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Endemic Kaposi's Sarcoma Vs. Kaposi's Sarcoma in AIDS : A Brief Communication

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ABSTRACT

Five cases of Kaposi's Sarcoma are presented to highlight the clinical course of the African or Endemic Kaposi's Sarcoma and point out the differences against the Epidemic form of Kaposi's Sarcoma common in AIDS patients in the United States and Europe. Groups at risk of developing the Epidemic form of Kaposi's Sarcoma are well identified in the United States whereas there is no special group at risk of developing the Endemic form in Africa. Genital Kaposi's Sarcoma of the Endemic form, a rare site for the disease is also presented. Three clinical and histological types of the endemic form are described.

Kaposi's Sarcoma was first reported by Dr. Moritz Kohn Kaposi in 1872. He described a disease that presented as multiple nodular tumors in the skin of hands and feet ranging in color from blue to purple[13]. These were usually small, reddish-purple, or hyperpigmented brown macules, papules, plaques, or nodules ranging in size from 2 mm to 2 cm. The disease is common among Ashkenazi Jews and people of Mediterranian origin especially men in their 6th or 7th decade of life[5]. This disease is also common and endemic in an area across Equatorial Africa where its relative frequency has been reported to vary from 9.1% in Zaire, 3.4% in Uganda and 4% in Tanzania[15]. In east Africa, three clinical forms of the disease are clearly defined: ① Nodular; in which males predominate and the prognosis is usually good. ② Aggressive; in which the tumor deeply infiltrates the surrounding tissues and ulceration is common. ③ Generalized; which is more common in females who usually present with visceral involvement, and in children in whom generalized lymphadenopathy is common. The generalized type has a poor prognosis[16,17]. In addition to sex, age and clinical types as prognostic indicators, anaplastic histology and impaired T-cell immunity show poor prognosis[17].

Recently in the United States and Europe, reports of an epidemic form of a widely disseminated and rapidly fatal Kaposi's Sarcoma have been published, which is seen mostly in active homosexual men ranging in age between 26 and 51[4,6,7,14]. These patients usually manifest a profound deficiency in cellular immunity, anergy to recall antigens on skin testing, lymphopenia and a reversal of the normal ratio of helper to suppressor/cytotoxic T-lymphocytes. This ratio, although not specific for this disorder, has become a marker for the acquired immunodeficiency syndrome (AIDS). They also suffer from a variety of viral, prostozoal, bacterial and fungal opportunistic infections[4,10,14]. The groups at risk of developing Kaposi's Sarcoma and AIDS now include homosexually active men, intravenous drug users, female prostitutes, Haitian

immigrants to the United states, and adult male hemophiliacs[10].

In Japan 2 suspected cases have been reported[1].

CASE PRESENTATIONS:

We present here five case summaries of patients with Kaposi's Sarcoma we experienced in 1980 at Muhimbili Medical Center, University of Dar-es-Salaam, Tanzania. No any of these patients had any history of neither homosexuality nor intravenous drug abuse. They highlight the typical features of the endemic form of the disease as against the Epidemic form as seen in the United States and Europe.

Case 1 (Fig. 1). 29 years old male presented with hardening and thickening of the skin and small nodular swellings in the lower legs, more on the right leg for the past 6 months. He had no other complaints. On examination he had scaly, non-tender hyperpigmented skin lesions with small nodules on the right lower leg. Routine laboratory investigations were normal. A biopsy showed Kaposi's Sarcoma. He responded well to treatment with Actinomycin-D and was discharged under Actinomycin-D follow up.

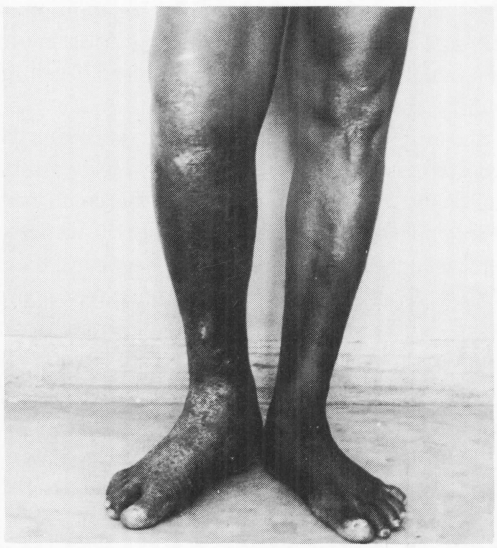
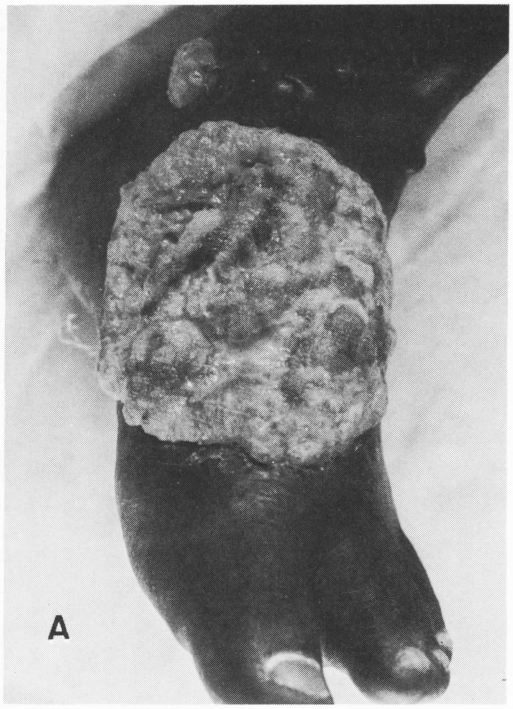


Fig. 1. A 29 year-old male with Endemic Kaposi's Sarcoma of the lower legs. It represents the nodular form of African Kaposi's Sarcoma that typically involves the extremities.

Case 2 (Fig. 2). 30 years old male came complaining of small hard swellings on the planter surface of the right foot for 1 year. He was otherwise in good health, and all laboratory investigations were normal. Skin biopsy revealed Kaposi's Sarcoma. Treatment was by Actinomycin-D and the response was a good one. Patient was discharged under Actinomycin-D follow up.

Fig. 2. A 30 year-old male with Endemic Kaposi's Sarcoma of the planter surface of the right foot. It represents the nodular form of African Kaposi's Sarcoma that typically involves the extremities.

Case 3 (Fig. 3A & 3B). 32 years old male gave a history of an ulcer and several small nodules over the left foot for 1 year. Physical examination showed a fungating ulcer with everted edges 3 × 2 cm (A). No bone involvement was found. Four other small nodules 0.5 to 1.5 cm were seen around the ulcer. No other significant findings were found from physical examination and all laboratory data were normal. Skin biopsy



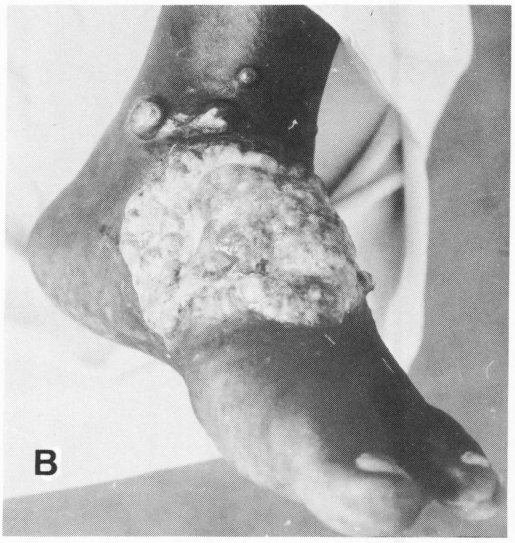


Fig. 3. (A and B).
A 32 year-old male with Endemic Kaposi's Sarcoma of the left foot. It represents the aggressive form of African Kaposi's Sarcoma. (A) shows the disease before treatment and (B) after initiation of treatment.

gave a histological diagnosis of Kaposi's Sarcoma. Administration of Actinomycin-D showed great improvement. Compare picture (A) before starting treatment and (B) after initiation of therapy. Patient continued to receiving treatment.

Case 4 (Fig. 4). 41 years old male came with a history of progressive multiple nodular swellings of the right foot of 18 months duration. On examination, the right foot was grossly swollen, several nodules were found three of which were ulcerated. There were no sinuses but an X-ray film of the lesion showed bone destruction. No other lesions or significant findings were detected. Kaposi's Sarcoma was the histological diagnosis after skin biopsy. An above knee amputation was done as treatment because the lesion showed poor response to Actinomycin-D and radiotherapy. Patient was discharged well.

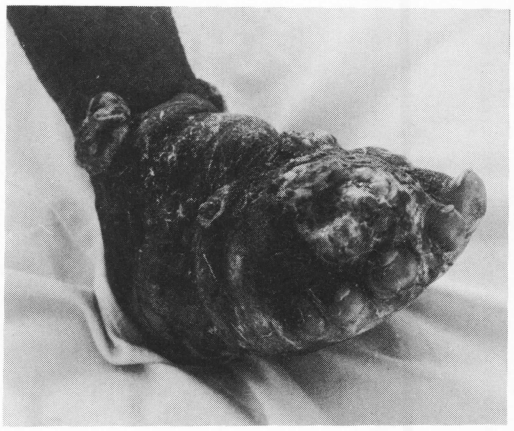


Fig. 4. A 41 year-old male with Endemic Kaposi's Sarcoma of the right foot. It represents the aggressive form of African Kaposi's Sarcoma.

Case 5 (Fig. 5). 34 years old male presented with a history of small nodules on the penis and pubic region accompanied with swelling of the shaft of the penis for 6 months. Examination showed edema of the penile shaft. Several small nodules, 1 — 5 mm, were seen at the base of the shaft and pubic region. The glans penis and sulcus region also had nodules. No lymphnodes were palpable. There were no other significant findings. Skin biopsy of the lesions gave a histological diagnosis of Kaposi's Sarcoma. Treatment by Actinomycin-D showed a poor response. Radi-

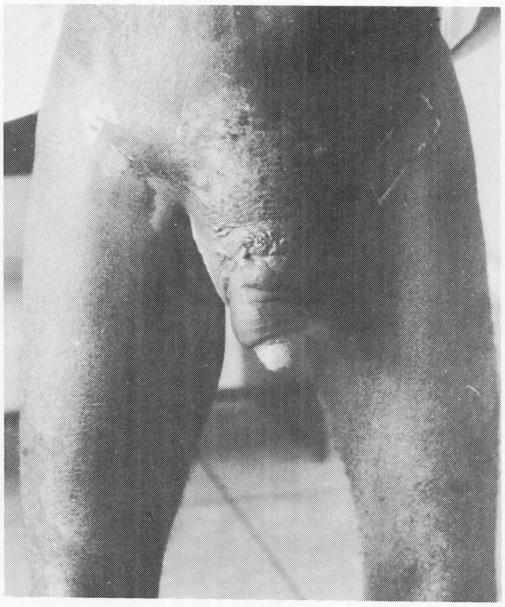


Fig. 5. A 34 year-old male with Endemic Kaposi's Sarcoma of the genitalia. This is a very rate site of the African Kaposi's Sarcoma and represents the nodular form.

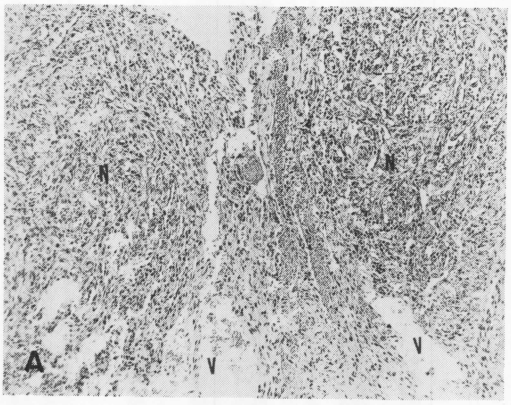
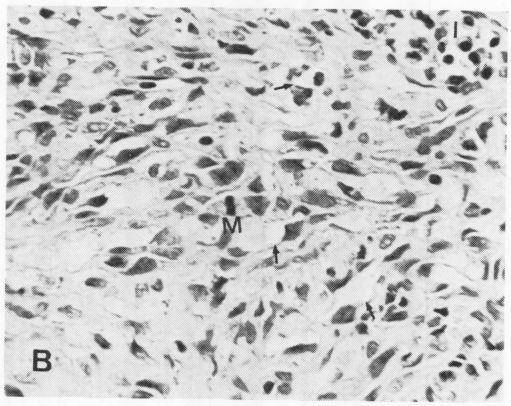


Fig. 6 (A and B)
(A) shows the histological appearance of the biopsy obtained from case No. 5 (Fig. 5). Nodular lesion N, surrounded by dilated sinusoidal vessels V, and hemorrhages are found. H and E × 100



(B) Higher magnification of (A). Irregular oval or spindle cells that partly line several vascular slits (arrow), some with mitosis M, are found. A few plasma cells and lymphocytes I, are associated. H and E × 470

otherapy was an effective treatment and patient was discharged under radiotherapy follow up. For comfirmatory purposes of this very rare case of the Endemic or African Kaposi's Sarcoma, description of the histological appearance of the biopsy obtained from this patients is given on Fig. 6 (A and B).

DISCUSSION

Cases 1 and 2 in our series represent the nodular form of African Kaposi's Sarcoma that typically involves the extremities, and has on excellent prognosis. Cases 2 and 4 represent the aggressive form of the disease in which local tissue destruction can be severe, but they still show a good response to appropriate treatment[18,20]. No example of the generalized form is seen in these cases. But case 5 is interesting in that the presenting site involved was the genitalia, a very rare site for Kaposi's Sarcoma. Of 624 cases seen in Uganda, only 4 had genital Kaposi's Sarcoma[16]. He had a nodular form of the disease with a good response to Radiotherapy. The mode of transmission of African Kaposi's Sarcoma is not very clear although an infectious etiology has been suggested[9,11]. Sexual transmission in African Kaposi's Sarcoma is unknown, and none of our patients was a homosexual. A sexual mode of transmission has been well documented in AIDS whose victims are homosexuals and among whom Kaposi's Sarcoma is common[5-7, 12]. Kaposi's Sarcoma in the genital area raises the possibility of sexual mode of transmission of endemic Kaposi's Sarcoma. Histology in Kaposi's Sarcoma is remarkable in that it is nearly constant in all three clinical forms. Three histological types

have been described: Mixed cellularity in which vascular slits are predominant, a monomorphic, and an anaplastic form which tends to have a poor prognosis. All our cases were of mixed cellularity type.

African or endemic Kaposi's Sarcoma is a disease entity in itself and patients with aggressive disease may show partial cellular immunodeficiency[17]. There is no group of people that is at special risk of developing Kaposi's Sarcoma although a geographic distribution has been reported[11]. Skin nodules on an extremity is the commonest site and there is a marked male preponderance. It has a broad spectrum of malignancy ranging from a prolonged indolent course to a highly malignant form seen in the generalized type mostly found in children and females[16,18,20]. On the other hand, the epidemic form of Kaposi's Sarcoma now seen in the United States is part of AIDS in which well recognized groups are at particular risk[2]. A severe defect in cellular immunity is the predominant factor[4-7,12]. The disease is usually generalized and opportunistic infections are frequent and as such it follows a fulminant clinical course[4,6,14].

Lymphadenopathy associated virus (LAV) and human T-cell leukemia virus type III (HTLV-III) were respectively identified by the Pasteur Research Institute of France and the National Institute of Health of the United States as AIDS-causing viruses[1,2,19]. They can be transmitted by close personal contact or injections of blood products. The immunodeficiency caused by these viruses in AIDS predisposes to repeated opportunistic infections, Kaposi's Sarcoma and other malignancies. However, in the African forms of Kaposi's Sarcoma, patients, show elevated serum antibody titres to cytomegalovirus (CMV) and herpes-type virus and the viruses have been isolated from tissue cultures of their Kaposi's tumor cells *in vitro*[3,8]. And more recently, antibodies to LAV virus have been detected in a few AIDS patients and healthy subjects from Africa[19]. The immunopathology of both the endemic and epidemic forms of Kaposi's Sarcoma seems to point to viral etiological role. Why the two forms of the disease are so different in their clinical course is as yet unclear.

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