

SPLENIC LYMPHOMA WITH HYPERSPLENISM - A CASE REPORT

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ABSTRACT

A 47-year old man had hypersplenism from massive splenomegaly, the cause of which was undetermined for 2 years. He was initially asymptomatic though there was mild pancytopenia. However, 18 months after presentation he manifested both clinical and haematological deterioration, almost succumbing to sepsis. Splenectomy finally provided a definite diagnosis of follicular lymphoma and also restored his blood counts to within normal range.

Keywords: Massive undiagnosed splenomegaly, follicular lymphoma.

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INTRODUCTION

Splenomegaly is associated with a great variety of diseases, most of which can be diagnosed after clinical and some laboratory evaluations. However, in rare instances, the cause of splenomegaly is not apparent until splenectomy is performed. We report a patient whereby the cause of massive splenomegaly was established 2 years after presentation when splenectomy had to be done for deteriorating hypersplenism.

CASE REPORT

I M was a 47-year old manual worker who was referred to the University Hospital, Kuala Lumpur for investigation of massive splenomegaly. Except for epigastric discomfort, he was otherwise asymptomatic. He had no loss of appetite nor weight loss. There was no history of arthritis, jaundice or malaria previously. He did not drink or smoke. On examination, his general condition was good, although he was slightly pale. There was no jaundice or pedal edema nor other stigmata of chronic liver disease. The spleen was massively enlarged and extended right down to the right iliac fossa (vertical span of 18 cm). It

was firm, non-tender with smooth surface and no bruit was heard over it. The liver was not palpable. His lymph nodes were not significantly enlarged. No other abnormalities were detected in other systems.

His blood count revealed mild pancytopenia with a haemoglobin concentration of 11.7g/dl, white cell count of $3 \times 10^9/L$ and a platelet count of $75 \times 10^9/L$. The peripheral blood film showed normochromic normocytic red cells. There were no poikilocytes and no evidence of a leucoerythroblastic picture. The liver function tests were normal. Hepatitis-B surface antigen was not detected. Bone marrow aspiration showed erythroid hyperplasia and focal lymphoid nodules which were interpreted as benign lymphoid aggregates then. Barium swallow study was normal, with no evidence of oesophageal varices. Antibody to Plasmodium falciparum was positive (titre of 1:64). A provisional diagnosis of Tropical Splenomegaly Syndrome (TSS) was made and he was given a trial of proguanil 100mg daily for 1 year. Serum Immunoelectrophoresis did not show any monoclonal protein and Immunoglobulin levels were within normal limits. As expected then, he did not show any clinical or haematological response to the treatment. However, 18 months after presentation, his haemoglobin started to drop rather sharply and splenectomy was advised.

He defaulted but had to be admitted in December 1985 because of bronchopneumonia as well as congestive heart failure secondary to severe anemia. He recovered with treatment and subsequently agreed to splenectomy. Bone marrow aspirate and trephine biopsy performed pre-operatively showed a cellular marrow and again lymphoid nodules were seen. Splenectomy was performed on 16 January 1986 following which he made an excellent recovery (Fig 1). No enlarged abdominal nodes were noted intraoperatively and the liver appeared normal. The spleen was markedly enlarged weighing 3600 gm (Fig 2). The capsule was smooth. On sectioning, several small whitish nodules were seen throughout the spleen. Histology of these nodules (Fig 3) revealed small lymphoid cells with cleaved nuclei and scanty cytoplasm. In addition, there was characteristic subendothelial collection of lymphoid cells. The pathologic diagnosis was low grade follicular malignant lymphoma, predominantly small cleaved cell type (Working Formulation) (1). His blood count returned to normal range at the fourth post-operative day. He remained well on last follow-up a year after operation.

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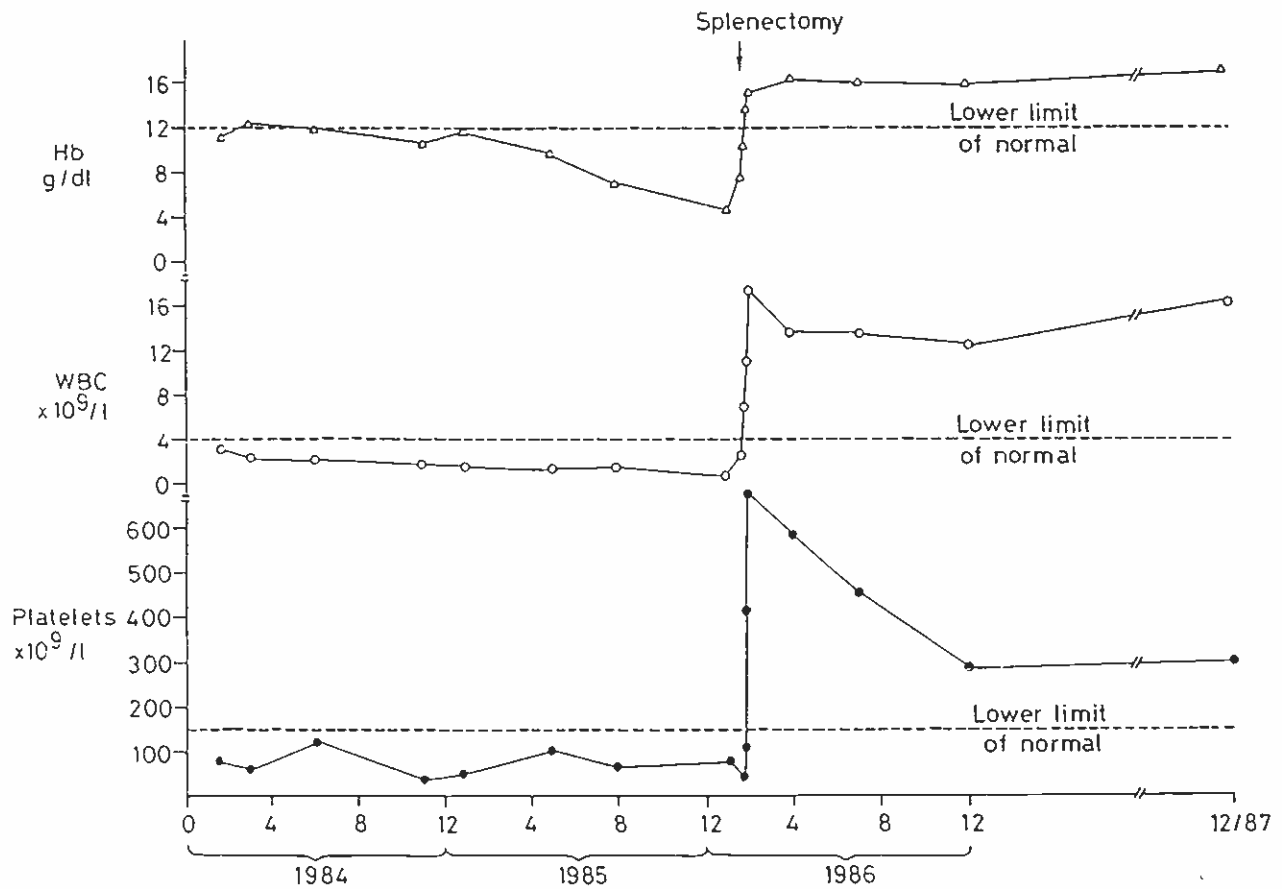


Fig 1 Clinical course of patient

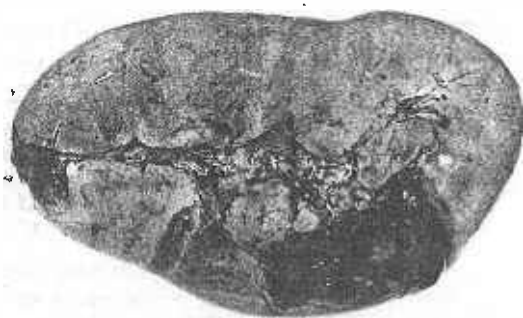


Fig 2 Low power view of spleen shows many neoplastic follicles (Hematoxylin-eosin, x 40)

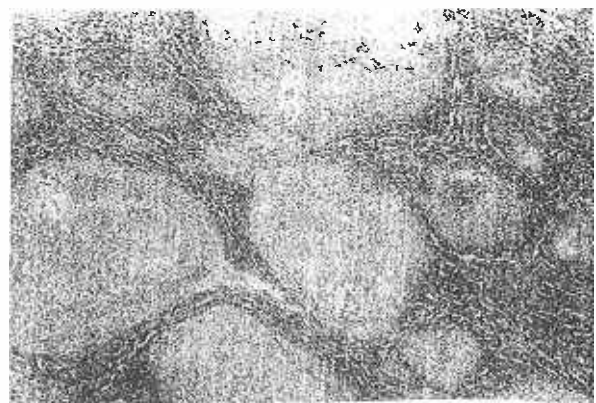


Fig 3 Enlarged Spleen

DISCUSSION

Although the spleen is clinically enlarged in at least 50% of patients with generalised lymphoma (2) and the incidence would probably be greater if all cases were subjected to post-mortem examination, primary involvement of the spleen by lymphoma is extremely rare. It has been estimated that 1% of all lymphoma present with splenomegaly as the initial finding, without evidence of lymphoma in peripheral sites (3).

Splenectomy was performed in this patient for both diagnostic and therapeutic grounds. He exhibited features of hypersplenism namely cytopenia, splenomegaly, hyperplasia of bone marrow and correction of cytopenia by splenectomy - the "Sine qua non" in the diagnosis of hypersplenism. The discovery of focal areas of lymphomatous infiltration in bone marrow is not a contraindication to splenectomy.

Gil et al (4) noted reversal of cytopenia after splenectomy in all the 19 patients with Non-Hodgkin's Lymphoma

who had marrow involvement by lymphoma. Skarin et al (5) reviewed the clinical and pathological findings of 11 patients in whom the diagnosis of lymphoma had been made at the time of splenectomy for undiagnosed splenomegaly. Histologically they were considered to be infiltrated by nodular or diffuse lymphocytic or lymphoblastic lymphoma. The histological typing of lymphoma is important as the patients in Skarin's series had relatively good prognosis and some patients survived a long time eg. 23 and 27 years. In contrast, Harris et al(6) reported 10 patients with diffuse large cell lymphoma of the spleen who generally had a poor prognosis. Interestingly, pa-

tients with idiopathic splenomegaly in whom splenectomy revealed no definitive diagnosis, had increased incidence of lymphoma (7, 8). Hence, there is a spectrum of lymphoid hyperplasia affecting the spleen predominantly and it ranges from diffuse large cell lymphoma to lymphocytic lymphoma and patients with a similar clinical syndrome but in whom the neoplastic nature of the lymphoid hyperplasia is not apparent.

This patient needs to be followed up as 6 out of the 11 patients in Skarin's series developed the blood picture of chronic lymphocytic leukaemia at a later stage.

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