

Peripheral Lymphocytosis In Tropical Splenomegaly Syndrome

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INTRODUCTION

Splenomegaly (a spleen greater than 10 cm) is a common finding in patients in Africa and can be caused by many diseases (See Table). In Malawi, schistosomiasis due to *S. mansoni*, and the Tropical Splenomegaly Syndrome (TSS) are believed to be the major causes¹.

Table: Some common causes of massive splenomegaly in Africa.

TROPICAL SPLENO-MEGALY SYNDROME
HEPATIC CIRRHOSIS
HEPATO-SPLENIC SCHISTOSOMIASIS
VISCERAL LEISHMANIASIS
CHRONIC GRANULOCYTTIC LEUKAEMIA
CHRONIC LYMPHOCYTTIC LEUKAEMIA
MYELOFIBROSIS
LYMPHOMA
HAEMATOLOGICAL DISEASES, eg THALASSAEMIA

TSS results from an abnormal immunological response to malaria in certain genetically predisposed individual^{2,3}. Diagnosis is largely by exclusion of other known causes of splenomegaly. In the majority of patients with TSS a massive spleen, often accompanied with some degree of hepatomegaly, is the only dominant clinical feature.

A number of laboratory investigations (high malarial antibody titres and high serum levels of IgM) are helpful in confirming the diagnosis. Unfortunately these are generally unavailable in Malawi. A blood film does not contribute towards the diagnosis because it can be either positive or negative. However, a full blood count and white blood cell differentiation are often available, and these may show certain abnormalities which clinicians should be aware of. The following case history illustrates this point.

Case Report

A 60-year old woman presented in the outpatients department with a three month complaint of fullness in the abdomen after meals. She had no other complaints; more specifically she had not experienced any weight loss or fever. On examination, she was an elderly woman in good condition, moderately pale and not jaundiced. There was no lymphadenopathy. On abdominal examination a massively enlarged spleen was found (Hackett grade 5. i.e. reaching beyond the level of the iliac crests); the liver was not palpable and no ascites could be detected.

Laboratory investigations showed:- Haemoglobin 6.8 g/dl;

Total white cell count 52,800/m³, of which 94% were lymphocytes; red cell morphology normocytic and normochromic; platelet estimation normal.

The differential diagnosis included TSS or chronic lymphocytic leukaemia (CLL). There was nothing in the clinical evaluation to suggest CLL, and she was therefore treated for TSS with antimalarial chemoprophylaxis (chloroquine 300 mg base once a week).

On review 3 months later, she reported significant subjective improvement. On examination the size of her spleen had remarkably decreased to Hackett grade 3

(umbilical level). The total white cell count had decreased to 50,300/m³, of which 98% were lymphocytes. It was explained to her that treatment with antimalarial chemoprophylaxis must continue for life.

DISCUSSION

It is not common knowledge that TSS can be associated with marked peripheral blood lymphocytosis, and given this laboratory picture many patients will be diagnosed as having chronic lymphocytic leukaemia CLL. The usual haematological finding in TSS is pancytopenia, resulting from hypersplenism. However, in Nigeria it is not uncommon to see patients with TSS who have high peripheral blood lymphocytosis², and surveys done in Africa show that as many as 15% of patients have a lymphocytosis within a range of 4.5 - 100.0 x 10⁹/l³.

CLL is the commonest leukaemia in Africa as well as in the industrialised world. Lymphocytosis, splenomegaly and lymphadenopathy are characteristic features, although the latter may be absent. The total blood lymphocyte count in CLL is often higher than 100.0 x 10⁹/l, whereas in TSS it is generally lower. Bone marrow aspiration may not allow differentiation of the two conditions, as both may show excessive lymphocytic infiltration. Liver histology can be diagnostic by showing the characteristic infiltration in hepatic sinusoids of TSS, but generalised lymphocytic infiltration occurs in both conditions. More conclusive techniques to differentiate between the two diseases include testing whether the lymphocytes rosette with mouse red cells and whether the lymphocytes are mainly B cells with weak staining monoclonal surface immunoglobulin, both characteristic of CLL. In addition to this, determination of serum IgM concentrations could be useful^{2,3}, being characteristically elevated in TSS. None of these sophisticated investigations are readily available to clinicians in Malawi, so in some patients who present with splenomegaly and lymphocytosis, it may be very difficult to distinguish TSS from CLL. Patients with TSS respond to antimalarial chemoprophylaxis, although some patients with CLL may show a partial and transient response as well⁴.

If in doubt about the diagnosis, I suggest that such patients be treated with antimalarial chemoprophylaxis. In cases of TSS, this will greatly reduce morbidity and mortality and will eventually lead to complete regression of splenomegaly, which confirms the diagnosis. The patient feels better after a few weeks of treatment, and the spleen decreases in size and becomes impalpable in 1 or 2 years. The lymphocytosis, however, may persist³. Treatment must be continued for life.

REFERENCES

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