CASE REPORT

KAPOSI'S SARCOMA OF RARE ANATOMICAL SITE:

A report of two cases.

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SUMMARY
Kaposi's Sarcoma (KS) of unusual sites are commonly associated with immunodeficiency and it is therefore known as one of the AIDS defining tumors. KS of the conjunctiva and traumatized areas of the foot especially the sole are listed as some of the uncommon sites for this tumor.

One of the patients developed oral thrush (moniliaisis) at the age of 60 years while the other developed diarrhoeal disease and desquamative skin lesions. Both of them are positive to human immunodeficiency virus (HIV) screening (retroviral screening).
Key Words: **Kaposis Sarcoma, rare sites, immunodeficiency**

INTRODUCTION

Kaposis Sarcoma (KS) is a vascular neoplastic lesion consisting of idiopathic multiple pigmented Sarcoma of the skin.[1] The highest number of this tumor is reported in the Democratic republic of Congo in tropical Africa. In Nigeria, majority of the documented cases come from Cross Rivers and Akwa Ibom States.[2] The lesion predominantly affects elderly men of Jewish or Mediterranean discent and currently, it is the commonest AIDS (acquired Immunodeficiency Syndrome) defining tumor in patients with acquired immunodeficiency syndrome.[3] The commonest viruses associated with KS are the human herpes virus (HHV), HIV (human immunodeficient virus) I and II, human T-cell leukemic virus serotype III (HTLV III) and to a lesser extent, hepatitis B virus (HBV) and cytomegalo virus (CMV)[4].

In contrast to adult patients in whom the disease is predominantly cutaneous, among the pediatric patients with AIDs, the disease is primarily limited to the lymphadenopathic form.[5] The authors diagnosed two cases of KS of the conjunctiva and the sole of the foot which are rare anatomic sites for this tumor. We report two cases of biopsy proven dermatopathic and ocular KS to highlight these unusual sites coupled with its association with HIV positivity because, no report of such has been made in this environment to the best of our knownledge.

CASE – ONE: A 60 year old female presented in the Ophthalmology department of the University of Port Harcourt Teaching Hospital (UPTH) with 6 months history of weakness, weight loss and inability to see clearly with the right eye. She had been receiving treatment in a private hospital prior to presentation without relief. The eye lesion was diagnosed as pterigium in a chronically ill patient by the Ophthalmologists. Her past medical history revealed that, she had serious and persistent oral thrush which was difficult to treat. Surgery was done on the eye and the tissue was very hemorrhagic which is atypical of pterigium. The tissue was then sent for histological analysis.

Gross Findings: A fibromuscular and hemorrhagic soft tissue was received

measuring 1.5x1x0.6cm. It was pigmented and micronodular in appearance. All the tissue was submitted in a single block.

Histological Findings: Sections of the tissue showed dilated thin-walled vascular spaces with irregular outline, rimmed by flattened endothelial cells. There were collagen bundles and prominent spindle and fusiform cells between them and extravasated red blood cells as well as hemosiderin deposits. A diagnosis of Kaposi's sarcoma was made.

A request for retroviral screening was suggested and it turned out to be positive. Anti retroviral medication was started but the patient was unable to comply due to financial difficulties and she died four months after presentation.

CASE – TWO: A 52 year old Negroid male with multiple pigmented nodules on the lower limbs was referred from a private clinic to University of Port Harcourt Teaching Hospital for management. He was observed to be wasted and weak. He complained of an itchy lesion in the sole of the left foot of three months duration. A working diagnosis of Melanocarcinoma in a patient with neurofibromatosis was made. The surgical and medical teams were consulted. Biopsy of the lesion on the sole of the foot was taken and sent for histologic study along with other base line investigations.

Gross Finding: The specimen consisted of an irregular brownish tissue measuring 3x2x1.8cm. The cut surface showed hemorrhagic tissue that was punctuated by grayish foci. The tissue was submitted in two blocks.

Histological Findings: Section of the tissue showed bands of spindle cells embedded in a network of reticular fibres and a maze of vascular spaces. There were numerous abnormal mitotic figures with extravasated red blood cells and hemosiderin landden macrophages. There was infiltration of the stroma by chronic inflammatory cells. A diagnosis of Kaposi's Sarcoma was made.

A request for retroviral screening was suggested. The result came out to be HIV positive. This findings prompted the review of his past medical history of which the patient had prolonged diarrhoeal illness along with desquamative dermatitis. The patient was placed on anti-retroviral regimen but he signed against medical advice and left the hospital.

DISCUSSION

Kaposi's Sarcoma usually affects the lower limbs, the mucus membrane of the mouth, anus and the intestine where it presents as a nodular, pigmented lesion that bleeds easily. It is slightly more frequent among males[2,5] and is usually accompanied by diarrhoea, chronic fungal and other opportunistic infections when associated with immuno deficiency.[6,7] It also occurs in odd sites such as the penis, conjunctiva, tip of the nose and areas of previous trauma[7] which our patients presented.

KS exists in four basic forms made up of the classic, endemic, iatrogenic and AIDS related types.[3] These types are made up of the mixed cellular, anaplastic or pleomorphic variant which are commoner in adults and the forth, the lymphadenopathic variant is known to be commoner in children.[8] KS in what ever form is believed to arise from vascular endoethlium based on histochemical and ultrastructural evidence in a manner similar to other vascular tumours.[5,7,8] These types vary in clinical features but in either cases, the histologic patterns are similar; implying that, host factors are determinants of the clinical behavior of the tumor.[7]

The classic form is common in the mediteranean area and Africa, its association with AIDS has increase with more patients, widespread distribution, variety of clinical manifestation and a more aggressive course[5,7] while the endemic and the sporadic form followed a more protracted benign clinical course.[9] KS is the most frequent clinically defined tumor in AIDS patients[10] and one of the classic manifestations of AIDS since it is found in nearly a third of the adult patients with AIDS in USA.[6] As in our patients, 95% of adult patients with AIDS and KS presented with visible cutaneous or oral lesions.[6,7]

Before the advent of HIV and AIDS, KS was well known in Central and East Africa which virtually posed no problems.[2] One of our patients died because she lacked funds for drugs treatment. This is also cited by the WHO[11]. The other problem is the social stigma attached to HIV and AIDS. These people are often ostracized by family members and the society. They are usually treated as outcasts and can be evicted from their homes, dismissed from work, be avoided or even refused medical attensions.[12] This was probably the reason why the second case sign against medical advice after he was informed of HIV status.

The felling of guilt, regret and sorrow are escalated because of the shame on them and family. As a result, denial syndrome develops. It is therefore necessary to mount a public awareness campaign through different media about HIV and AIDS and measures to control it. The government should also see to the high cost of retroviral drugs and advice means of making them available to victims of this scourge. The HIV and AIDS patients have the right to exist as any other person by law. Some of them have taken to the option of committing suicide. This is because of lack of awareness and enlightenment of the public. However, many non-governmental organizations are springing up to meet the challenges posed by this raging pandemic. They should be encouraged to work in partnership with the Federal organs set up to specifically tackle this all important problem.

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