

CASE REPORT


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DISSEMINATED ENDEMIC KAPOSI'S SARCOMA IN A YOUNG MAN WITHOUT EVIDENCE OF HIV INFECTION.

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ABSTRACT

We report a case of disseminated endemic Kaposi's sarcoma in an HIV- negative young man, involving the head, face, eye, tongue, upper and lower limbs, trunk, glans penis and peripheral lymphadenopathy. Tests for human immunodeficiency virus were negative. The clinical and histologic features with absence of immunosuppressive drugs in patient history confirmed the African endemic Kaposi's sarcoma. To the best of our knowledge this might be the first case reported in this part of the world.

KEYWORDS: Disseminated, Endemic, Kaposi's sarcoma, HIV-negative.

INTRODUCTION

In 1872, Moritz Kaposi, a Hungarian dermatologist, described odd skin tumors in five men in their sixth and seventh decades of life as "idiopathic multiple pigmented sarcoma of the skin (1)." There are five subtypes of Kaposi's sarcoma which include: 1) classic; 2) endemic African cutaneous; 3) endemic African lymphadenopathic; 4) acquired immunodeficiency virus-associated; and 5) those associated with immunosuppressive therapy. Although it is not understood, epidemiologic and biologic evidence has suggested that the pathogenesis of classic Kaposi's sarcoma involves the transmission of an infectious viral agent such as human herpes virus 8 or some other unique agent that is transmitted independently (2,3).

The most common form that is associated with Acquired Immunodeficiency Syndrome has been well documented since first described in the early 1980's (4). This form appears in up to 40% of AIDS patients and may account for up to 90% of all cancers found in the AIDS population (5). The African type, primarily

found in regions of equatorial Africa, is a much more common entity within its geographic setting, with findings of up to 9% of all malignancies in eastern Zaire (6). While usually less severe than the AIDS-associated condition, the African-type is the only form known to regularly occur in children (6). African-Endemic Kaposi sarcoma is the second most prevalent malignancy in Africa representing about 10% of all malignancies (7).

We report a young man with aggressive Kaposi sarcoma with no evidence of HIV infection.

CASE REPORT

A 30-year-old male farmer presented with a history of progressively asymptomatic multiple skin nodules on the left lower limb and face of 10 months duration. This is followed shortly by similar lesions on the ipsilateral leg and foot, forehead, face, right eye, right hand, glans penis and tongue. No associated itching or preceding history of trauma. No past history of immunosuppressive therapy. Significant peripheral

lymphadenopathy (cervical and inguinal), oedema of the left lower limb (fig.1 and 2) were observed. Chest, cardiovascular, abdominal and central nervous system examinations were essentially normal. A complete blood count and metabolic profile were within normal with the exception of mild normochromic anaemia

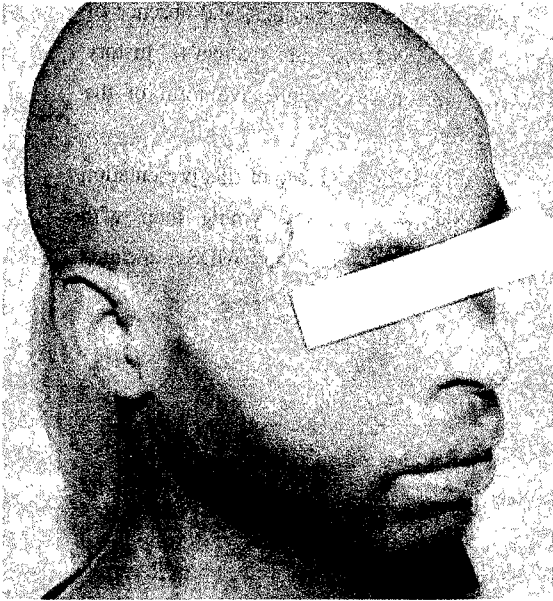
(Packed Cell Volume=30%), Scanning of abdomen and pelvis did not show any abnormality. Biopsy of one of the skin nodules (fig. 2) showed Kaposi's sarcoma (fig. 3). The patient was screened for HIV by the following tests. Table I.

Table I. Tests for HIV infection

KIT NAME	TYPES OF TEST	DESIGNED TO DETECT	RESULT
DETERMINE	RAPID IMMUNO- CHROMATOGRAPHY	ANTIBODIES TO HIV-1 AND HIV-2	NEGATIVE NEGATIVE
CAPILLUS	RAPID AGGLUTINATION	ANTIBODIES TO HIV-1 AND HIV-2	NEGATIVE
GENIE II	RAPID IMMUNO- CHROMATOGRAPHY	ANTIBODIES TO HIV-1 AND HIV-2	NEGATIVE
ORAQUICK	RAPID IMMUNO- CHROMATOGRAPHY	ANTIBODIES TO HIV-1	NEGATIVE
GENSCREEN	ENZYME - LINKED IMMUNOSORBENT ASSAY	ANTIGEN P24 OF HIV-1, ANTIBODIES TO HIV-1 GROUPS M AND O, AND ANTIBODIES TO HIV-2	NEGATIVE
NEW LOVBLOT 1	IMMUNOBLOT (WESTERN BLOT)	ANTIBODIES TO SPECIFIC HIV-1 ANTIGENS	NEGATIVE (NO VISIBLE BAND)
AMPLICOR HIV-1 MONITOR	NUCLEIC ACID AMPLIFICATION	HIV-1 RNA	NEGATIVE (VIRAL RNA NOT DETECTED)

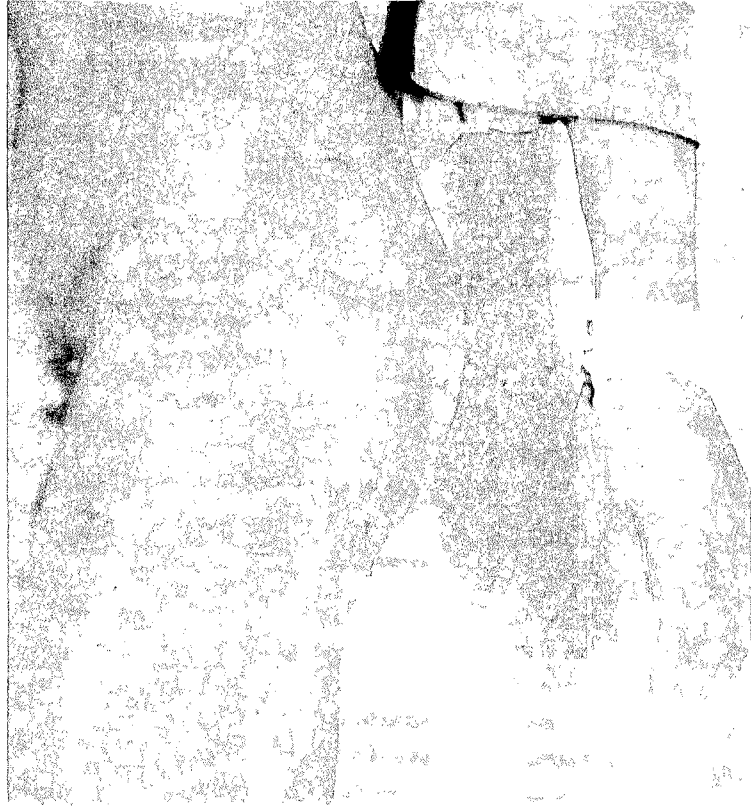
LEGEND 1

Photograph showing multiple nodules of Kaposi's sarcoma on the head and face.



LEGEND 2

Photograph showing multiple nodules of Kaposi's sarcoma on the left lower limb and severe oedema.



LEGEND 3

PHOTOMICROGRAPH OF KAPOSI'S SARCOMA. (H&E X40).



DISCUSSION

The pattern of AIDS-associated Kaposi sarcoma favours the lower extremities that are similar to the classical type. However, in our previous study, we reported aggressive KS in AIDS patients with multiple lesions on the body (8). This patient also presented with features of aggressive KS but was HIV-negative with the various tests of HIV at our center, table I Esu-Williams et al (9) carried out a study on the distribution of 330 seropositives patients and found 314 (95.2%) for HIV-1 group M, 1 (0.3%) for HIV-1 group O, 3 (0.9%) for HIV-2 and 12 (3.6%) for dual HIV-1 and HIV-2. This patient had similar tests to rule out HIV-1 group O that may be HIV negative by other routine tests.

The presentation of this patient is widespread Kaposi sarcoma involving the head and face, right eye (conjunctiva), tongue, right hand, left leg, glans penis and cervical and inguinal lymph nodes. The short course of this disease and wide spread of the lesions made it aggressive. In contrast to classic Kaposi sarcoma, which evolves very slowly and runs a benign course, even in the face of extracutaneous disease (10), the African endemic lymphadenopathic Kaposi sarcoma affects mainly children (6). This patient is a young man (30 years), which is also in contrast to the classic KS that was seen in elderly (older than 60years).

Kendrich et al (11) reported a similar case of widespread Kaposi's sarcoma in an elderly HIV-negative African-American. In the same center, Kagu et al (12) also reported a case of African Endemic Kaposi Sarcoma involving only the lower extremity in a 62-year-old farmer. Both types of African KS typically affect the lower extremities with the cutaneous type comprising 90% of cases (7).

The histological features of classic KS are not different from other forms of the disease(2). The short course of disease, and clinical features in our patient led to diagnosis of aggressive KS, with a negative HIV serology, normal laboratory findings, and absence of immunosuppressive drugs in patient's history confirmed the diagnosis of aggressive form of the disease.

In conclusion, the importance of this presentation is that of a negative-HIV young man with aggressive KS as seen in AIDS-associated Kaposi's sarcoma patients.

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