Orbital Kaposi Sarcoma in a Female HIV-Seronegative Nigerian and a Review of Literature

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Introduction: Kaposi Sarcoma, first described by the Hungarian Dermatologist, Moritz Kaposi in 1872[1] is a rare, multifocal, vascular lesion of low-grade malignant potential typically presenting in muco-cutaneous sites such as the skin of the lower extremities, face, trunk, genitalia and oropharyngeal mucosa[2]. The tumour became more common following the spread of Human Immuno-deficiency virus /Acquired immune-deficiency virus syndrome (HIV/AIDS)[3,4,5]. It is usually associated with Herpes virus 8 (HHV8) infection and the eye is an unusual location[2]. The commonly reported ocular locations are the eyelids, bulbar conjunctiva and lacrimal gland[2,6]. An orbital location of this tumour is rare and warrants reporting[3]. We report the case of a HIV-seronegative woman with orbital Kaposi sarcoma. Case Report: A 50-year-old female farmer from South-South Nigeria, presented to our clinic with a 2-year history of painless, progressive swelling of the left eye associated with redness, and total loss of vision in eye 10 months prior to presentation. On examination, the right eye was essentially normal with a visual acuity (VA) of 6/9 while the left eye had no light perception. There was a non-axial proptosis and slight medial displacement of the left globe which was embedded in a huge orbital mass measuring about 60x70x40mm.(Figure 1). There was no evidence of local spread beyond the left orbit and other systems were essentially normal. A cranial computed tomography scan showed an extensive left orbital mass without calcifications or areas of lucency, it was mildly enhancing and isodense, causing antero-medial displacement of the globe with associated expansion of the orbit without gross intracranial extension [Figure 2]. An initial diagnosis of orbital lymphoma to rule out

Fig. 1

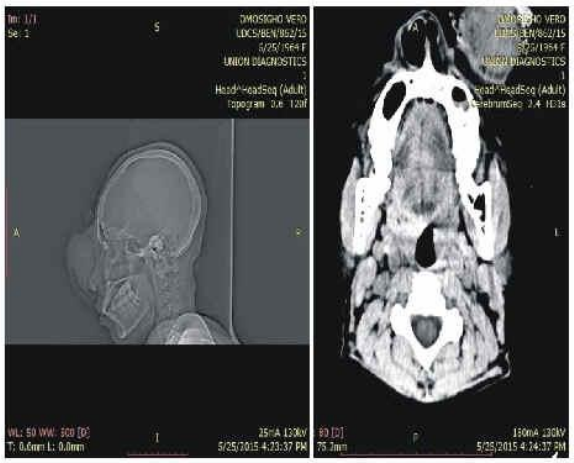

Fig. 2

pleomorphic adenoma of the lacrimal gland was made. All heamatological tests done were normal, fasting blood sugar was 82mmg/dl and she was RVS negative to HIV I and II antibodies (ELISA test) on repeated testing. She had an incisional biopsy of the orbital tissue and had marked intra-operative haemorrhage with an estimated blood loss of 500mls. Histopathological examination of the tissue biopsy showed features in keeping with Kaposi sarcoma [Figure 3]. She was planned for External Beam Radiotherapy to the left eye 40Gy in 20 fractions over 4 weeks using a direct field and 6 courses of combination chemotherapy

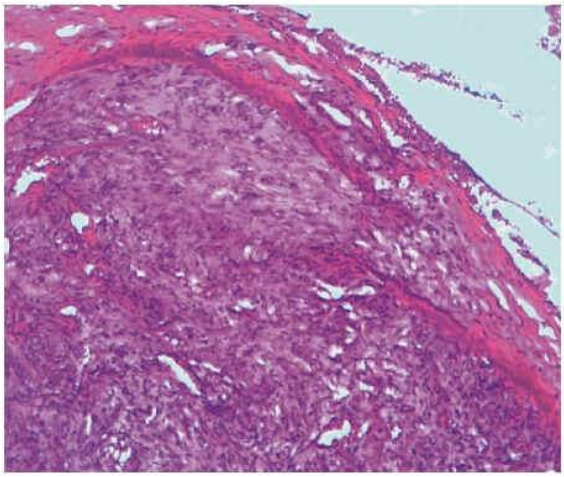

Fig. 3

SPECIMEN: *Received a soft, irregular grey brown tissue that measures 4x3.5x3cm and weighted 22g. Cut sections show tan brown microcystic appeareances. PEx4*

MICROSCOPIC DESCRIPTION: *Sections shows a fairly encapsulated lesion composed of mostly capillary-sized vascular channels containing red blood cells with foci showing dilated vascular channels at the periphery of the lesion. The stroma is fibrous and infiltrated by mononuclear cells. Other areas shows lobules of mature adipocytes and haemorrhage.*

comprising Intravenous Bleomycin30IU, Vinblastine 10mg, Doxorubicin 50mg. She only received 2 courses of chemotherapy due to financial constraints and 8 fractions of External Beam Radiotherapy before the equipment broke down, after which she was referred to continue treatment at the only center in the country with a functional machine at the time. She was unable to do so on account of lack of funds and logisticconstraints. She re-presented a year later and examination at this last visit showed the left orbital tumor had increased in size with associated fungation. The patient was still constitutionally stable and a repeat retro-viral screen was negative. All efforts are being made to assist her through social channels..

Discussion and Literature Review: There are four variants of KS: classical, African (endemic), immunosuppression (transplant)-associated and AIDS-associated KS. Classical KS tends to be indolent, while the African- and AIDS-KS are more aggressive with frequent involvement of mucous membranes and viscera[4,6,8,9]. Risk factors associated with classic KS include topical steroid use, trauma, infrequent bathing, asthma, and a history of allergy in men[4,5]. Our patient's disease ran an indolent course, was localized to the orbit and most likely to be the classic type. Although there is no definitive cure, lesions of Kaposi sarcoma are highly radiosensitive, the treatment is well tolerated and temporarily controls large localized lesions[10,11]. The tendency toward multimodality makes radiation therapy or chemotherapy, or both the preferred mode of treatment[11,12].

Conclusion and Recommendations: Orbital occurrence of KS is rare. Although there is no definitive cure, classical KS istreatable with timely diagnosis and treatment.Costs and availability of treatments especially radiotherapy were the problems our patient had but these can largely be solved with an efficient, well funded health system and better coverage of health insurance for all.

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