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Clinical Features of Kaposi's Sarcoma Amongst Rhodesian Africans

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Kaposi's sarcoma, a relatively rare malignancy in the Caucasian world has proved to be surprisingly common in certain parts of Africa particularly the Congo, East Africa and Rhodesia. There is still a great deal of mystique surrounding this particular form of malignancy in its origin, in its presentation, and in its treatment. This is probably because even the largest centres in Europe and America have little experience of the condition and it is a relatively new disease to research.

There are few clues as to the origin or etiology of this disease though a great deal has been written on the subject and discussed at the International Symposium held on Kaposi's sarcoma at Kampala. To the credit side, the treatment of Kaposi's sarcoma has been rewarding. It would appear that success can be claimed in certain cases with chemotherapy and certainly, the tumours are radio-sensitive. Kaposi's sarcoma is an ideal form of malignancy for therapeutic research. The lesions are cutaneous and subcutaneous in the majority of cases and are therefore visible. They are multicentric in origin and the natural history of the disease is slowly progressive with remissions and regression. Thus a clinical trial can be undertaken with the fore-knowledge that time is on the side of the researcher.

There has been speculation on an association between Kaposi's sarcoma and other tumours and between Karposi's sarcoma and tropical diseases prevalent in the same areas as Kaposi's sarcoma. Unfortunately, there is still no constant link or constant feature running through all affected geographical areas.

Fifty-eight new cases presenting at Harare Hospital between 1967 and 1971 were reviewed on their clinical features. These features are to be compared with the features described by workers from other centres.

SEX INCIDENCE

Fifty-five cases were males and three cases were females. This corresponds with all other areas where 90 per cent. of the cases occur in males.

AGE

Table 1 shows the spread of occurrence of the tumour throughout all age groups but the maximum incidence is at 60 years of age and over. It is of interest to notice that ten cases occurred between the ages of 20 and 29.

Table 1: Ages of cases seen

Ages 1 year to 9 years	: 3 cases
Ages 10 years to 19 years	: 2 cases
Ages 20 years to 29 years	: 10 cases
Ages 30 years to 39 years	: 7 cases
Ages 40 years to 49 years	: 8 cases
Ages 50 years to 59 years	: 4 cases
Ages 60 years and over	: 24 cases

Only three patients were females and the age range was 13 years, 25 years and 65 years.

CLINICAL FEATURES OF THE DISEASE

There are probably five major divisions in the presentation of Kaposi's sarcoma.

- (1) The common presentation.
- (2) Presentation in the female.
- (3) Kaposi's sarcoma in children.
- (4) The fulminating form in both sexes.
- (5) Unusual presentations.

Commonly, it is a disease of middle age or of the elderly. There are few cases in which any form of prodromal symptom is admitted. Itching has been described in only four of the cases and in one case an itch was the definite presenting feature that drew attention to a small nodule under the skin. Only four cases complained of pain in the area involved. The basic presenting feature was the formation of nodules.

NODULES

The nodule is the commonest lesion of Kaposi's sarcoma. Twenty-eight of our cases presented with typical nodules. The nodules occurred on the extremities in most cases and varied from a small verrucous nodule, rather like a dried up wart, to a large red turgid nodule the size of a cherry or

small walnut. The average lesion was approximately 0.5 cm. in diameter lying in the dermis and bulging through the epidermis which was thinned over the surface. Many nodules are felt rather than seen and the nodule may eventually ulcerate through the skin. This ulceration may take the form of a true sarcomatous ulceration bursting through the skin or it may actually involve the skin and grow within the skin. Nodules may be single or confluent and are often associated with plaques within the skin.

The nodule often gives the impression of being encapsulated but it is only the tip of the iceberg. At biopsy it becomes apparent that the nodule extends into the subcutaneous tissue and that other nodules actually occur within the subcutaneous tissue. Nodules may regress and it is often found that the nodule becomes flat and wrinkled after several months or after treatment and may actually disappear leaving a slightly pigmented scar.



Fig. 1.—Typical verrucous lesions of Kaposi's sarcoma on the heel of an African male. The lower lesions are fleshy while the lesions on the pigmented surface tend to be more wart-like.

PLAQUES

Plaques may be associated with the nodule or occur alone. The classical plaque described in European literature as of a violaceous hue will, in the African, be a black plaque surrounding verrucose or reddish nodule. The plaque may however occur without the nodules and involve the whole of the dermis and epidermis producing a pachydermatous effect.

ULCERATION

Nodularity may lead to ulceration. In all our cases it would appear that the ulceration had originated in nodules. These nodules may coalesce and then break down and ulcerate.



Fig. 2.—A typical sarcomatous lesion bursting through the skin of the palmar surface of the hand. On the ring finger a lesion which is invading the skin can be seen. Biopsy revealed Kaposi's sarcoma.

The ulcers as previously described may burst through the skin or involve the skin. Only the earlier lesions appear to burst through the skin. Once the ulceration has become established the skin is rapidly involved in the process and a typically malignant ulcer may form. Eight cases showed ulceration in this series. Figures 2 and 3.

Interestingly enough, it is with the formation of nodules and ulceration that pain is often associated. When the nodules are closely grouped together, or when there is severe oedema then pain may be experienced. The more vascular the nodules the more likely they are to be tender to pressure. No fully developed lesion is really ever itchy. Despite the presence of ulceration and the formation of nodularity there is little constitutional disturbance at this stage except in advanced cases.



Fig. 3.—The fingers of the patient with Kaposi's sarcoma showing the invasion of the skin by a Kaposi's nodule which has ulcerated, and on the adjacent finger Kaposi's sarcoma which has not yet ulcerated.

OEDEMA

In the classical case oedema is associated with Kaposi's sarcoma. It has been stated that the oedema is very rarely a pitting oedema but in our experience this is not true. The oedema is almost invariable if a patient is examined with this in mind and it is ab-initio a pitting oedema becoming a true lymphatic oedema with progression of the disease. Eventually it becomes a classical non-pitting variety similar to elephantiasis as the disease progresses within the limb.

Ulceration may lead to an increased swelling of the tissues which is not due to central lymphatic obstruction. The oedema that occurs in Kaposi's sarcoma is also thought not to be due to central lymphatic obstruction. It may become severe enough to produce vesiculation in the hands and over the wrist. The oedema tends to disappear after successful treatment with cytotoxic drugs or radiotherapy.

PRESENTATION IN FEMALES

In all descriptions the presentation in the female case is a more rapid, severer onset and progress than in the male. The onset is associated with nodule formation, moderately severe oedema and in most cases the patient is a young woman. Two patients in this series were young (13 years and 25 years).



Fig. 4.—A young male with fulminating Kaposi's sarcoma illustrating the generalised oedema involving the face and chest wall. The presence of nodules on the chest wall and upon the face including the upper eyelid.

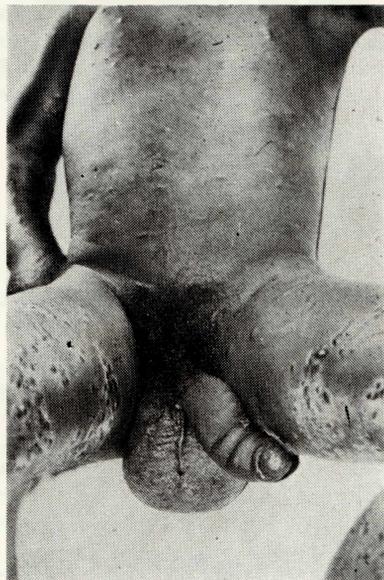


Fig. 5.—The body, scrotum and thighs of the same case with severe oedema, nodules and plaque formation.

The youngest female patient in this series at the age of 13 years presented with pain in both feet and developed a typical Kaposi's nodule. These nodules were biopsied and histological proof obtained. Subsequent course of this patient was one of rapid progression of symptoms with further nodules appearing and oedema spreading up both the lower limbs eventually involving the hands and eventually the face. The body did not appear to be oedematous. Treatment by radiotherapy produced a definite regression but this was not as effective as was expected in the male case, and cytotoxic drugs were instituted. Cytotoxic therapy produced a further regression and the patient is still under observation.

KAPOSI'S SARCOMA OCCURRING IN CHILDREN

In keeping with experience of all other African territories two children seen in this hospital under the age of ten years had presented with a cervical lymphadenopathy. These children presented with swelling of the cervical and inguinal lymph nodes. They were obviously ill, oedematous and had a pyrexia. There was anaemia and the picture resembled that of a lymphoma. However, when the biopsy was carried out it demonstrated the classical picture of Kaposi's sarcoma within the gland structure. Treatment with cytotoxic drug and transfusion produced a slight regression.

A third child however presented with numerous nodules that resembled subcutaneous lesions of cysticercosis. However, close examination revealed nodules and plaques typical of Kaposi sarcoma. Biopsy of a gland and of a nodule confirmed the diagnosis.

THE FULMINATING FORM

In this form of Kaposi's sarcoma the patient is usually a young person. It is a carapace of tumour involving bones, viscera and skin as described by Dr. Templeton of Kampala. In this group it is said that males occur only three times as commonly as female cases.

The whole of the patient is oedematous and the hands and feet show numerous nodules with nodules and plaques over the limbs. These nodules will often appear to follow the superficial veins. They may also be scattered all over the rest of the body involving the eyelids and encroaching even upon the conjunctive. The oedema will involve the genitalia. Two such cases were seen in this series, one of a man aged 25 years and one aged 70 years. Both patients also complained of severe abdominal pain. The lesions tended to ulcerate and produce large sloughing ulcers. It appeared that the ulceration developed in nodules but there was close association between the nodules and the numerous plaques occurring within the skin all over the patient. The progress



Fig. 6.—The dorsum of the hand in the same case as Fig. 4, showing an admixture of plaques and nodules with severe oedema.

varied in that the younger patient responded extremely well to cytotoxic drug therapy while the older patient died within four days. The length of history in the two cases were extremely short, the younger patient having a total history of four months and the elder patient giving a history of two months. Both cases had a history of pulmonary tuberculosis and were receiving sanatorium treatment when they developed Kaposi's sarcoma.

UNUSUAL PRESENTATIONS

Kaposi's sarcoma was discovered on biopsing a laryngeal nodule in a young woman of approximately 25 years of age. The sarcoma had, in fact involved the pharyngeal wall in the sub-glottic area and the patient was treated with radiotherapy and cytotoxic therapy, but a further biopsy confirmed Kaposi's sarcoma to be still active in this area in December 1970.

Gelfand in 1955 reported a case of Kaposi's sarcoma which was diagnosed at postmortem. Postmortem findings in this man revealed that he had a small fleshy tumour on the heart muscle and the histology was that of Kaposi's sarcoma.

Visceral presentations of Kaposi's sarcoma have included a young male who presented with an acute appendicitis and on histology the appendix was found to be involved by Kaposi's sarcoma and that further tumours had been noted at operation in the region of the terminal ileum. In a patient with a typical Kaposi's sarcoma of the leg hepatomegaly together with gynaecomastia led to the diagnosis of hepatic involvement by the tumour.

The so-called normal features in children is that of a lymphadenopathy but two adults who presented with inguinal adenopathy were on biopsy proved to be Kaposi's sarcoma involving the inguinal glands.

The incidental finding of Kaposi's sarcoma occurred in several cases. Three cases complained of cough and shortness of breath with chest pain, while one presented with a cataract. The incidental biopsies of various nodules found in these cases led to the diagnosis of Kaposi's sarcoma. In a diabetic who presented with nodules on the feet biopsy proved the nodules to be Kaposi's sarcoma. A male patient was admitted to hospital with a tentative diagnosis of malignant melanoma because he had a mass on the sole of the foot together with pulmonary metastasis on X-ray. Biopsy proved this to be a Kaposi's sarcoma. In the case of a man with persistent backache and pain in the leg, resistant to any form of therapy, diagnosis of Kaposi's sarcoma was made by biopsing a small verrucose growth and pain was

ascribed to involvement of the spinal cord by a Kaposi's metastasis. The puzzling onset of oedema in a middle aged man was finally ascribed to Kaposi's sarcoma when a very small suspicious area was biopsied.

X-RAY FINDINGS

Despite the work done by Palmer and his colleagues who describe various bony changes, routine X-ray of the cases seen in this hospital have produced in fact very few abnormal films. The hands and feet of all patients admitted were X-rayed for the so-called classical signs. In one case alone has there been any suggestion of the rubbing out of the head of the metacarpal bones and the phalanges or the metatarsals and phalanges. The more common sign in this hospital has been that of infiltration of bone in the formation of what appeared to be radiological bone cysts. These occur in the severe and fulminating forms of Kaposi's sarcoma in which there is an actual metastasis of the tumour into the bone, or pressure from the nodule outside causing a pressure erosion of bone. All cases have chest X-rays carried out on admission and in only three cases have pulmonary metastases been noted.

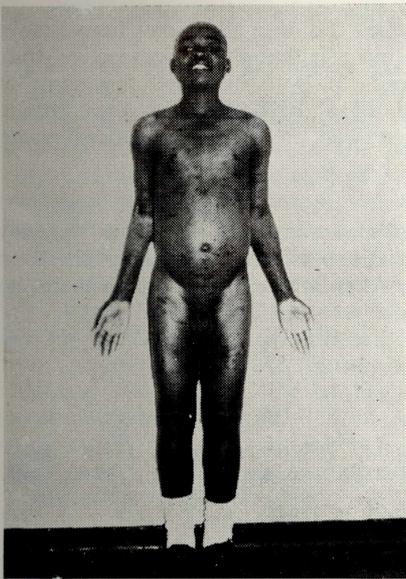


Fig. 7.—The same patient after initial Nitrogen Mustard therapy combined with Lasix showing early improvement with loss of some oedema but still illustrating the multiplicity of lesions.

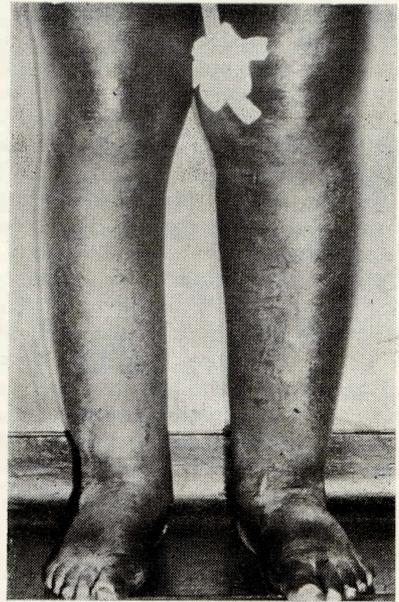


Fig. 8.—The legs of a middle aged African male showing bilateral lymphodema. Kaposi's sarcoma was detected when a small node at the site of the dressing was biopsied.

HAEMATOLOGICAL INVESTIGATIONS

There is no characteristic blood picture of a Kaposi's sarcoma apparent in this series. Many of the patients are anaemic and may show an iron deficiency type of anaemia. However, these findings compare with those in the rest of the population and there is nothing of particular note in the blood count. Eosinophilia has not been noted in these cases.

DISCUSSION

By far the most common presentation of Kaposi's sarcoma is the nodular form. This usually occurring in the middle-aged and elderly. It is obvious though that there are very many variations of this and the wider our experience the greater number of variations we are seeing. Basically, there is marked variation between the elderly and the young. In children the presentation usually resembles that of the lymphoma and the nodular form has in our experience only been seen once as described.

In the young adult the tumour is usually more aggressive and the presentation a florid one. The occurrence in women leads one to believe that there is a possible hormonal difference influencing the tumour in that the age group in the experience of all workers is usually lower than the average age

of presentation with Kaposi's sarcoma, and that the disease is aggressive and the normally rather slow progress is speeded up. It was therefore very interesting to see that the one case of florid presentation in a young male very closely resembled that, in fact, superseded that of the so called normal presentation in the female case.

The diagnosis of Kaposi's sarcoma is not a matter of a spot diagnosis. Although with experience the majority of the cases may be diagnosed clinically the disease closely resembles many other diseases and even clinicians with extremely wide experience may be badly caught out. This has been particularly true in our case with nodular leprosy. Other conditions that may closely resemble Kaposi's sarcoma are simple ganglion, angiomas, neuro-fibromatosis, melanoma, granuloma pyogenicum, lymphosarcoma and lymphangiosarcoma, onchocerciasis, filarial elephantiasis, maduromycosis, syphilis, sarcoidosis of the skin, and postmastectomy lymphangiosarcoma.

There has been a great deal of discussion on the association of Kaposi's sarcoma with other tumours and with other diseases (O'Brien & Brasfield, 1967). It is patently evident to us in this country that there is an association with tuberculosis. This association is probably on an immunological basis. As pointed out in this series of patients and in reviewing the known cases of Kaposi's sarcoma that have passed through this hospital, in a high percentage of cases there

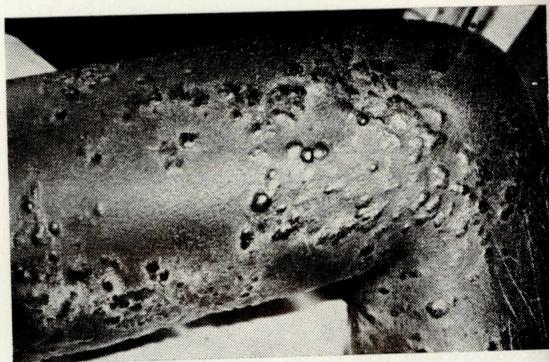


Fig. 10.—A fulminating case of Kaposi's sarcoma showing gross involvement of the thigh with severe oedema, plaques and nodules.

is an associated pulmonary tuberculosis. In view of the fact that there may be certain association with immunity inoculation of some of the tumours with vaccinia virus has produced regression of the surface portion of the Kaposi's nodule but the tumour in deep tissues has remained unchanged. Association with diseases such as diabetes is probably entirely fortuitous but Kaposi's sarcoma has been detected incidentally in cases with diabetes and in a patient with a cataract.

This intriguing tumour has in numerous presentations and it is becoming apparent that there are associations with other diseases. A rethink is probably due upon the etiology and presentation of this disease. To date all studies have been retrospective but it is time that a prospective study was introduced. It is to be noted that some of the "classical signs" of Kaposi's sarcoma, particularly those described on X-ray, are indeed rarities.

SUMMARY

Kaposi's sarcoma in its normal presentation and variations is described upon these presentations. Attention has been drawn to its importance as a research disease and that its progress is such that time is on the side of the research worker.

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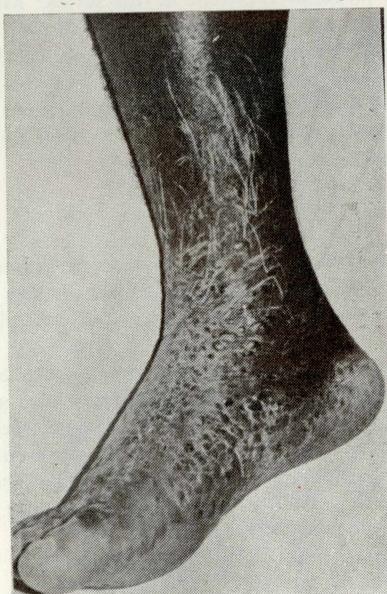


Fig. 9.—Numerous nodular lesions of Kaposi's sarcoma in the right foot of an African male with only moderate associated oedema.