

# Kaposi Sarcoma with Visceral Involvement in a HIV- Neg Female Patient

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**Abstract:** *Background:* Kaposi sarcoma is a rare multifocal malignancy of hemo-lymphatic tissue that ,its precise etiology is unknown .There are reports showing the role for an infective viral agent. In HIV disease ,its prevalence is far more common as well in immune- suppressed patients it is more prevalent.

*Report of the case:* We presented a 78 year- old woman with multiple tender purplish plaques on her extremities for a month. In her past history she was healthy until 2 years ago when she noted two plaques on her left foot and a similar one on her right hand. In skin biopsies, there were spindle shaped cells presented with hypervascularity consistent with Kaposi's sarcoma.She had refused any kind of treatment . No positive signs were found in physical examination.

Routine laboratory data was normal. Anti-HIV ab test was negative although she showed significantly lower levels of CD4+ T- lymphocytes in her blood profile. Thoraco-abdominal CT demonstrated lesions highly suggestive of hepato-splenic involvement as multiple hypodense nodules.

Because of patient's disagreement and the reluctance of GI surgeons to remove the masse, we refused to refer her for invasive procedures .

This is the only kaposi sarcoma female patient in our department since 10 years ago.

## INTRODUCTION

Kaposi sarcoma is a multicentric, malignant neoplastic vascular proliferation which is characterized by the development of bluish-red cutaneous nodules, usually on the lower extremities, most often on the toes or feet, and slowly increasing in size and number and spreading to more proximal areas [1]. Kaposi sarcoma (KS) is the most frequent neoplasm found in AIDS patients. The disease preferentially involves the skin and the lymphatic and digestive systems. KS is rarely the cause of death in these patients, although it can cause significant morbidity [2].

Four forms of KS are known: classic or Mediterranean, endemic or African, post transplant, and epidemic or acquired immunodeficiency syndrome-associated KS [3]. The tumors often remain confined to the skin and subcutaneous tissue, but widespread visceral involvement may occur. Kaposi's sarcoma occurs spontaneously in Jewish and Italian males in Europe and the United States. An aggressive variant in young children is endemic in some areas of Africa. Endemic KS (EKS) is the most common entity in tropical Africa [4]

A third form occurs in about 0.04% of kidney transplant patients. It has perhaps viral etiology ,HHV-8 is the suspected cause [5].

## REPORT OF THE CASE

Here,we presented an 78 year- old Iranian woman with multiple pruritic , tender purplish blue nodules and plaques on her feet and hands (Figs. 1 & 2) from one month ago.In her past medical history she was healthy until 2 years ago when she had been noted of two purple plaques appeared on her left foot and a similar plaque on the dorsum of her right hand .On two consecutive skin biopsies from these plaques, there were spindle shaped cells presented with extensive hypervascularity of dermis and an inflammatory infiltrate consistent with Kaposi's sarcoma(H&E stain magnification X 40) (Fig. 3). The patient had no symptoms except for itching and pain in the plaques. She did not agree with any kind of treatment though the previous physicians suggested radiotherapy for her lesions .In physical examination ,no positive signs (organomegaly or lymphadenopathy...) were detected.

Routine laboratory data was normal.Occult blood (Guaiac test) and anti-HIV Abs were all negative.Chest X ray and CT scan were normal but she had significantly lower levels of CD4+ T- lymphocytes and a low CD4:CD8 ratio in her peripheral blood profile. The thoracoabdominal CT scan revealed lesions as multiple hypodense nodules ,highly suggestive of hepato- splenic involvement by KS.

Because of the patient disagreement and old age as well as the GI surgeons statement that the liver and spleen mass lesions are not easily accessible ,we did not refer her for any invasive procedures or laparotomy, so, she was discharged.

This is the only HIV-Neg female patient with kaposi sarcoma in our department since 10 years ago.

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Fig. (1).



Fig. (2).

## DISCUSSION:

It is known that immunosuppression and human herpesvirus-8 (HHV-8) are associated with almost all cases of KS, including AIDS-KS, and they are believed to be necessary factors for the development of KS [6]. Other factors, however, such as trauma and physical stimuli (e.g. the Koebner phenomenon), may play a major role in the development of KS [7] It is believed that the latter factors may act by causing the displacement of basic fibroblast growth factor (bFGF) from the basement membranes also, the Koebner phenomenon may involve the release of inflammatory cytokines, such as interleukin-1 (IL-1) and IL-6 and tumor necrosis factor (TNF-), to induce normal endothelial cells to acquire features of KS spindle cells and also to induce KS cells to produce and release bFGF [8].

The current systemic treatments for KS revolve around the newer liposomal anthracyclines, paclitaxel, and vinorelbine [9]. Older agents which had been used include bleomycin, vinblastine, vincristine, and etoposide. [10]

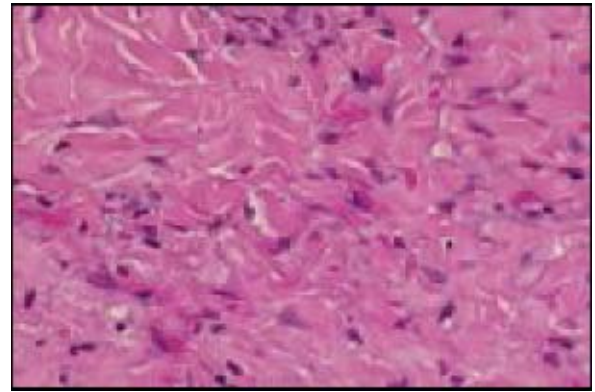


Fig. (3).

Treatment of AIDS-related KS should be integrated into the overall management of the patient [9].

In large Haramati' *et al* survey, they only identified seven women with biopsy-proven or autopsy-proven pulmonary Kaposi's sarcoma [11]. Although uncommon, pulmonary Kaposi's sarcoma should be considered in the differential diagnosis of diffuse lung disease in women with AIDS. Hepatosplenic involvement which is considered as a frequent autopsy finding is rarely diagnosed at life. Of 8 cutaneous cases of this disease, 5 also presented visceral manifestations. Seven patients died. Those with visceral tumors died because of the tumor itself [12].

Our patient was otherwise healthy and despite low ratio of CD4:CD8 ,serial monitoring of HIV Abs were negative and her past medical history was not high risk for AIDS.

It is unclear why KS is so rare in women. Malignant vascular tumors of the liver are very rare and the imaging findings are non -specific in general. Because hepatic malignant vascular tumors are often multiple, the main differential diagnosis is metastatic disease [13]. Kaposi's sarcoma in patients with rheumatologic conditions is rare. The clinical features are similar to those with classical Kaposi's sarcoma. Tumor regression usually occurs with decreasing corticosteroids and/or immunosuppressive drugs, local irradiation, or cytotoxic therapy. Kaposi's sarcoma involvement of the head and neck is often the presenting symptom of AIDS, making accuracy in diagnosis critical if intervention is to begin early. Treatment is currently directed toward palliation for pain, bleeding, dysphagia, airway obstruction, severe disfigurement, and prophylaxis for rapidly progressive tumor.

In patients with advanced AIDS, Kaposi's sarcoma will often disseminate to involve the oropharynx, larynx, tracheobronchial tree, lungs, and other viscera.[12] Hepatosplenic involvement by KS, while the patient is alive would be rare. The imaging techniques are useful to diagnose visceral involvement of KS with high probability [13]. Systematic search for visceral involvement in KS patients would lead to a marked increase in the cases such as described [14].

## REFERENCES

- [1] Friedman-Kien, A.E.; Saltzman, B.R. Clinical manifestations of classical, endemic African and epidemic AIDS-associated Kaposi's sarcoma. *J. Am. Acad. Dermatol.*, **1990**, 22, 1237-1250.

- [2] Babál, P.; Péč, J. Kaposi's sarcoma – still an enigma. *J. Eur. Acad. Dermatol. Venereol.*, **2003**, *17*(4), 377-380.
- [3] Bakari, M.; Pallangyo, K.; Kitinya, J.; Mbena, E.; Urassa, W. The importance of clinical features in differentiating HIV related from non-HIV related Kaposi's sarcoma: experience from Dar-es-Salaam, Tanzania. *Trop. Doct.*, **1996**, *26*(3), 104-7.
- [4] Pitson, G.A.; Aw, T.J.; Rodger, A. Classical Kaposi's sarcoma involving bone. *Aust. Radiol.*, **1999**, *3*, 391-393.
- [5] Szende, B.; Toth, A.; Perner, F.; Nagy, K.; Takacs, K. Clinicopathologic aspects of 8 Kaposi's sarcomas among 1009 renal transplant patients. *Gen. Diagn. Pathol.*, **1997**, *143*(4), 209-13.
- [6] Schofer, H.; Roder, C. Kaposi sarcoma in Caucasian women. Clinical, chemical laboratory and endocrinologic studies in 8 women with HIV-associated or classical Kaposi sarcoma. *Hautarzt*, **1995**, *46*(9), 632-7.
- [7] Kurt, A.; Massey, Bryon Mc Neil. Ulcerative Kaposi sarcoma in the lower extremity of an AIDS patient. *Hosp. Physician*, **2003**, *39*(9), 37-40.
- [8] Fossati, S.; Boneschi, V.; Ferrucci, S.; Brambilla, L. Human immunodeficiency virus negative Kaposi sarcoma and lymphoproliferative disorders. *Cancer*, **1999**, *85*(7), 1611-5.
- [9] Dezube, B.J.; Pantanowitz, L.; Aboulafia, D.M. Management of AIDS-related Kaposi sarcoma: advances in target discovery and treatment. *AIDS Read*, **2004**, *14*, 236.
- [10] Lee, F.C.; Mitsuyasu, R.T. Chemotherapy of AIDS-related KS. *Hematol. Oncol. Clin. North. Am.*, **1996**, *10*, 1051.
- [11] Haramati, L.B.; Wong, J. Intrathoracic Kaposi's sarcoma in women with AIDS. *Chest*, **2000**, *117*(2), 410-4.
- [12] Gonzalez-Lopez, A.; Fernandez-Martin, J.I.; Gordillo, I.; Drona, F. Disseminated Kaposi's sarcoma with hepatosplenic involvement in AIDS. *Med. Clin. (Barc)*, **1996**, *106*(16), 622-3.
- [13] Buetow, P.C.; Buck, J.L.; Ros, P.R.; Goodman, Z.D. Malignant vascular tumors of the liver: radiologic-pathologic correlation. *Radiographics*, **1994**, *14*(1), 153-66.
- [14] Caccialanza, M.; Marca, S.; Piccinno, R.; Eulisse, G. Radiotherapy of classic and human immunodeficiency virus-related Kaposi's sarcoma: results in 1482 lesions. *J. Eur. Acad. Dermatol. Venereol.*, **2008**, *22*(3), 297-302.

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