CASE REPORT

SPLEENIC LYMPHOMA WITH VILLOUS LYMPHOCYTES: CASE REPORT AND LITERATURE REVIEW.

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ABSTRACT

BACKGROUND: *Splenic lymphoma with villous lymphocytes (SLVL) is a rare but recognized distinct disease entity among chronic B lymphoproliferative disorders. It is frequently misdiagnosed as chronic lymphocytic leukaemia, (CLL) Prolymphocytic leukaemia or Hairy Cell leukaemia. Few cases have been reported worldwide.*

METHOD: *The case records of a sixty year old Nigerian male with a splenic lymphoma and a review of the literature on the subject using MEDLINE, other internet sources and manual library search was utilized.*

RESULT: *A sixty-year-old male with splenic lymphoma with villous lymphocytes, seen at the Ahmadu Bello University Teaching Hospital Zaria is presented. However review of the blood film, bone marrow and Splenic aspirates showed absolute lymphocytosis, consisting of "villous" lymphocytes. He was commenced on an alkylating agent and a glucocorticoid, with partial remission in the first three months but was lost to follow up.*

CONCLUSION*: Adequate morphologic evaluation is advocated particularly in the resource limited settings were Cytogenetics, immunohistochemistry and immunophenotyping are not available.*

Key Words: SLVL; CLL; Peripheral Blood/bone Marrow Lymhocytosis; Rare Presentation.

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INTRODUCTION

Splenic lymphoma with villous lymphocytes (SLVL) is a benign or indolent non-Hodgkin's lymphoma with a distinct clinical, morphological and immunophenotypic pattern. It is a disease of the elderly between 57 to 88 years of age and may produce anaemia, thrombocytopenia and lymphocytosis with massive splenomegaly.[1,-3] In 1987 Melo et al[3] reported a series of 22 patients while Matutes et al[3] in 1994 describes the immunophenotypic characteristics of 100 cases.[3] However no literature in Nigeria has been documented.

There is minimal involvement of the liver and lymph nodes. Blood and bone marrow are often involved. Villous lymphocytes are found in the peripheral blood and bone marrow.[1, 2, 3] This lymphoma has been previously described as "splenomegalic immunocytoma with circulating hairy cells", "malignant lymphoma" and "chronic lymphoproliferative disorder reticuloendotheliosis".[3,4,5]

There may be co infection with Hepatitis C virus.[6, 7] Bone marrow examination is an important diagnostic tool as it provides distinction from other lymphoproliferative disorders. Spleen histology however shows a variable white and red pulp involvement, with some evidence of lymphoplasmacytic differentiation.[3, 8, 9] There may be elevated levels of immunoglobulin in the serum or urine but often not greater than 30g/dl.[6, 7] Cytogenetics and Membrane marker (immunophenotyping) studies are additional diagnostic tool.[3, 10]

No therapy may be required for a long time in a minority of patients. Splenectomy is the treatment of choice, provided there are no clinical contraindications to surgery. Chlorambucil with glucocorticoid may be effective.[3] Fludarabine has been effective, partial responses have been reported with 2 Deoxycoformycin (DCF).[3, 6] Interferon is effective in the presence of Hepatitis C virus co-infection.[7] Transformation to a large cell lymphoma may occur and respond to CHOP.[3,]

In this report we describe a case of SLVL, elucidating the diagnostic and management constraints in our resource limited setting.

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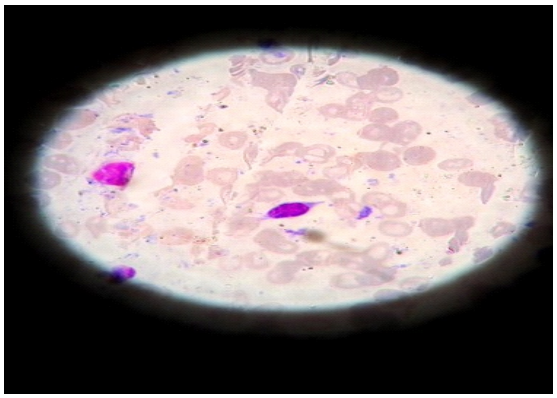
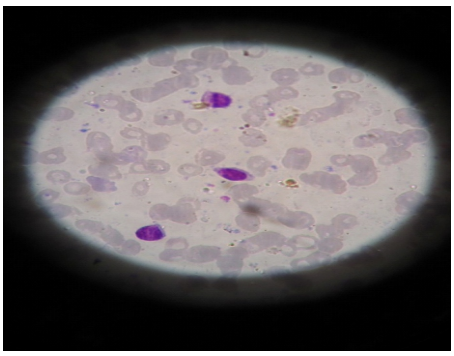
CASE REPORT

A 60-year-old male Nigerian farmer was seen at Ahmadu Bello University Teaching Hospital (ABUTH) Zaria. He presented with a 3-year history of progressive left sided abdominal swelling associated with weight loss. Systemic review revealed a poor appetite, no bone pains, no fever, no drenching night sweats and no bleeding from any visible external orifices. Urinary and bowel habits were normal. He had 1 pint of packed cell transfusion about 1 year prior to presentation in a peripheral hospital.

Physical examination revealed a chronically ill elderly man, not in any distress, afebrile, moderately pale, anicteric, acyanosed, no finger clubbing well hydrated but with palpable discrete peripheral bilateral inguinal lymphadenopathy. There was a massive splenomegaly extending towards the left iliac fossa and measured about 28 cm. The liver was also palpably enlarged by about 4 cm below the right costal margin at the midclavicular line.

Laboratory data were as follows: Packed cell volume 28%, total white cell count 56.4 x 10^9/l, platelet count of 120 x 10^9/l and a dimorphic red cells with 97% lymphocytes on blood film. Stool microscopy, serum IgM, Bence Jones proteins, hepatitis C virus (HCV serology), Serum electrolytes, urea and creatinine estimation and liver function results were unremarkable. However close examination of the blood film showed that the lymphocytes were "villous". Bone marrow histology revealed a hypercellular marrow and confirmed lymphocytosis of about 30%, consisting of lymphocytic infiltrates interspersed between other haemopoietic precursor cells; which were of intermediate size with high nuclear cytoplasmic ratio, irregular cytoplasmic extensions at one pole "villous" an oval nucleus and a dense nuclear chromatin pattern with an occasional nucleoli. Similar picture was observed following a splenic aspiration. Cytogenetics and immunohistochemistry were not done due to unavailability of such diagnostic tool.





He was commenced on low dose continuous oral Chlorambucil 6mg daily and oral prednisolone 20mg three times daily for 7 days. In addition he had other supportive therapies including blood transfusion. Partial response was achieved in the first three months as was evident by reduction of the splenic size to 18 cm and normalization of the peripheral blood lymphocytes count. However he was lost to follow-up.

DISCUSSION

SLVL is a slowly progressive clonal lymphoid disorder, in which massive splenomegaly posses a significant problem. The occasional patient may be diagnosed by chance on investigation for another complaint. Massive splenomegaly causes cytopenias particularly anaemia occasionally with thrombocytopenia, enlarged plasma volume, infections, portal hypertension and severe abdominal discomfort. This leads to increase in transfusion demand.

No treatment may be required for prolonged periods of time in a minority of patients. When symptoms make treatment necessary, measures directed against the spleen seem to be beneficial. In this report there was a massive splenomegaly, which was causing anaemia and abdominal discomfort. This calls for measures against the spleen using the available chemotherapy such as Chlorambucil, although it is said to be ineffective in several patients and prednisolone was therefore added. Fludarabine and DCF, which are a better choice, are not available. Mulligan et al. 1991 stated that splenectomy is the treatment of choice provided there are no clinical indications for surgery.[3] These was not feasible in our case due to socioeconomic factors.

Finally these treatment modalities are palliative and therefore therapy is for life. Cure is only achieved by stem cell transplantation, which is still far from our reach.

CONCLUSION

Adequate morphologic evaluation of patients presenting with lymphocytosis and splenomegaly, is mandatory even in the absence of Cytogenetics, immunophenotyping and histochemistry in our locality to prevent diagnostic pitfalls.

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