Case Report

Kaposi Sarcoma in HIV negative patient - A case report and review of literature

Prashanthi Nemani[1*], Abdul Rehman Siddiqui[2], Lakshmi S Rao[3], Erella Madhusudan Rao[4], Tanmay Tapasee Praveen[5], Chinmaya Ranjan Behera[6]

[1, 2, 3, 4, 5]Post Graduate Trainee, [6]Associate Professor
Department of General surgery Kalinga Institute of Medical Sciences, Bhubaneswar, Odisha, India
[*]Corresponding author email: **prashanthi2292@gmail.com**

International Archives of Integrated Medicine, Vol. 8, Issue 9, September, 2021.
Available online at http://iaimjournal.com/
ISSN: 2394-0026 (P) **ISSN: 2394-0034 (O)**
Received on: 05-09-2021 **Accepted on:** 13-09-2021
Source of support: Nil **Conflict of interest:** None declared.

How to cite this article: Prashanthi Nemani, Abdul Rehman Siddiqui, Lakshmi S Rao, Erella Madhusudan Rao, Tanmay Tapasee Praveen, Chinmaya Ranjan Behera. Kaposi Sarcoma in HIV negative patient - A case report and review of literature. IAIM, 2021; 8(9): 71-74.

Abstract

Kaposi sarcoma is angioproliferative malignancy affecting skin and internal organs. This is common in advanced stages of HIV infection. Incidence of Kaposi Sarcoma in HIV negative patients is rare with indolent course, varying from small cutaneous lesion to wide spread lesion involving internal organs. The current article is a case report and review literature of Kaposi sarcoma. A 21 year old male presented to OPD with a 5 month old chronic wound involving left foot and ankle with a tendency to bleed. Biopsy revealed Kaposi sarcoma and serology negative for HIV. Patient was treated with below knee amputation and one and half year followup showed no recurrence.

Key words

Kaposi Sarcoma, Non HIV patient, Clinical features and management.

Introduction

Kaposi sarcoma is index for suspecting AIDS/HIV infection or immune-compromised state. But its incidence in immune-competent individual is rare, which limits the adequate understanding on etiopathogenesis and treatment modalities. Kaposi Sarcoma was first described in 1872 by the Hungarian dermatologist Moritz Kaposi. It is common in Mediterranean and Eastern European population. Later its incidence increased in immune-compromised patients. Four variants of Kaposi sarcoma has been described

based on clinical features and epidemiological distribution. A close association of HHV8 (Human Herpes virus) in etiopathogenesis of Kaposi sarcoma has been studied which helps in developing new treatment modalities.

Case report

A 21 year male presented to surgery OPD with complaints of wound in the left ankle and foot for past 5 months. It started with a trivial injury over left foot which was accompanied by a swelling and gradually progressed to a big ulcer involving the sole, ankle and the lower leg (**Photograph – 1**). Patient was receiving regular dressings at a local hospital with minimal relief. Later the wound started bleeding, initially small, and then progressing to profuse bleeding for 2 days, warranting a referral to our tertiary centre where biopsy revealed Kaposi Sarcoma. Patient had no other symptoms or lesions in the body. Routine investigation revealed mild anemia and serology negative for HIV. Patient was managed by below knee amputation. Post-operative period was uneventful and with no recurrence in next one and half year follow up. At present the patient is doing better with prosthesis.

Photograph – 1: Ulcer on lower limb.

Discussion

Kaposi sarcoma is neoplasm of lymphatic endothelial cells that usually indicates underlying immune-compromised conditions like AIDS, post transplantation immune suppression, but rarely seen in immune-competent individuals. HHV-8 (human herpes virus) is commonly associated with Kaposi sarcoma. Based on clinical and epidemiological features there are 4 variants [1].

Classical Kaposi sarcoma is most common in Mediterranean and Eastern European regions with varying incidence. Usually presents in elderly population in 5^{th} to 6^{th} decade of their life with male preponderance (10:1), not associated with HIV [1, 2]. Clinically presents with macules, plaques and nodules in the lower extremity (foot and ankle) and have chronic course with slow progression. They rarely metastasize. Risk factors include male gender, and steroids use. Prognosis is good when detected early [1].

Iatrogenic Kaposi Sarcoma in post transplantation immune suppression presents with cutaneous and extra cutaneous lesions. Has very good prognosis once medications are modified. Endemic KS is seen in Central Africa, presents commonly as extra-cutaneous lesions. Prognosis is good in early stages just as with classical Kaposi Sarcoma [1]. Epidemic KS seen in HIV positive individuals, both homosexual and heterosexual patients. They present both as multifocal extensive cutaneous and extra-cutaneous lesions of GIT, lymph nodes, liver, spleen and Lungs. It is an aggressive disease with poor prognosis.

Etiopathogenesis of Kaposi sarcoma is closely associated with HHV 8. HHV 8 virus is transmitted through saliva and sexual route. According to literature (KSHV) Kaposi sarcoma herpes virus acts on multiple cellular pathways like increasing cellular proliferation by promoting chromosomal instability, uncontrolled cell division, inhibiting apoptosis (increase anti-apoptotic proteins, inhibit p53 and Rb genes), inducing growth factors, cytokines and chemokine production, along with angiogenesis, lymphocytic infiltration. There is a strong correlation between viral load and tumor progression [1]. Mediterranean, Eastern Europe, Africa, Jewish region males are predominantly affected with classical Kaposi Sarcoma [1].

Corticosteroids and Diabetes are associated risk factors.

Classical KS includes 4 stages. I. Maculo-nodular, II. Infiltrative, III. Florid, IV. Disseminated. Stage I, II have more of cutaneous involvement with slow progression. Stage I and II are slow growing with cutaneous lesions. Stage III, IV have visceral involvement like oral cavity pharynx, GIT, bone marrow lymph nodes, spleen, liver. Stage III and IV have rapid progression that is rapid increase in number of Plaque and nodules or increase in total area of lesion. Complications of KS include pain, ulceration, hemorrhage, lymphedema, lymphorrhea, functional impairment [1].

Treatment modality is different for AIDs associated Kaposi sarcoma and HIV negative Kaposi sarcoma as course of disease is different. The management in HIV negative KS depends on the stages.

In stages I and II which is slowly evolving, different therapeutic strategies are opted, depending on the features of KS like clinical monitoring, surgical excision, intra-lesional chemotherapy with Vincristine, radiotherapy, and supportive measures like elastic stockings for the prevention of lymphedema.

In our case which is locally aggressive Classical Kaposi sarcoma of the Ankle involving distal leg without any other lesion on skin and viscera, below knee amputation was done. Biopsy revealed stage 2 classical Kaposi sarcoma with margins free from tumor. Follow up for next 1 year showed no skin lesion or visceral involvement, hence No chemo or radiotherapy was advised.

For rapid tumor evolution or complications or advanced stages (III and IV)
First line medications include different multidrug regimen like

(i) Adriamycin, Bleomycin plus Vincristine or Vinblastine,
(ii) Etoposide, Vinblastine and Bleomycin,
(iii) Pegulated liposomal Doxorubicin or Daunorubicin, Interferon-α2b.

Second line medications include Taxanes (Paclitaxel and Docetaxel), Pegulated liposomal Doxorubicin or Daunorubicin [1, 2, 3].

Systemic chemotherapy is considered as a standard approach and has better result in terms progression free interval and reduced remission. This includes therapy with Interferon, PEG-interferon and standard chemotherapy (bleomycin, anthracyclines, etoposide, vinca alkaloids, paclitaxel, liposomal doxorubicin or daunorubicin). Additionally drug groups include Taxanes (paclitaxel), pegylated liposomal doxorubicin, Antivirals (gancyclovir, cidofovir), Thalidomide. Recent new treatment modality under trials include Interferon (most promising in long term treatment), VEGF inhibitor (pentosan, tecogalan), Thyrosin kinase inhibitors (imatinib), matrix metalloproteases, mamalian target or rapamycin inhibitors [1, 2, 4].

Conclusion

Kaposi Sarcoma is a common neoplasm in AIDS and has an aggressive course of disease. It is rare in HIV negative or immune-competent individuals. High index of suspicion and Biopsy proven diagnosis is required for early diagnosis. The prognosis in these patients is good with complete remission if detected early and treated accordingly. Many Systemic treatment modalities targeting the pathogenesis of the disease are still under trial and hold a potential to deliver promising results for these patients.

References

1. Akasbi Y, Awada A, Arifi S, Mellas N, El Mesbahi O. Titre du document ici. Bulletin du Cancer, 2012 Oct 1; 99(10): 92-9.
2. Jan RA, Koul PA, Ahmed M, Shah S, Mufti SA, War FA. Kaposi Sarcoma in a Non HIV Patient. Int J Health Sci (Qassim)., 2008; 2(2): 153-156.

3. Ozmen H, Baba D, Kacagan C, Kayikci A, Cam K. Case report: HIV negative isolated scrotal Kaposi's sarcoma. International journal of surgery case reports, 2014 Jan 1; 5(12): 1086-7.
4. Jakob L, Metzler G, Chen KM, Garbe C. Non-AIDS associated Kaposi's sarcoma: clinical features and treatment outcome. PLoS One, 2011; 6(4): e18397. Published 2011 Apr 12. doi:10.1371/journal.pone.0018397