

Kaposi Sarcoma in Non-immunocompromised Adult

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Fig. 1: Swelling and redness over both peri-orbital

A 27 year old female patient, with no known co-morbidities or drug allergies presented to us with complaints of painful swelling and redness over both peri-orbital regions, of insidious onset, gradually progressive for past 6 months. Initially, unilateral it progressed to other side with associated peri-orbital redness and swelling.

There were no associated eye discharge, epiphora or bleeding suggestive of nasolacrimal obstruction. However, vision was compromised later on as a result of severe peri-orbital swelling (Figure 1).

She was administered antibiotics, local application and systemic steroids, and other Intra-lesional injections which were largely unsuccessful. As the symptoms of peri-orbital swelling increased, pain and color of lesion changed to black, and skin dryness with scaling appeared and increased progressively, she was brought to our hospital.

Evaluation for Hepatitis B, C and HIV was negative. Cultures of the blood, urine and swab culture from the wound were sterile. Swabs taken from the skin lesions around



Fig. 2: Substantial reduction in lesions after 6 months

the eyes were unproductive for any infective pathology on Gram stain and fungal stain. MRI of the face revealed diffuse and extensive skin, subcutaneous and soft tissue thickening and edema involving the scalp and soft tissues of face and neck and features consistent with angioedema. Globes and intra-orbital structures were normal. Work-up for vasculitis was negative. A skin biopsy was planned to ascertain the diagnosis. Skin biopsy was consistent with a diagnosis of atypical vascular proliferative lesion, low to intermediate grade, most likely Kaposi sarcoma (KS).

KS is a multifocal, vascular lesion of low-grade malignant potential that presents most frequently in mucocutaneous sites and commonly involves lymph nodes and visceral organs. The musculoskeletal system, central and peripheral nervous system, larynx, eye, major salivary glands,

endocrine organs, heart, thoracic duct, urinary system and breast represent unusual locations for the development of KS¹. Peri-orbital edema may occur with KS of the face.^{2,3} KS of the conjunctiva and ocular adnexa has been reported in association with Classic and AIDS-related KS.⁴⁻⁷

In view of the diagnosis Oncology opinion was requested and due obvious cosmetic disfigurement and widespread facial disease, our patient was planned for treatment with Pegylated Liposomal Doxorubicin (PLD). She received 6 cycles of PLD over a period of 6 weeks and responded very well to the treatment. Her lesions subsided substantially and vision also improved with treatment (Figure 2).

This case represents an unusual location for the development of KS in an immune competent host. It also stresses on the value of suspecting KS at atypical sites and also the importance of systemic therapy for KS when site and extent of the disease involvement make local treatments rather less attractive options.

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