

# Lympho-epithelioid Cellular Lymphoma (Lennert's Lymphoma)

## A Case Report

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### SUMMARY

A patient with Lennert's lymphoma (malignant lymphoma with a high content of epithelioid histiocytes) is described. The case is unique in that hepatosplenomegaly was the presenting feature, the diagnosis was made on bone marrow biopsy and the disorder appears to have run a benign course.

*S. Afr. med. J.*, 51, 555 (1977).

Lympho-epithelioid cellular lymphoma was originally described by Lennert,<sup>1</sup> who considered it a variant of Hodgkin's disease, but has since categorized it as a non-Hodgkin's lymphoma.<sup>2</sup> Burke and Butler<sup>3</sup> have recently described 15 patients seen at the M.D. Anderson Hospital. In this article we describe another case with some unusual features.

### CASE REPORT

The patient, a 69-year-old White woman, first presented in 1961 with a urinary tract infection and nausea, and was found to have a liver palpable 3 cm beneath the right costal margin, and a soft spleen, palpable 3 cm below the left costal margin. There was no past history of note or other significant findings. Her blood picture was normal and a bone marrow aspirate revealed no abnormality. No cause for the hepatosplenomegaly was found and she was discharged for follow-up. Examination of a bone marrow aspirate 1 year later again revealed no abnormality. The moderately enlarged spleen remained palpable and in late 1973 she was again noted to have hepatosplenomegaly, which was confirmed by <sup>99</sup>Tc scan. The length of the spleen, as determined from the scan, was 25 cm. The uptake of <sup>99</sup>Tc in the liver was mildly irregular but no filling defects were present.

In January 1974 she was seen in the Haematology Department for the first time because of hepatosplenomegaly with anaemia and a low platelet count. A low serum folate concentration (1,7 ng/ml) was found and oral replacement therapy was commenced. A bone marrow aspirate again showed no abnormality and red cell survival was minimally shortened at 25,2 days (normal 26-30 days) but no meaningful sequestration by the spleen could be

demonstrated. Results of liver and renal function studies were within normal limits.

The patient was admitted in September 1976 because of symptoms of anaemia and discomfort due to massive splenomegaly. There was no lymphadenopathy, and no tonsillar enlargement. The haemoglobin concentration was 4 g/100 ml, the platelet count was 15 000/ $\mu$ l and the white cell count varied between 2 400/ $\mu$ l and 4 900/ $\mu$ l with a leuco-erythroblastic blood picture. The serum copper was 32,3  $\mu$ mol/l (normal 10-26  $\mu$ mol/l), and serum chemistry showed no abnormalities. Total serum protein and serum globulin concentrations were at the lower limits of normal. At this time a clinical diagnosis of myeloproliferative syndrome was made and a splenectomy was carried out. A liver biopsy, bone marrow aspirate, and trephine biopsy were also performed. Postoperatively her haemoglobin, white cell, and platelet values rose to normal levels, and a temporary thrombocytosis followed splenectomy. Further recovery was uneventful.

### Histological Features

The bone marrow displayed scattered foci of bland epithelioid histiocytes with vesicular nuclei, small nucleoli and abundant acidophilic granular cytoplasm (Fig. 1). In and around the clusters of epithelioid histiocytes were lymphocytes and in some foci there were eosinophils in a peripheral location. The lymphocytes showed slight nuclear irregularities and were rounded, oval, or elongated. An

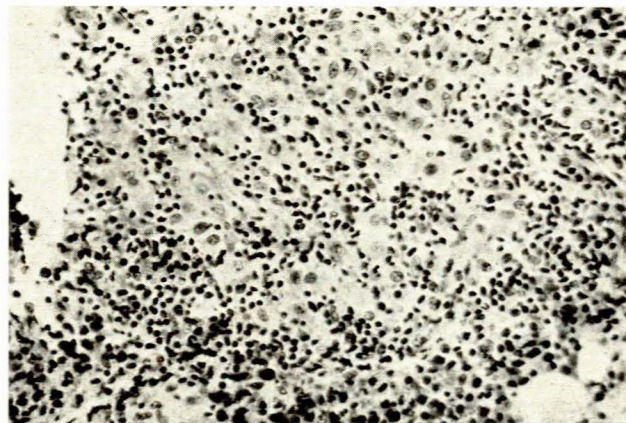


Fig. 1. A cluster of bland epithelioid histiocytes. Scattered among them are atypical lymphocytes. At the lower border of the illustration are peripherally located eosinophils (H and E  $\times$  400).

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Date received: 29 October 1976.

occasional immunoblast was seen but they were not a feature of the infiltrate. Reed-Sternberg-like cells were not seen.

The spleen weighed 1200 g. Histologically it showed diffuse involvement of the white pulp and peri-arteriolar lymphoid sheaths with foci identical to those seen in the bone marrow. The liver showed portal and sinusoidal infiltration with atypical lymphocytes, but epithelioid histiocytes were not as prominent as in the bone marrow or spleen.

### DISCUSSION

The histopathological features in this patient were identical to those originally described by Lennert<sup>1</sup> as 'epithelioid type of lymphogranulomatosis' and more recently as 'lympho-epithelioid cellular lymphoma'.<sup>2</sup> Burke and Butler<sup>3</sup> recently documented 15 cases seen at the M.D. Anderson Hospital and coded as 'malignant lymphoma, Lennert's type'. Our patient, however, displayed some unusual features not observed in Burke and Butler's series. There was long-standing hepatosplenomegaly with no lymphadenopathy, whereas all their patients presented with lymphadenopathy. Tonsillar tissue was not involved in our patient, although this was relatively commonly involved in Burke and Butler's series. Moreover, they had only 1 patient with splenomegaly and none with hepatomegaly, whereas 3 of our 4 patients who underwent a staging laparotomy had splenic involvement. Although in our series only 1 patient had bone marrow involvement, the patient's condition was first diagnosed on bone marrow biopsy. Subsequent splenectomy and liver biopsy confirmed hepatic and splenic involvement.

Apart from abdominal discomfort due to an enlarged spleen our patient had neither systemic symptoms nor a polyclonal gammopathy. In fact, the total protein and  $\gamma$ -globulin concentrations were at the lower end of the normal range. She has a long history, and it may be that in some of the reported cases the condition is relatively benign, since the follow-up of these patients to date has

been short. Burke and Butler described 1 patient who was in stage IIIB and who has remained well for 18 months with no therapy. Dorfman and Warnke<sup>4</sup> described a patient with stage IV disease who has remained asymptomatic without treatment for 2 years.

Rappaport described Hodgkin's granuloma with a focal histiocytic reaction.<sup>5</sup> This has a superficial resemblance to Lennert's lymphoma in that clusters of large pale histiocytes are prominent throughout the affected tissue. The histiocytes are bland, with abundant pink-stained cytoplasm and ovoid vesicular nuclei. Reed-Sternberg cells are present but are difficult to find in this type of histiocytic reaction. It is interesting that the first patients were all thought to be suffering from Hodgkin's disease and were in fact treated as such with the MOPP regimen. In the tissue sections of our patient there were no Reed-Sternberg or Reed-Sternberg-like cells nor was the cellular environment compatible with Hodgkin's disease.

The fact that there was no lymphadenopathy is unusual, but we have seen cases of lymphoma with hepatosplenomegaly and bone marrow involvement with no lymphadenopathy. These patients are usually elderly and are treated conservatively. The long interval between the documentation of hepatosplenomegaly and eventual presentation with anaemia, thrombocytopenia and a leucoerythroblastic peripheral blood picture can only be ascribed to the benign nature of the neoplasm. This may be analogous to follicular lymphoma which often has a prolonged asymptomatic course.

We wish to thank Prof. H. P. Wassermann for allowing us to study his patient.

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