# TWO SOUTH WEST AFRICAN CASES OF BURKITT LYMPHOMA*

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SUMMARY

*Two cases from South West Africa presenting with intra-abdominal Burkitt tumours are described. One patient, after an initial dramatic response to vincristine sulphate therapy, relapsed and died, apparently insensitive to further chemotherapy. The second patient died before the histological diagnosis became known. These are, as far as is known, the first cases from South West Africa to be described. The climatic and geographical conditions of the territory of South West Africa are discussed in view of the differences as compared with those of countries in which Burkitt's tumour occurs as a common malignancy in childhood.*

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The African lymphoma syndrome has featured prominently in the literature since attention was drawn to a number of unusual features surrounding the disease.[1-5] From the numerous observations and studies on the so-called Burkitt lymphoma, those of the high cure rate by means of chemotherapy[6-8] and those of the roles played by epidemiological,[7,9] immunological[10] and viral agents in the aetiology of the disease[11] have probably been the most fascinating. New hope and interest have been stimulated in cancer research following these findings.

Two cases of Burkitt's tumour syndrome from South West Africa were recently seen. One patient came from the Kamanjab district in South West Africa while the other developed the lymphoma in the Kavango territory. (Fig. 1).

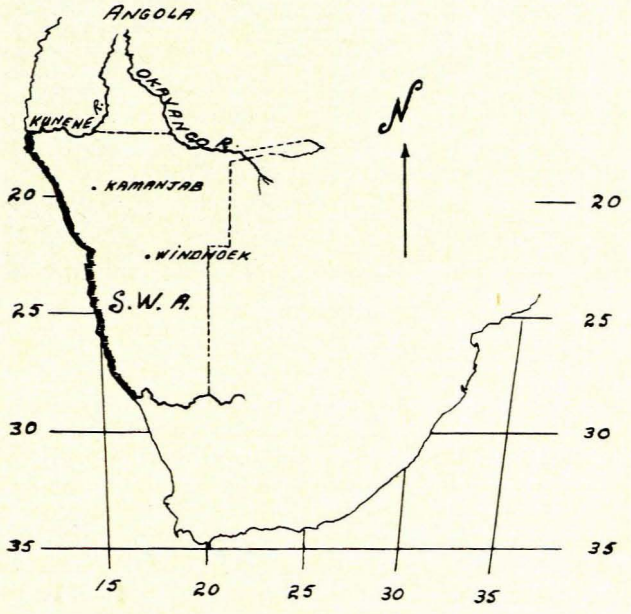


*Fig. 1.* Localities in South West Africa from where the two cases of Burkitt lymphoma came.

The occurrence of 2 cases with the syndrome in South West Africa within a period of 6 months is reported because of the exceptional geographic and climatic conditions prevailing in this territory compared with that of other countries where this malignant disease is common in childhood.

## CASE REPORTS

*Case 1*

A 10-year-old Bantu girl was admitted to hospital in Windhoek from the north-western area of South West Africa with symptoms of vomiting, diarrhoea and abdominal pain for 14 days. The parents had noticed considerable weight loss and abdominal distension for probably one month before admission. The patient had lived all her life in the Kamanjab area where her father worked as a farm labourer. On admission to hospital, she was found to be a frail-looking girl in whom there was obvious marked weight loss and considerable abdominal distension. On examination it was found that the distension of the abdomen was due partly to ascites and partly due to large firm irregular abdominal masses. Peripheral lymphadenopathy was absent.

A full blood count showed a haemoglobin level of 9·1 g/100 ml. The white cell count was 8 000/mm³ and the differential count was within normal limits. Features suggestive of a leukaemia were not observed at any time during regular haematological investigations.

Radiography of the skeleton did not at any time show evidence of bony metastases.

Intravenous pyelography on different occasions did not indicate any abnormalities of the genito-urinary system.

A laparotomy carried out soon after admission to hospital showed extensive neoplastic masses, most of which involved the mesentery of the small intestine. A large mesenteric growth which infiltrated the ileum was resected, but removal of further growth deposits in the pelvic and abdominal cavities was not attempted (Fig. 2). Histological examination of the resected tumour showed the well-known features of Burkitt's African lymphoma of the poorly differentiated lymphocytic type (Fig. 3).

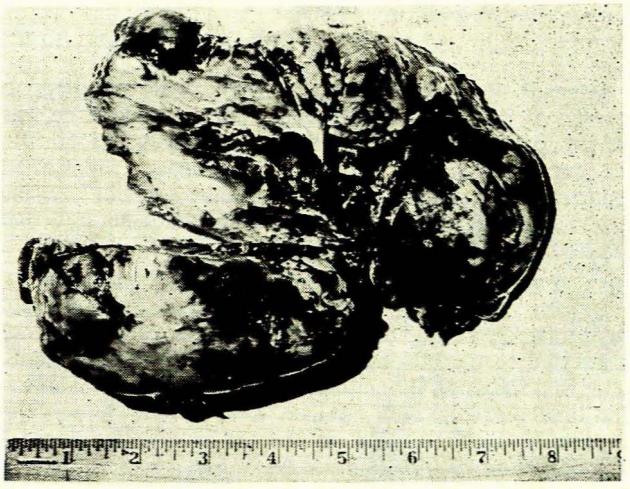


*Fig. 2.* The laparotomy specimen of the tumour in Case 1.

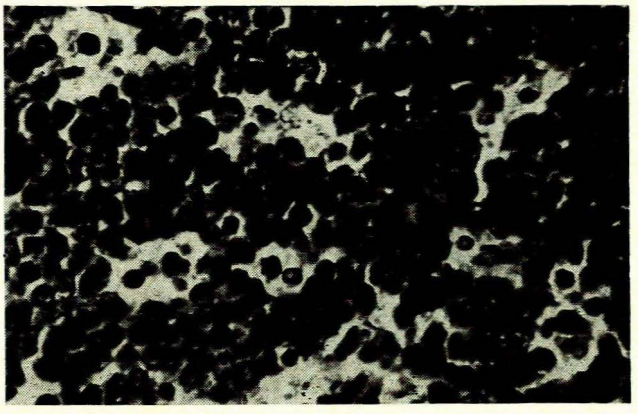

*Fig. 3.* Section of the tumour in Case 1 in which a typical 'starry-sky' appearance was seen.

Intravenous injections of vincristine sulphate in 1-mg doses weekly were instituted. This treatment caused the extensive intra-abdominal tumour to disappear dramatically within a period of 4 - 5 days. Following on the satisfactory response to the drug, the general condition of the patient improved. She developed an appetite and was able to get out of bed for the first time since admission. However, some 3 weeks later, while still receiving weekly injections of vincristine sulphate, the abdominal tumour masses reappeared rapidly. Methotrexate therapy was started but her condition deteriorated rapidly, and the neoplasm appeared to be completely resistant to chemo-

therapeutic drugs. The disease from the time of its recurrence progressed relentlessly to a rapid fatal termination.

A postmortem examination showed intra-abdominal tumours involving the mesentery, intestines, retroperitoneal tissues and pelvic organs. The growth had caused matting of intra-abdominal lymph nodes and organs, making it impossible to determine a primary site of the growth with any precision. The kidneys, spleen, liver, suprarenals and pancreas did not show tumour deposits. There was no evidence of any bony metastases or of peripheral or intrathoracic lymph-node involvement.

*Case 2*

A small Bantu boy aged 5 years was admitted to hospital at Rundu in the Kavango territory where he was treated for urinary schistosomiasis. Shortly before he was due for discharge from hospital, a mass, which had apparently developed rapidly, became palpable in the abdomen. Peripheral lymph nodes were clinically of normal size and consistency.

At laparotomy growths in the small intestinal mesentery infiltrating the ileum were found. These tumours had apparently developed in mesenteric lymph nodes and had progressed to partial obstruction of the wall of the ileum. After the operation the patient's general condition deteriorated unexpectedly and he died a few days later. The laparotomy specimen (Fig. 4) showed histological features of a lymphoma belonging to the same group of poorly differentiated lymphosarcomas as in case 1 (Fig. 5).

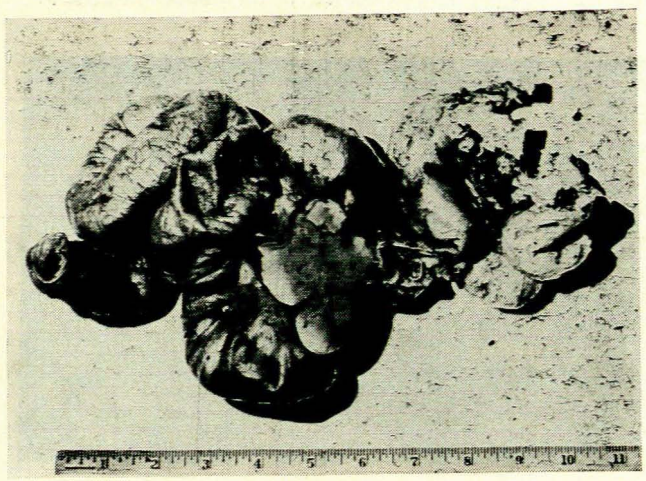

*Fig. 4.* Naked-eye view of the mesenteric tumour mass infiltrating the ileal wall in Case 2.

Radiographic investigations for skeletal or renal metastases were not carried out as the histopathological report on the operation specimen was received only after death. Full blood counts carried out before the operation did not show any abnormality of the peripheral blood.

### CLIMATIC AND PARASITOLOGICAL DATA OF SWA

The annual rainfall over most of South West Africa is sparse. The Kamanjab area, whence patient No. 1 was sent, lies at an altitude of 1 300 m above sea-level and has an average annual rainfall of probably not more than

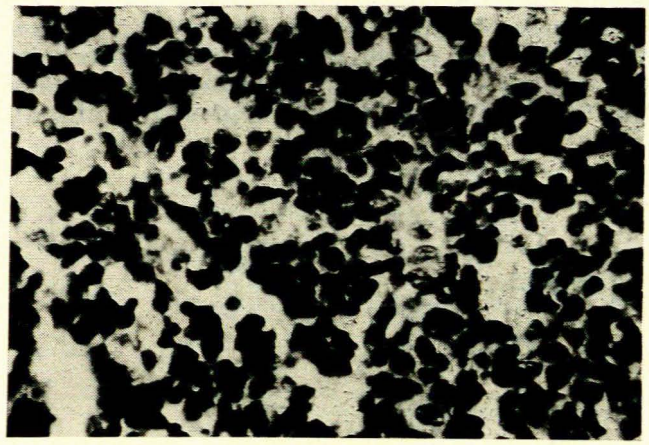

*Fig. 5.* Section of the growth in the intestine showing an undifferentiated lymphosarcoma; the 'starry-sky' pattern was less marked in the second case.

300 mm. The Okavango river region, of which patient No. 2 was a native, has, with more than 600 mm per annum, the highest rainfall in South West Africa. The Kavango is a thickly-wooded country lying at an altitude of approximately 1 000 m above sea-level. Malaria is hyperendemic in the Okavango River region,[12] but only sporadic cases occur annually in the Kamanjab district. Vigorous malarial control measures are in constant operation in the Kavango territory where the population live for the most part, because of fishing, along the banks of the Okavango River. The river which forms the border between South West Africa and Angola is infested with bilharzia, and a very large section of the population suffers from the disease.

### DISCUSSION

Two cases which fall clinically and histopathologically into the group of lymphosarcomas referred to as Burkitt's African lymphoma occurred in South West Africa within 6 months of each other. To our knowledge the occurrence of this form of lymphosarcoma has not been previously recognized in South West Africa. It seems feasible, however, that as the medical services in the territory are extended and as transport facilities improve, further cases may be expected in future. Cases of the Burkitt type of lymphoma have been described from Portuguese East Africa[13] and from various centres in the Republic of South Africa.[14-17]

Recently a 3-year-old child, domiciled in Sa da Bandeira in Angola and of white Portuguese parentage, was referred to Windhoek, suffering from a typical facial type of Burkitt lymphoma. A biopsy specimen of the tumour examined histologically confirmed this clinical diagnosis but the child returned to Angola before adequate treatment was given and has since been reported to have died of the disease. While in Windhoek, peripheral lymphadenopathy was at no time observed.

The incidence of the so-called African lymphoma in South West Africa could probably be regarded as 'occasional' or 'rare'.[7] Certain sections of the Okavango River with its swamps and the Caprivi Zipfel may well prove, on climatic, geographical, epidemiological and entomolo-

gical grounds, to have a higher incidence of the African lymphoma syndrome than elsewhere in South West Africa or the Republic of South Africa. The Okavango River area, along which the entire Bantu population of Kavangoland is concentrated, is a mosquito-infested country. For mainly geographical reasons the intensive malarial control measures in the Kavango probably do not produce the same striking success as in other parts of the territory. Bilharzia infestation with both *S. haematobium* and *S. mansoni* in different localities is present in a varying but always very high percentage of the Kavango people. Sporadic cases of trypanosomiasis occur along the western Caprivi, but all have occurred in members of nomad tribes who probably acquired the disease in countries to the north of the South West African border.

There seems to be considerable evidence that the Burkitt type of poorly differentiated lymphosarcoma in children described from some parts of Africa, merits its being placed into a special category for several reasons. The commonest clinical presentation of the growth is in the maxilla and/or mandible or intra-abdominally.[2,18,19] Peripheral lymphadenopathy and leukaemic changes are notably absent in patients with the Burkitt lymphoma.[20,21] The Burkitt lymphoma, which is the commonest malignant tumour of children in some parts of Africa, exhibits rapid growth and responds to chemotherapeutic treatment resulting in what is for lymphomas a high and unique percentage of cures.[6,8]

An association with the epidemicity of malaria and with the immunological response evoked by malarial infestation further serves to emphasize the individuality of the Burkitt tumour in the lymphoma group of cancers. Morbid anatomical findings indicate a pattern of development of the disease in children which is different from that of the classical lymphosarcoma at this age. The World Health Organization's memorandum on the histopathological definition of Burkitt's tumour concludes, among other things: 'the eponym Burkitt's tumour is best applied to a malignant neoplasm of the haemopoietic system composed of a predominant and characteristic cell type'. The same memorandum emphasizes, however, that the prominent so-called 'starry-sky' feature seen histologically is not specific or pathognomonic of Burkitt's tumour.[21] It could be argued, therefore, with some justification, that the Burkitt lymphoma merits consideration as a separate independent tumour entity within the lymphosarcoma subdivision of malignant lymphomas. Burkitt's tumour is seen, notwithstanding its special features, also in other parts of the world besides Africa and it cannot be denied that certain histopathological features of Burkitt's tumour may be found in lymphomas of other types as well.

Both the Bantu cases reported showed intra-abdominal tumour masses. Case 1 did not show any bony lesions and diffuse tumour growth in the abdominal and pelvic cavities would have obscured any primary site of the growth had it been present. An autopsy was not carried out in case 2 because the patient died at Rundu before the histological findings were known. Both cases fitted into the group presenting with abdominal tumours and reported to comprise 40% of all cases with the disease in Africa.

In hyperendemic or holo-endemic malarial areas strict preventive control measures of the vector mosquitoes could result in a diminution of the incidence of Burkitt lymphoma. It has not, as far as can be judged, been established whether other chronic parasitic disease processes play a role in the aetiology of the disease in the same way as malaria seems to do. Could chronic bilharzia be of importance in some areas of Africa as regards an altered immunological response to a virus causing lymphoma? Does an immunological reaction to the mosquito toxins of repeated bites over a long period change the response to a known viral agent so that Burkitt tumours result? It may well be that we are not dealing, as is thought, with a new malignant disease but that there are other aetiological routes along which it may be acquired and that only the expression of the disease differs, i.e. a change of the who, where, why and how of the illness. It could be that, if the Epstein-Barr virus is responsible,[10,11] a shortcut opens up for a malignant transformation of cells by simultaneously operating immunological factors. Other less potent and as yet undiscovered factors in other communities of the world may operate to produce lymphosarcomas as a result of an infection by the same, or other, virus. All histopathologists of experience have at some time examined a lymph node of suspected glandular fever cases with some misgiving about the benignity of the condition. Many of us have been worried by the marked atypicality of the glandular fever cells in peripheral blood smears of such patients and have remembered Damashek's remarks with some uneasiness of mind.

Whatever the final word on the Burkitt lymphoma may be, no one can deny the importance of the outstanding work which has been done in many parts of Africa, which leaves one, when reviewing the extensive literature, with the impression that important progress has been made in the solution of some of our cancer problems.

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