THE SO-CALLED BURKITT LYMPHOMA IN A WHITE CHILD FROM KIMBERLEY

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The Burkitt lymphoma has captured the imagination of clinicians and pathologists and, led by Burkitt, it has in some cases induced flights of fancy regarding aetiology, distribution and basic pathology, out of proportion to the facts.

In collaboration with Haddow, Burkitt postulated a climatic area in which the condition was said only to occur and they excluded from this areas above 5,000 ft. in altitude. The latter was said to be a reflection of temperature, of which a minimum mean seasonal value of 60°F was said to be the limiting factor. Burkitt diagrammatically showed that cases in the Republic of South Africa were only to be expected to occur in the coastal plain below 1,000 ft. and north of Natal.

The purpose of this publication is to report a case of this condition in a White child, a male aged 3 years, from Kimberley. This child had never been out of the town. The altitude of Kimberley is 3,950 ft. and the lowest mean seasonal temperature is 36°F.

The presentation was sudden. Apparently the child had been quite well until the day of admission, when he complained of pain in the right lower abdomen. On examination there was no fever. No anaemia was found and no lymphadenopathy or splenomegaly. The liver was slightly enlarged. The total leucocyte count was 7,000/cu.mm. A tender mass was felt in the right iliac fossa and intussusception was suspected. Laparotomy was performed. The appendix was found more or less encased in a mass of soft, friable, grey-white, haemorrhagic glands. The mesentery of the small bowel as well as the omentum and surface of the small bowel were found studded with umbilicated nodules about 2 cm. in diameter. The visible surface of the liver showed the same picture. All the nodules had a haemorrhagic appearance. Biopsy of the glands around the appendix was sent for histopathological examination. This showed malignant lymphomatous infiltration with destruction of gland architecture and extension beyond the capsule. There was an abundance of large macrophages with

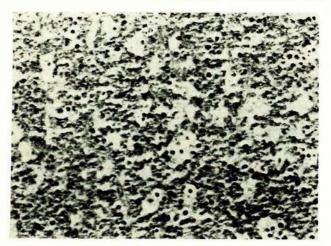


Fig. 1. See text.

vacuolated cytoplasm, in which cell debris could be seen. The periodic acid-Schiff stain was negative. Fig. 1 is a high-power view. The diagnosis of the so-called Burkitt lymphoma was made and the histological sections were seen by 3 referees, who agreed. Subsequent radiological examination showed no abnormality of the skull, thoracic cage and long bones. The child died 7 days after the laparotomy. Unfortunately permission for autopsy was not obtained.

DISCUSSION

Burkitt and O'Conor³ described the clinical presentation, limited geographical distribution and pathological features of this condition. Only half of their cases had jaw tumours, and yet they considered this a characteristic feature. O'Conor' stated that with the exception of jaw tumours, lesions occur in all sites where malignant lymphoma can be expected to occur. Burkitt⁵ states that clinical lymphnode enlargement and splenic enlargement is exceptional. This statement is open to question, because in the same article one reads that cases with obviously malignant abdominal tumours are seldom transported over long distances to hospitals in North Africa. Many cases are therefore not seen. The author concludes that abdominal swelling may be an even more common presenting feature than jaw tumours. It is also stated that the epigastric mass, which is often felt, has at autopsy been found to consist of neoplastic tissue and is believed to have originated in lymph glands. It is stated that although clinical enlargement of lymph glands is uncommon, they are often found to be affected on histological examination.

From the foregoing it appears that organ involvement is not basically different in this condition than in any other malignant lymphoma.

The rapid progression to a fatal termination in the case described, reflects the malignant propensity. The umbilication seen in the deposits, is a morbid anatomical manifestation seen in other rapidly growing malignancies. The so-called 'starry sky' effect, seen in the lymph glands, is due to the large numbers of giant macrophages. Their presence must be due to rapid growth leading to increased cell necrosis and consequent necessity for phagocytosis of cell debris. Similar phagocytes may be seen in varying numbers in any reactive lymph gland and in malignant lymphomas, although in lesser numbers. The question may be asked, whether the Burkitt lymphoma is an entity or merely a very malignant form of ordinary lymphoma.

Other cases⁶⁻¹⁰ have been reported in areas of the Republic of South Africa where the climatic conditions that have been postulated do not obtain. The evidence which Burkitt put forward in his writings to support the suggestion that the aetiological agent was a virus, transmitted by an arthropod which thrives in certain climatic regions, appears to be unfounded.

SUMMARY

A case of the so-called Burkitt lymphoma in a White child from Kimberley is described. The rapid progression to a fatal termination is outlined. The morbid anatomical features which

were seen in the form of haemorrhagic umbilicated nodules in the abdomen are noted. The typical histological appearance of the 'starry sky' effect in the lymph glands was found, and this is presented as a manifestation of rapid growth leading to cell necrosis requiring large numbers of phagocytes-what these cells have been found to be. It is suggested that this malignant lymphoma is essentially not different from other malignant lymphomas and could merely be an extremely malignant form. The occurrence of another case in an area where the climatic conditions originally postulated for this tumour, do not obtain, indicates that the aetiology that has been put forward is probably incorrect.

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REFERENCES

- 1. Burkitt, D. (1962): Ann. Roy. Coll. Surg. Engl., 30, 211.
- Idem (1962): Brit. J. Cancer, 16, 379.
 Burkitt, D. and O'Conor, G. T. (1961): Cancer (Philad.), 14, 270.
- 4. O'Conor, G. T. (1961): Ibid., 14, 270.
- 5. Burkitt, D. (1962): Postgrad. Med. J., 38, 71.
- Gluckman, J. (1963): S. Afr. Cancer Bull., 7, 7.
 Chapman, D. S. and Jenkins, T. (1963): Med. Proc., 9, 320.
- Correspondence (1963): *Ibid.*, 37, 476.
 Brüning, E. (1963): *Op. cit.*¹⁰
- Schmaman, A. G., Gampel, B. and Luntz, C. H. (1965): S. Afr. Med. J., 39, 741.