A CASE OF KAPOSI'S HAEMANGIOSARCOMA

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(RECEIVED FOR PUBLICATION JULY 20, 1951)

In 1872 Kaposi described a dermatological condition which he called 'idiopathic multiple pigment sarcoma'. At the present time it is often known as Kaposi's haemangiosarcoma. Kaposi described it as affecting the skin, and it certainly affects this part most often, but for years it has been known to involve other organs. In 1940, Choisser and Ramsey reviewed the literature and mentioned a total of 600 cases since 1872. Stats (1946) lists the sites which, after the skin, may be affected most often. He mentions the glans penis, the submucosa of the gastro-intestinal tract, the respiratory tract and the lymph nodes. Less common sites are the spleen, liver, heart, diaphragm and bladder. Seagrave (1948) has described a case in which the submucosa of the gastro-intestinal tract and the skin were involved. In such cases blood is often found in the stools. The disease is said to be rare in negroes, but Kaminer and Murray (1950) record 38 cases in the South African Bantu occurring in the period 1942-49. It is certainly very uncommon in children and appears to occur mostly in adults over the age of 40. MacKee and Cipollaro (1936) mention an incidence of less than 1% under the age of 10 years.

The sex incidence is predominantly male. Denzer and Leopold (1936) recorded the clinical aspects of the disease in a male child of 4½ years; the necropsy findings were subsequently described by Stats (1946). The case recorded here is that of a boy, and it illustrates the very uncommon form of the disease without skin manifestations. Choisser and Ramsey (1940) described two cases of Kaposi's tumour of the heart without skin lesions; in these cases the right auricle was involved. Cardiac nodules have also been described in cases with cutaneous involvement. Tedeschi, Folsom and Carnicelli (1947) mention three cases affecting viscera without skin manifestations. These authors consider the disease to be systemic in nature

affecting various parts at the same time. Aegerter and Peale (1942) consider the condition to be a vascular cancer and regard the visceral lesions as true metastases. However, lymph node involvement is often not regional to any affected organ, and supports the view that the disease is a systemic affection like a reticulo-endotheliosis, although the disease is generally considered to be neoplastic.

The case recorded here illustrates this point.

Case Report

A Bantu male child aged 6 years was admitted to the Baragwanath Hospital in July, 1950. He had developed a sore throat and swelling of the neck a week before. There were no other symptoms, and his previous history was satisfactory.

The parents were healthy and three siblings were all apparently well.

The patient was a fairly well nourished child lying comfortably in bed. He did not appear ill or distressed. The temperature was 97.4°, the pulse 78 per minute, and respiration 26 per minute.

Careful examination of the entire skin and the genitalia revealed nothing abnormal. There was no oedema of the extremities. (Swelling of the extremities has been noted by various authors as the first sign of this disease.) The tongue and mucosa of the mouth were normal. The tonsils were slightly congested but not enlarged.

Nothing abnormal was detected in the clinical examination of the cardiovascular or respiratory systems. The blood pressure was 100/65.

The central nervous system was normal. Examination of the abdomen revealed a palpable liver one finger breadth below the costal margin. It was firm, regular and not tender. The spleen was palpable two finger breadths below the costal margin. It was firm but not tender.

The lymph nodes of the occipital, cervical, submental, right axillary and both inguinal regions were all palpably enlarged. The largest was about 3 cm. by 2 cm. They were discrete, felt distinctly rubbery, were freely mobile, and were not tender.

Investigations. The Mantoux test was negative up to 1:10.

A full blood count on admission gave: Hb. 14.9 g.%, leucocytes 9,800 per c.mm. (neutrophils 47%, monocytes 9%, lymphocytes 39%, eosinophils 5%), packed cell volume 44%, sedimentation rate 23 mm. per hour. The corrected sedimentation rate was slightly increased. Malaria parasites were not observed. A subsequent blood count, five weeks later, gave: Hb. 12.6 g.%, leucocytes 8,800 (neutrophils 42%, monocytes 3.5%, lymphocytes 46.5%, eosinophils 7.5%, basophils 0.5%).

The standard Eagle test and the modified Ide test were both negative.

The Paul-Bunnell test was negative.

The benzidine test for occult blood in the stool was negative.

Radiographs of the chest showed no abnormality and no evidence of mediastinal glandular enlargement. The heart was normal in size and shape.

The urine had a specific gravity of 1020. Tests for albumin were negative.

An electrocardiogram was normal.

A sternal marrow puncture gave the following results: blast cells 0.5%; myelocytes (neutrophil 2%, eosinophil 1%) 3%; metamyelocytes (neutrophil 3%, eosinophil 1.5%) 4.5%; staff cells 4%; polymorphonuclears (neutrophil 9%, eosinophil 9%) 18%; monocytes 1%; lymphocytes 23%; erythroblasts 3%; normoblasts 41%; plasma cells 1%; reticulum cells 1%; total nucleated cell count 42,400 per c.mm.; megakaryocytes 11 per c.mm. Myeloid: erythroid ratio 0.7:1; myeloid: lymphoid ratio 1.3:1.

The bone marrow was of normal cellularity with slightly increased reactions in the lymphoid and erythroid series. This was borne out by the reduced myeloid: lymphoid and the reversed myeloid: erythroid ratios.

A special feature was the increase in eosinophils in all stages of maturation.

A skin biopsy taken from the anterior abdominal wall did not reveal any abnormality on histological examination.

A needle liver biopsy showed no pathological lesion.

An inguinal lymph node biopsy showed, on histological examination, loss of the normal architecture and its replacement by a fibro-sarcomatous type of tumour with numerous blood spaces in which mitotic figures were plentiful. The histological features were strongly suggestive of Kaposi's haemangiosarcoma (Dr. J. Higginson).

An enlarged axillary lymph node was subsequently removed and its microscopical examination confirmed the presence of a similar lesion to that found in the inguinal gland. Here the report indicated the presence of a haemorrhagic sarcoma with numerous blood vessels and moderate activity (Figs. 1 and 2). Some plasma cell infiltration was also noted.

Progress and Treatment. The diagnosis of a haemangiosarcoma of the Kaposi type was established. It was decided, before instituting general treatment, to apply x-ray therapy to a single enlarged lymph node and assess the effect of the treatment by its subsequent

removal and examination. Therefore, a single dose of 1000 r of medium voltage x-ray therapy (135 K.V.) was applied to an enlarged submental gland of about 2 cm. in diameter. This gland became smaller, and four weeks after the treatment it was removed and examined, disclosing the presence of interstitial fibrosis which indicated a definite response to the therapy (Figs. 3 and 4).

Four weeks after the biopsy, the child was given general x-ray therapy as follows:

Left groin	..	total dose of 1500 r (180 K.V.)
Right groin	..	total dose of 1000 r
Right lateral neck	..	total dose of 1200 r
Left lateral neck	..	total dose of 1200 r
Right axilla	..	total dose of 2000 r
Spleen	..	total dose of 1500 r

This treatment was applied for seven weeks, and after it was completed the liver and spleen were no longer palpable, and only small, shotty glands could be felt in the groin, axilla and neck.

Discussion

The presenting features in this child were generalized lymphadenopathy and hepatosplenomegaly. Various diagnostic possibilities, such as tuberculosis, syphilis, glandular fever, leukaemia, were excluded by appropriate investigations. The final diagnosis of Kaposi's haemangiosarcoma could only be made by lymph node biopsy and histological examination. This was confirmed by the presence of the same lesion in another gland taken from a different region. This case appeared to be remarkable because it showed none of the cutaneous lesions which commonly occur in this disease.

The patient had an eosinophilia, which has been found in other cases. Dörffel (1932) mentions secondary anaemia as a common feature but this was not present. The sternal marrow puncture specimens showed an increase in the eosinophils in all stages of maturation. There was normal cellularity with slightly increased reactions of the lymphoid and erythroid series. Stats found an active bone marrow with a normal cellularity in two cases. In this case the spleen was enlarged and the palpable enlargement disappeared after x-ray therapy. We might assume that the disease was present in this organ, although there was no absolute proof. Symmers (1941) reported a case in which the spleen, weighing 1,000 g., was filled with the Kaposi's lesion. But in other cases of splenomegaly in this disease, the haemangiomatous deposits were not present; Dalla Favera (1911) records such a case.

The needle liver biopsy did not reveal any abnormal histology, but this need not preclude the presence of the disease in this organ.

FIG. 1.

FIG. 2.

FIGS. 1 and 2.—Kaposi's tumour before treatment (low power). High power view of Fig. 1.

FIG. 3.

FIG. 4.

FIGS. 3 and 4.—Low power view of lymph node of Kaposi's tumour after x-ray therapy showing a definite response by the presence of interstitial fibrosis. High power view of Fig. 3.

Clinical, electrocardiographic and radiological examination of the heart did not reveal any abnormality, but, here again, one cannot exclude the presence of lesions in the heart. Occult blood is often found in the stools of patients with gastrointestinal involvement, but this patient had no such abnormality. A skin biopsy showed no abnormality and there was no clinical evidence of any skin lesions.

In this case most of the superficial lymph nodes were enlarged, but there was no radiological evidence of mediastinal glandular involvement or clinical evidence of abdominal glandular enlargement. We have only evidence of generalized superficial lymph node involvement and possible splenic affection, and this might be in favour of relating this disease to the reticulo-endothelioses. X-ray therapy given to the affected glands and the spleen led to a great diminution in their size.

Summary

A case is presented of Kaposi's haemangiosarcoma occurring in a South African Bantu boy of 6 years. The superficial lymph nodes and spleen were affected without cutaneous involvement. The diagnosis was made by lymph node biopsy and histological examination. A single lymph node was first treated with x-rays, and the effect on the disease process assessed by its subsequent removal and microscopical examination. General x-ray

treatment was applied, and the enlarged lymph nodes and spleen gradually became impalpable. The literature is briefly reviewed.

I should like to thank Dr. J. D. Allen, Medical Superintendent, Baragwanath Hospital, for permission to publish this case. I am also indebted to Dr. S. C. Heymann, Head of the Paediatric Department, University of the Witwatersrand, for his helpful criticism; to Drs. M. P. Shapiro and L. Cohen, Radiotherapy Department, Johannesburg Hospital, for their helpful advice and for allowing me access to their therapy notes; to Dr. J. Higginson for histopathological reports; to Dr. H. Hirsch for laboratory investigations; and to Drs. M. Shnier, P. Marchand, M. Goldberg and D. Warnes for providing biopsy specimens.

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ADDENDUM

This patient has been followed up at regular intervals for a year after the completion of x-ray therapy, and has remained completely free of any recurrence of the disease. He has no palpable lymph nodes and his spleen cannot be felt. Details of the x-ray treatment given to this patient was as follows:—

Left groin	total dose of 1500r in 5 daily fractions
Right groin	total dose of 1000r in 2 daily fractions
Right lateral neck	total dose of 1200r in 6 daily fractions
Left lateral neck	total dose of 1200r in 6 daily fractions
Right axilla	total dose of 2000r in 10 daily fractions
Spleen	total dose of 1500r in 10 daily fractions
The submental gland was treated with one dose of 1000r.	