

UNUSUAL PRESENTATION OF LYMPHOMA

(A Case Report)

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Summary

A case of diffuse non-Hodgkin's lymphoma (DNHL) of poorly differentiated lymphocytic type (Stage IV) in a 63 year old African male with obstructive cardiomyopathy, massive left sided renomegaly and ipsilateral malignant retroperitoneal fibrosis is reported. An important feature was the absence of superficial lymphadenopathy.

Introduction

The lymphomatous spread is known to involve among others, the heart (Roberts et al 1968) kidney (Kielly et al 1969) and retroperitoneal space (Nitz et al 1970). The dominant clinical symptoms are usually related to the organ most severely involved. Following is the report of a case of DNHL who presented with a symptom complex related to the metastatic involvement of heart, kidney and retroperitoneal space without peripheral lymphadenopathy. Such clinical presentation, as far as we are aware, has not been previously reported.

Case Report

A 63 year old African male was admitted to a Medical Ward of the University Teaching Hospital, Lusaka, Zambia, with a four month history of left sided abdominal pain, progressive dyspnoea and weight loss. On physical examination the patient was moderately dyspnoic but without any cyanosis. There were bilateral pedal edema, clubbing of all fingers of both hands and a raised J.V.P. The pulse was 100/minute and irregular while BP was 150/100 mm Hg. There was no peripheral lymphadenopathy. Apex beat was located in the 5th intercostal space just lateral to the midclavicular line with faintly audible but normal 1st and 2nd heart sounds. There was clinical evidence of left sided pleural effusion. The liver was tender and enlarged to 6 cms below the right costal margin with a smooth surface. The left subcostal space

contained a ballotable and tender mass which extended to 14 cms below the costal margin. Investigations included HB 12.8 gms%, total nucleated count 5600/mm³ with a normal differential count, platelets 152000/mm³ and ESR 105 at the end of 1st hour; ECG suggested atrial fibrillation. A chest X-Ray confirmed left pleural effusion and also showed enlargement of the right heart (atrium and ventricle). A straight X-Ray of the abdomen suggested the presence of a soft tissue mass in left upper abdomen which also contained multiple gas containing loculi. An intravenous pyelogram demonstrated a normal right kidney (Fig. 1); the left kidney, however, remained non-visualised at 24 hours. Further investigations which included biopsy of the liver, left pleura and cytology of pleural effusion, were non contributory. The patient refused to undergo any surgical procedure, hence his management was essentially symptomatic which also included digitalisation. He died shortly afterwards.

Autopsy Findings

At autopsy, there was 700 ml of left sided pleural effusion. Pericardium was thick and adherent to the heart (830gms). The right anterolateral surface of the heart was pale white and unusually firm. On cut section this was found to be due to a diffusely infiltrating tumour. The tumour had involved the right atrial and ventricular myocardium (Fig. 2); the corresponding chambers were markedly narrowed. The left ventricle also showed tumour infiltration of the apical portion; thickness of the myocardium (after stripping the pericardium) below the mitral valve was 1.8 cms. The left atrium and mitral and aortic valves were uninvolved. Major branches of the coronary arteries were patent.

An enlarged spleen (300 gms) and a loop of the ileum were adherent to a massively enlarged left kidney (1500 gms) and thus, resulted in the formation of the left subcostal mass. The

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left kidney and proximal ureter were encased in thick fibrous tissue. The para-aortic, mesenteric and right perirenal lymph nodes were variably enlarged with fish flesh appearance on their cut surfaces. There was no lymphadenopathy in the mediastinum, axillae and in the neck. Rest of the organs showed congestion which was most marked in the liver (1600 gms, with nutmeg appearance) and spleen.

Histology

Sections from the enlarged abdominal lymphnodes revealed histologic features of DNHL of poorly differentiated lymphocytic type. There was extensive infiltration of the heart (fig. 3) and the left kidney (fig. 4) with malignant lymphoid cells; necrotic areas in the kidney were of ischemic nature. Histology of the retroperitoneal tissue showed similar infiltrations with fibrocellular proliferation. These malignant cells were also noted in the sections from the liver, spleen and bone marrow.

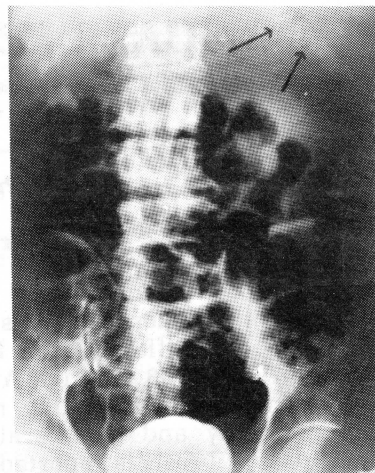


Figure I

Intravenous Urogram showing normal right kidney and nonvisualisation of left kidney. Arrows pointing to the gas containing loculi overlying a soft tissue mass.

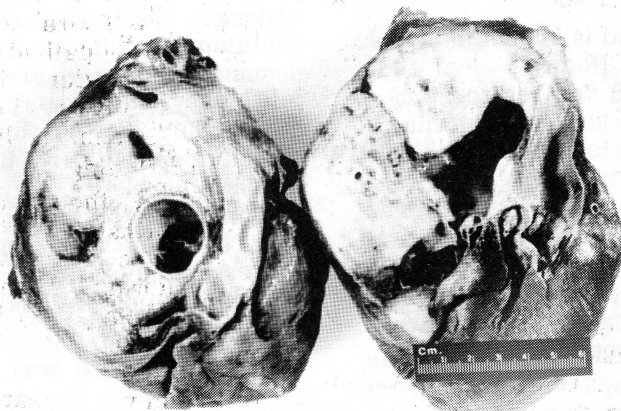


Figure II Extensive infiltration of the myocardium with malignant lymphomatous tissue.

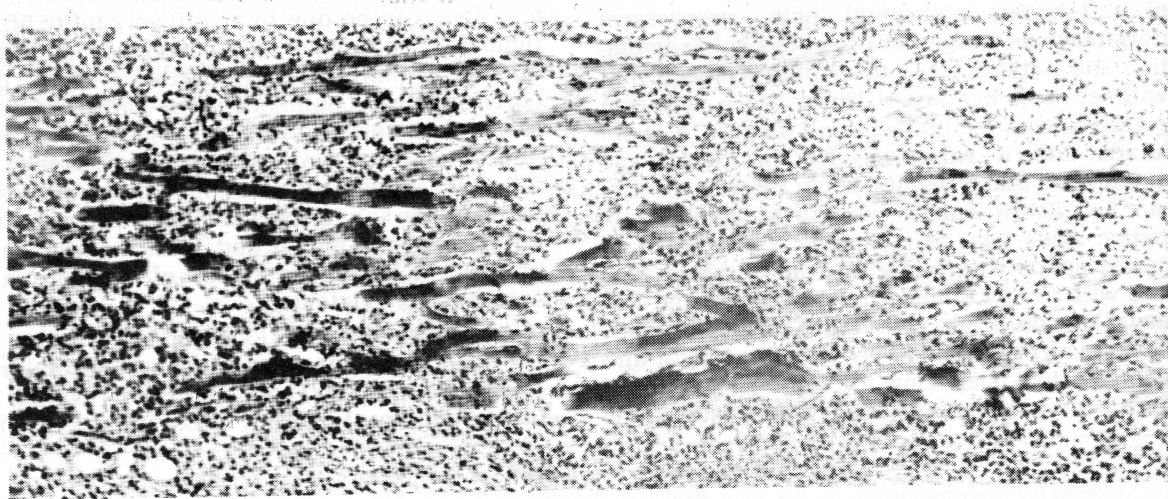


Figure III Infiltration of the myocardium with the malignant lymphoid cells.

(H & E X 250)

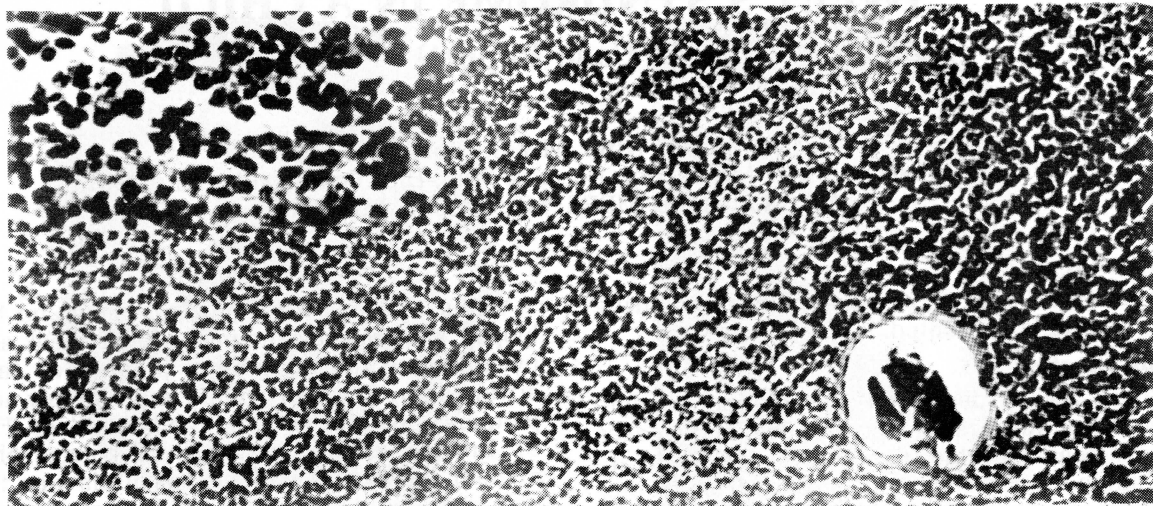


Figure IV Almost complete replacement of the renal tissue by the malignant lymphoid cells (H & E X 250); inset shows the cells at higher magnification (X 400).

Discussion

Cardiac involvement in lymphoma is either nodular or infiltrative in type. Roberts et al (1969) found pericardium and right sided myocardium to be the most common sites of lymphomatous infiltration. In most cases, cardiac abnormalities are usually limited to the ECG changes alone; however, patients with extensive cardiac involvement as in this case are likely to be symptomatic. In addition to infiltrative constrictive pericarditis, this patient also had bulky tumour growths in the right atrium and ventricle; obstructive cardiomyopathy with congestive cardiac failure was the final outcome. This type of cardiac involvement is rare in DNHL even with systemic spread (Davis, 1975, Roberts et al 1969).

Malignant retroperitoneal fibrosis is an infrequent complication of lymphoma (Nitz et al, 1970; Thomas and Chisholm, 1973, Treves, 1958). In the present case the reactive process besides the proximal ureter, had surrounded the left kidney as well.

Both renal parenchymal infiltration and malignant retroperitoneal fibrosis contributed to radiological nonfunction of the left kidney. Whilst renal involvement is not uncommon in DNHL (Kielly et al 1969), infiltration of this magnitude seems to be uncommon.

The present case is unique because it combines three rarely observed anatomic lesions due to massive metastatic infiltrations. This report also serves to emphasize the importance of including DNHL in the differential diagnosis

of a case presenting with multiple organomegally even in the absence of peripheral lymphadenopathy.

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