hepatitis B Virus infection diagnosed 15 years back. Clinically, he had a very large spleen extending almost up to his umbilicus. Ultrasound scan of the abdomen confirmed splenomegally with a maximum oblique diameter of 25cm. He was mildly anaemic with haemoglobin of 10.1 gm/dl, platelets of $54x10^{9}$ /L, WCC of $6x\ 10^{9}$ /L, neutrophil count of $1.9x10^{9}$ /L and a lymphocyte count of $1.2x\ 10^{9}$ /L. Liver function tests revealed mildly raised AST of 146 IU/L and ALT of 124 IU/L (rest were within normal limits). Peripheral smear showed few hairy cells. Bone marrow aspiration revealed normal haemopoesis but there was infiltration with medium sized lymphoid cells. Trephine biopsy showed infiltration with small lymphoid cells. A provisional diagnosis of primary splenic lymphoma was made.

Pneumovax injections were given two weeks prior to explorative laparotomy and splenectomy specimen was sent for histopathological assessment. The spleen weighed 3.2 kg and measured 260x 185x 80mm. On microscopy the sections of the spleen showed nodules with clusters of macrophages at the centre (Fig 1). Surrounding this central zone there were neoplastic cells that have biphasic appearance and contains intermediate sized cells with more abundant cytoplasm. The neoplastic cells stained with antibodies to CD20, CD79a and CD43.

Figure 1

Figure 1: Low power view of splenic white pulp with marginal zone expansion (X40)



The patient received post operatively single agent chemotherapy using Fluderabine. After 6 months follow up, his symptoms of tiredness, night sweat and abdominal discomfort have significantly improved.

DISCUSSION

Secondary involvement of the spleen from various nodal lymphomas is common. Primary malignant lymphomas of the spleen are very rare and represent only 1-2% of all malignant lymphomas involving the spleen ([1]). Splenic marginal zone lymphoma (SMZL) is a recently described primary splenic lymphoproliferative disorder that mainly affects older men ([2]).

The etiology of SMZL is less well understood. Hepatitis B virus is both hapatotropic and lymphotropic, and has been implicated in the development of hepatocellular and hematopoietic malignancies. The mechanism by which the malignancy develops after the HBV infection is unclear although it could be similar to its involvement in the pathogenesis of hepatocellular malignancy. Primary hepatic lymphoma has been identified at autopsy in a patient with hepatocellular carcinoma associated with HBV infection ([3]). HBV gene products have been detected in the endothelial cells of tumour tissues of leukaemia and lymphoma patients and it is probable that HBV-infected endothelial cells support and stimulate the tumour growth by producing cytokines ([4]).

Splenectomy is the treatment of choice in patients with SMZL, even in those with blood and bone marrow involvement ([5,6]). Chemotherapy has a role in the management following splenectomy, especially in those patients with progressive disease ([5]). The prognosis seems to be very good and there have been series where the 5-year survival rate is approximately eighty percent ([7,8]).

LEARNING POINTS

Primary splenic lymphoma could be considered as a differential diagnosis of extrahepatic disorders in patients with chronic hepatitis B virus infection.

CORRESPONDENCE TO

Mr John Mathew 8 Fern House Amberley Drive Wythenshawe Manchester M23 2RW E-mail: mathewjohn@aol.com

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Author Information

John Mathew, FRCS
Trafford General Hospital

Ismail M. Aldean, FRCS
Trafford General Hospital