

KAPOSI'S SARCOMA – Common but elusive cancer

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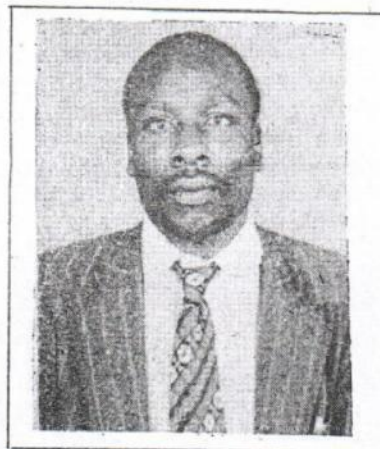
By Dr. Julius A. Ogeng'o

Kaposi's sarcoma was first described by Moritz Kaposi in 1872 among the Jews and Italians. Since then, it has been found in many parts of the world. In Africa, the first reports came from the West Coast in 1922. Following this, it has been described in many African countries. By 1950, evidence had accumulated to sub-Saharan Africa, accounting for 4% of all malignancies. Indeed, the highest incidence is in Zaire and Uganda where it is the fifth commonest malignancy. In the 1970s, it was found to occur more commonly among those individuals whose immunity was suppressed and in the western societies by 1981, it was found common among homosexuals who later turned out to have AIDS. In Kenya, Kaposi's Sarcoma is among the top ten cancers, accounting for 2-3 per cent of the total but with increasing numbers of AIDS patients, we are now seeing more and more of Kaposi's Sarcoma.

OCCURRENCE

Though all ages are susceptible, the sporadic form occurs in elderly men, with a peak around age 45-50 years. Children below 15 years, approximately twice as many females as males suffer the illness. However, above 15 years, in Kenya about thrice as many males suffer the illness.

The disease occurs more commonly in the low income groups who spend most of their time in the field



Dr. Ogeng'o: "In Kenya, Kaposi's sarcoma is commonest on the slopes of Mount Kenya among the Kikuyus and on the shores of Lake Victoria among the Luo."

with bare feet. Geographically, in Africa, the highest incidence appears to be in the countries along the equator. Indeed, gross figures indicate that warm, wet climate favours higher incidence.

Thus in Kenya, the tumor occurs on the slopes of mount Kenya among the Kikuyus and on the shores of lake Victoria among the Luo though other areas are not exempted especially with the onslaught of AIDS.

Though there is evidence for genetic predisposition, analysis in Uganda tribes implicate environmental rather than genetic factors.

More recently, the association of Kaposi's Sarcoma with lymphomas, leukaemia and AIDS have stimulated further research into the possibility of viral infections. So far it is not clear.

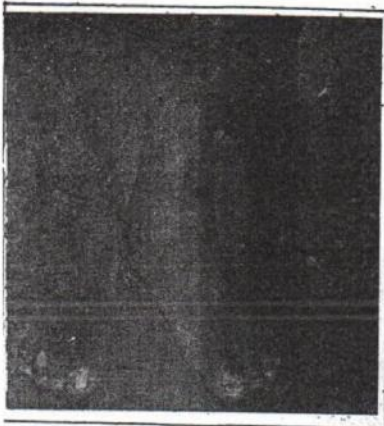
CAUSES

The final word about the causation of Kaposi's Sarcoma has not yet been said, neither can it be now. Familial, trauma - related, geographical, immunological and viral factors have all been implicated but it appears that the disease depends on a complex relationship between cancer causing agents yet unknown, acting on susceptible individuals.

PRESENTATION

Today, one must distinguish between the rapidly fatal aggressive kaposi's Sarcoma which is strongly associated with AIDS and the endemic variety in Africa. The latter is relatively better understood and in this article, I shall concentrate on it.

The commonest presentation is the nodular type. In most cases, the patient first experiences cold in the legs during or after rains. After a period varying from weeks to months, the leg swells. The swelling is initially soft and pits on pressure and is usually confined to below the knees. Two to three months later, little hard swellings appear, followed by hardening of the swollen leg till eventually the affected leg and foot becomes "woody". This progression occurs in about 12 months and if untreated affects the opposite leg in 6 - 12 months and in another 4 - 6 months, the arms and hands get affected one followed by the other in a matter of weeks. From this stage, the disease may progress to the rest of the body or remain confined to the extremities. This pattern of evolution



Swelling and infiltration of endemic Kaposi's Sarcoma. (Source: Surgery International Edition 2)

may however be influenced by the age and sex.

In women, the skin nodules tend to spread and involve the whole body in a short time. In young adults, 15 - 20 years, the disease often affects the alimentary canal. In this latter group, the patients survive and the skin nodules may show after the age of 30 years and then progress slowly, often allowing the patient to progress through life without further deterioration.

When it develops in children before the age of 10 years, it is almost always confined to the lymph nodes - in the neck, armpits, in the chest and sometimes generalised involving the intestines. In these cases it resembles other diseases that cause lymph node enlargement. At this age, and in this form if untreated, the outcome is poor, the victim dying within less than a year.

Kaposi's Sarcoma frequently affects internal organs. The commonest is the alimentary canal and the liver. When this occurs, the commonest presentation is internal bleeding with fluid accumulation, causing gross abdominal distension. Other internal organs likely to be affected are the lungs, the heart, blood vessels and even the brain.

Kaposi's Sarcoma may involve bones especially those of the foot, but other bones, for example, those of the hands, the long bones and ribs give rise to deep gnawing pain.

DIAGNOSIS

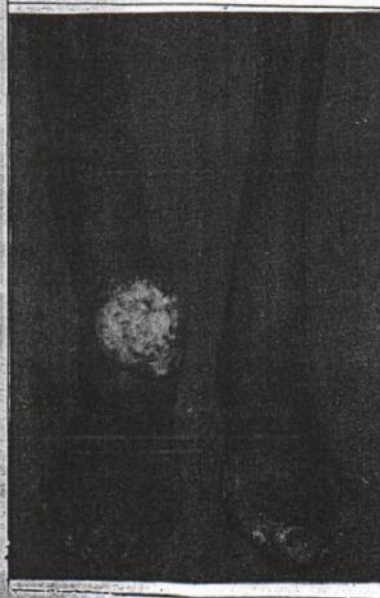
This is based on history, clinical examination and investigations such as specialised-rays of bones, the

blood vessels and the intestines. However, the most certain way is to take a small piece of suspected material for microscopic examination by a specialist.

Alternative Diagnosis

In children under the age of 10 years, the presence of enlarged lymph nodes may also be due to lymphomas, leukaemia, tuberculosis and bacterial infections among other things. In adults, though to the experienced eye the nodule is unmistakable, it can be confused for a jigger, fungal infections like "madura foot", simple wart or river blindness.

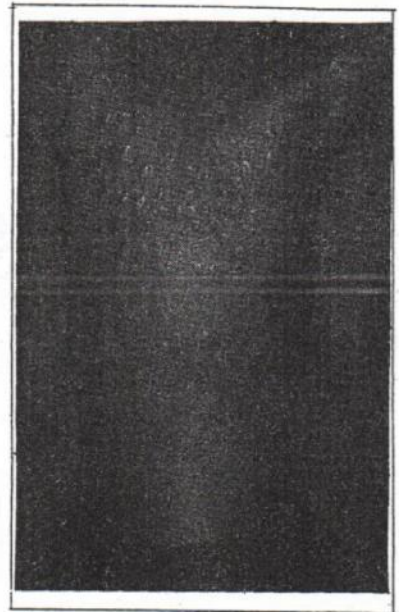
In cases which present as oedema especially in initial stages, it may be confused for renal, heart or liver disease or any other causes of oedema. However, the oedema of Kaposi's Sarcoma is always confined to below the knees, usually hardening within the year.



Florid Kaposi's Sarcoma. (Source: Surgery International Edition 2)

Treatment

Kaposi's Sarcoma is commonly a systematic illness and so treatment must be applied to the whole body. Of the three modes of treatment available, namely radiotherapy surgery and chemotherapy, the last is of first choice. It comprises of a combination of Vincristine and Actinomycin D, both fairly expensive.



Pigmented plaques of a typical African Kaposi's Sarcoma with Oedema of the trunk. (source: Surgery International Edition 2)

Outcome

When the tumor presents as nodular skin lesions in adults in its endemic form, the outcome is good. In fact cases of spontaneous recovery are known. In its epidemic form, when associated with AIDs, it runs an aggressive course and almost always kills. In children with lymph node involvement or in adults with florid or internal organ involvement, again the outcome is poor unless treatment is started early. The victims die from overwhelming disease or intercurrent infection resulting form depressed immunity and or anaemia. They may die also from complications of treatment or tumor inside the skull when this rarity occurs. Coexistence of Kaposi's sarcoma with lymphoma and leukaemia worsens the course and outcome.

However, with continuing research, it is hoped that better understanding of the disease and the nature of anti-cancer drugs will go a long way towards improving the outlook of Kaposi's Sarcoma. If you have features suggestive of the disease, consult your doctor now for it might be in the treatable stages or better still, it might be some other disease that is easy to treat.