Classical Kaposi’s Sarcoma in a Nigerian Farmer: A Case Report

Nijeryalı Bir Çiftçide Klasik Kaposis Sarkomu: Vak’a Takdimi

Michael O. Iroezindu1, 2, Izuchukwu B. Achusi3
1Department of Medicine, University of Nigeria College of Medicine, Enugu, Nigeria
2Federal Medical Centre, Owerri, Imo State, Nigeria
3Department of Anatomic Pathology, Federal Staff Hospital, Abuja, Nigeria

Abstract

Classical Kaposi’s sarcoma (KS) typically affects elderly men of Mediterranean and Jewish origin. We present an unusual case of classical KS in a human immunodeficiency virus (HIV) negative elderly farmer from rural Nigeria. He had multiple brownish nodules and plaques on both lower extremities associated with lymphoedema. Histopathological examination of a biopsy of the skin nodule confirmed the diagnosis of KS. The lesions only showed marginal improvement on chemotherapy which necessitated a referral for radiotherapy. There is a need to look beyond the traditional geographical distinction of KS variants as we continue to experience a dynamic role for environmental co-factors in the development of KS.

Keywords: Classical, Kaposi’s sarcoma, Nigeria

Introduction

Kaposi’s sarcoma (KS) was first described in 1872 by Moritz Kaposi, a Hungarian dermatologist [1]. It is a spindle cell tumour of endothelial origin. There are four variants of the disease namely classical, African-endemic, Acquired Immune Deficiency Syndrome (AIDS)-associated (Epidemic) and immunosuppression-related (transplantation-related or iatrogenic) KS [2]. Before the era of human immunodeficiency virus (HIV)/AIDS, KS was a rare disease. Human herpes virus 8 (HHV-8), otherwise known as Kaposi’s sarcoma-associated herpesvirus (KSHV), has been identified in over 70-90% of all KS tumours which suggests a causative role [2].

Classical KS is rare in sub-Saharan Africa. It is usually seen in individuals of Mediterranean and Jewish origin [2, 3]. We report the case of an HIV-negative 85 year-old farmer from rural Nigeria with classical KS affecting the lower extremities.

Case Report

EG, an 85 year old hypertensive male farmer from a rural community in South East Nigeria presented with a 5 year history of progressive skin nodules and painful swelling of both lower limbs. He later developed weight loss and limitation of movement. Further systemic review was unremarkable. Physical examination revealed a chronically ill-looking elderly man with brownish skin nodules and plaques on both feet (Figure 1). There was bilateral non-pitting leg oedema. He had no mucosal lesions or peripheral lymphadenopathy. Systemic examination was normal. A working diagnosis of cutaneous Kaposi’s sarcoma was made.

Histopathological examination of a biopsy of the skin nodule confirmed the diagnosis of Kaposi’s sarcoma (Figures 2 and 3). HIV-1/2 screening was negative and fasting blood glucose was normal (92 mg/dL). Complete blood count was normal: haemoglobin-11.9g/dL; total white blood cells (WBC): 6,800/µL, differential- neutrophils 60%; lymphocytes 40%; platelets 297,000/µL. Chest X-ray showed features of hypertensive heart disease only. Other relevant investigation results were normal. Informed consent was obtained from the patient to report the case.

He received three cycles of chemotherapy using a combination of adriamycin (Adrim, Dabur Pharmaceuticals Ltd, New Delhi, India): 20 mg/m², vincristine (Cristovin, Ansell
Pharmaceuticals Ltd, Nairobi, Kenya): 1.4mg/m² and bleomycin (Bleomycin, Divya Enterprises, Maharashtra, India): 10IU/m². There was significant reduction in the lymphoedema and pain and he became ambulant. There was only marginal improvement in the lesions (Figure 4). He developed chemotherapy-induced pulmonary oedema after the third cycle which resolved following the treatment. He was counselled and referred for radiotherapy.

Discussion

There is paucity of information in the literature about classical KS in people of sub-Saharan African origin. Classical KS primarily affects elderly men of Mediterranean, Eastern European and Jewish origin. In a comprehensive report of classical KS in Italy, [3] the mean age was found to be 72 years. It is by far more common in men than in women, with a ratio as high as 10-15: 1 in some series [2]. Classical KS often presents as multiple, bilateral purple-blue or reddish-brown nodules and plaques on the extremities which may become confluent over time. In a recent study in Germany [4], lesions were localised to the lower limbs in 70% of patients. It progresses slowly and visceral involvement is uncommon [2]. Lymphoedema may precede or follow the appearance of visible cutaneous lesions [2]. The indolent nature of classical KS distinguishes it from African-endemic KS and AIDS-KS both of which tend to be more aggressive with frequent involvement of mucous membranes and viscera. In addition, individuals with African-endemic KS and AIDS-KS are usually younger compared to those with classical KS. In a study involving both...
African-endemic KS and AIDS-KS in Kenya, the predominant age group was 31-40 years [5]. In Uganda, 65% of persons with African-endemic KS were less than 55 years [6].

The age, sex and clinical presentation of our patient are typical of classical KS. He had no obvious cause of immunosuppression. The fact that our patient is of sub-Saharan African origin is rather unusual for classical KS. Nevertheless, occurrence of classical KS in non-Mediterranean countries such as Peru and China has been documented [7, 8]. The striking geographical variation in the incidence of classical KS and African-endemic KS points to a role for environmental factors. Despite the fact that HHV-8 has been clearly linked with all variants of KS, the prevalence of HHV-8 in the general population in sub-Saharan Africa is substantial at the rate of 18-71% [9]. In sub-Saharan Africa, HHV-8 prevalence increases steeply with age and is higher in males and in rural areas [9]. In Uganda, although 79% individuals with African-endemic KS were seropositive for HHV-8, the seroprevalence of HHV-8 in a large population of HIV negative controls with malignancies other than KS was found to be as high as 50% [6]. These observations suggest that other contributory factors influence progression to KS in HHV-8 infected populations. Ethnicity, raring of domestic animals, walking bare foot on a long-term basis, and significant alcohol use are some of the environmental factors that were associated more with KS than other cancers in Uganda [6]. Exposure to iron in volcanic soils has also been postulated to have a role in the development of KS [10]. We could not test for HHV-8 in our patient due to non-availability of facilities. As a farmer in a rural community, it stands to reason that he had some of the co-factors mentioned above but their relevance in the development of classical KS is still uncertain.

Various treatment options exist for KS and the choice largely depends on the variant, site and extent of the disease. They include surgical excision, intra-lesional biologics such as interferon alpha-2b, local or extended field radiotherapy, cryotherapy and systemic chemotherapy [2]. Radiotherapy has increasingly become the standard treatment for localized classical KS. Systemic chemotherapy is often considered to have limited benefits in classical KS and its use is often limited by poor tolerance of the drugs by the patients who are elderly as was the case in our patient. In the absence of radiotherapy and other options, our patient was offered chemotherapy as a bridge modality to alleviate his symptoms which was achieved before he was referred for radiotherapy.

In conclusion, we have presented a case of classical KS in an elderly farmer residing in rural Nigeria where classical KS is unusual. Clinicians involved in the management of HIV negative patients with KS may need to look beyond the traditional geographical distinction of KS variants as we continue to experience a dynamic role for environmental co-factors in the development of KS.

Ethics Committee Approval: Ethical approval was obtained from the Ethics Committee of the Federal Medical Centre, Owerri.

Informed Consent: Written informed consent was obtained from the patient/patients.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept - M.O.I.; Design - M.O.I., I.B.A.; Supervision - M.O.I., I.B.A.; Materials - M.O.I., I.B.A.; Data Collection and/or Processing - M.O.I., I.B.A.; Analysis and/or Interpretation - M.O.I., I.B.A.; Literature Review - M.O.I.; Writing - M.O.I.; Critical Review - M.O.I., I.B.A.; Other - M.O.I., I.B.A.

Acknowledgement: We would like to thank the house officers and resident doctors in the Infectious Diseases unit of the Department of Medicine, Federal Medical Centre Owerri for their assistance with patient management. The staff of the Medical Records Department of the Hospital is appreciated for their assistance with folder retrieval.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study has received no financial support.

References