INTRODUCTION

Kaposi Sarcoma (KS) was first described in 1872 by Dr. Moritz Kaposi (1837-1902), a Hungarian dermatologist.\(^1\) It is a malignancy of skin, mucous membranes and blood vessels. It has a known association with Human Immunodeficiency Virus (HIV). In fact, this tumour was the first clue to the diagnosis of Acquired Immunodeficiency Syndrome (AIDS) in the late 1970’s.\(^2\) Previously, it was known as a slowly progressive tumour, mostly seen on the lower extremities in the elderly men of Mediterranean or Jewish origin or in Africa.\(^3\) In contrast, the tumour occurring in the new clinical situation of AIDS had a more virulent progressive course.\(^6\) Viral etiology for this malignancy has been implicated and Human Herpes Virus-8 (HHV-8) has been shown to be the cause of both AIDS-related and non-epidemic cases.\(^7\) The endemic or classical KS is not associated with HIV and is infrequently seen in the north west of Pakistan and rarely reported by clinicians of different specialities.\(^9\)

Here, we present a case of non-HIV Kaposi sarcoma and discuss it in view of current available literature and our own past experience with a similar kind of malignancy.

CASE REPORT

A 45-year stout Afghan patient presented to our outpatient department at the Institute of Radiotherapy and Nuclear Medicine (IRNUM), Peshawar, with multiple subcutaneous nodules. The sites of involvement were the peri-orbital region, retro-auricular region, forearms, legs, chest and back. Oral mucosa was spared at the nodules. The patient had no visceromegaly at the time of presentation.

A biopsy specimen from the retro-auricular region revealed a KS with dermal lymphatic involvement. His serum was negative for the common types of viral infections including Human Immunodeficiency Virus (HIV) on routine serology. His total B-lymphocytes (CD 19\(^+\)), total T-lymphocyte (CD3\(^+\)), total CD4+ lymphocyte (CD3\(^+\), CD4\(^+\)) and total CD8\(^+\) (CD3\(^+\), CD8\(^+\)) counts were all normal or borderline high. The patient was under treatment with 3 weekly chemotherapeutic regimens of Adriamycin, Bleomycin, Vincristine (ABV) keeping in view socioeconomical constrains, logistical difficulties in getting proper medical care and side effects of other options like radiotherapy for extended surface areas.

Key words: Kaposi sarcoma. Non-HIV. Subcutaneous nodules.
Due to socioeconomic and logistic constraints, the patient was put on three drug chemotherapy including Adriamysin, Bleomysin and Vincristine (ABV) on standard doses according to body surface area calculations. The patient is under monthly follow-up and for the last 4 months his response to chemotherapy is adequate with a regression in the size and hardness of most of the nodules. No new nodules have appeared since the start of chemotherapy. He is tolerating chemotherapy well and has a satisfactory haematological profile.

DISCUSSION

Kaposi Sarcoma (KS), an apportunistic sequela of HIV infection, is considered as the common AIDS associated neoplasm. The epidemiology of AIDS associated KS has suggested that an environmental and/or infectious sexually transmitted co-factor might contribute to the development of KS. The search for such a co-factor led in 1994 to the discovery of a novel herpes virus, human herpes virus-8 (HHV-8), also known as the Kaposi sarcoma-associated herpes virus (KSHV). Non-HIV Kaposi sarcoma is a rare entity in this part of the world and clinicians are mostly not familiar with its presentation.

In the past 10 years, we have received 10 cases of KS with a more or less similar kind of clinical presentation. Extensive and multifocal skin involvement by pinkish red macules and papules of variable sizes and no laboratory evidence of HIV infection positively were the main features of the KS variant in this region. The interesting part of their presentation was the sparing of the oral mucosa which is usually seen in classic KS of other areas. Three of these patients were treated with ABV chemotherapy along with photons to more symptomatic sites. One was treated with an electron beam only for selected sites. All of these patients were Afghan in origin and due to the worsening Pak-Afghan border situation have been lost to follow-up.

AIDS has taught us many lessons about tumour pathogenesis and malignancies which have viral implications like KS; certain aggressive non-Hodgkin's Lymphoma (NHL) and cervical cancers have all frequently seen in AIDS.

Laboratory and epidemiological evidence suggest that HIV is not the only viral etiology involved but another virus KS-associated Herpes virus (KSHV) is also involved. As KS was more commonly seen in gay men as compared to HIV positive cases due to other routes like blood transfusion, factor-VIII infusion recipients and drug abusers, a different virus was incriminated and detected which was later named as KSHV. The facilities for detecting this virus were not available in the country, hence its detection was not possible. Rather, CD-4/CD-8 ratio was done which favoured a non-HIV origin.

The three drug chemotherapy in these cases has been found to be useful in disease stabilization and causes lesser toxicity. Detailed follow-up studies of different chemotherapeutic regimen outcomes are not available for non-AIDS Kaposi sarcoma. In AIDS related KS, ABV chemotherapy has documented evidence of efficacy. Liposomal Donoublastine, Paclitaxil has also been used in KS.

REFERENCES


