Journal of the Turkish Academy of Dermatology
eISSN 1307-394X

Case Report
DOI: 10.6003/jtad.17112c1

Lymphangioma-like Kaposi's Sarcoma in an Elderly Woman

Aslı Aksu Çerman,[1] MD, Ezgi Aktaş,[1] MD, İlknur Kıvanç Altunay,[1] MD, Özben Yalçın,[2] MD, Fevziye Kabukçuoğlu,[2] MD

Address: [1]Department of Dermatology and Venereology, [2] Department of Clinical Pathology, Şişli Hamidiye Etfal Training and Research Hospital, İstanbul, Turkey

E-mail: ezgiaktasmd@gmail.com

** Corresponding Author:* Dr. Ezgi Aktaş, Department of Dermatology and Venereology Department, Şişli Hamidiye Etfal Training and Research Hospital, 34377, İstanbul, Turkey

Published:
J Turk Acad Dermatol 2017; **11 (2)**: 17112c1
This article is available from: http://www.jtad.org/2017/2/jtad17112c1.pdf
Keywords: Bullae, lymphangioma-like Kaposi's sarcoma

Abstract

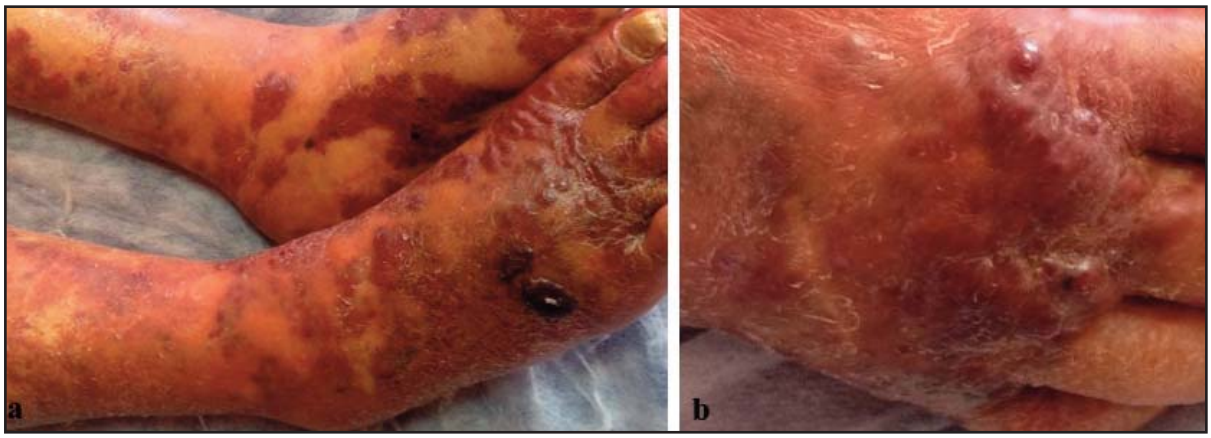
Observation: Lymphangioma-like Kaposi's sarcoma is a rare subtype of Kaposi's sarcoma which is characterized with lymphedema, swelling of the soft tissue and bullae and bulla-like lesions. Clinical appearence of lymphangioma-like Kaposi's sarcoma may cause confusion and delays with the diagnosis and the treatment. Its histologic features may mimic other benign and malignant vascular tumors such as spindle cell hemangioendothelioma, low-grade angiosarcoma, hemosiderotic hemangioma and benign lymphangioma. Here we report a case of lymphangioma-like Kaposi's sarcoma of an elderly woman presenting with deep purple coloured patchy lesions on the distal parts of upper and lower extremities with violaceous bullae.

Introduction

Kaposi's sarcoma (KS) is a multifocal vascular neoplasm characterized by clinical and histologic polymorphism. Lymphangioma-like Kaposi's sarcoma (LLKS) is a rare subtype, which can develop in all subtypes of KS [**1, 2, 3**]. LLKS presents similar to KS but is also associated with severe lymphedema and soft tissue swelling and bulla-like vascular lesions [**1**]. We report a case of LLKS in an elderly woman.

Case Report

A 90 year-old woman was presented with violaceous patch lesions on distal lower and upper extremities. Her family reported a gradual increase in the size of her hands and feet. On examination the distal parts of the lower and upper extremities were edematous and violaceous, deep purple coloured with partly petechia like patchy lesions. Multiple purple bullae on the dorsum of the hands and feet were noticed (**Figure 1**). The lesions were easily compressible and gradually resumed their original shape on release. The peripheral pulses were palpable and arteriel and venous doppler ultrasound showed no abnormalities. No lymphadenomegaly or hepato-splenomegaly was detected. In blood testing no abnormalities including HIV serology were present. A skin biopsy was performed from a bulla on the dorsum of the right hand. Histologic examination revealed an angiomatous proliferation composed of dilated vascular channels lined with banal- appearing endothelia in the upper dermis coexisting with focal areas of classic KS. Monomorphic spindle cells without nuclear atypia with a moderate inflammatory infiltrate and extravasated erytrocytes surrounding capillaries



Figures 1a and b. Edematous and violaceous, deep purple coloured with partly petechia like patchy lesions and bullae and bulla-like lesions on a) the legs and feet and b) Hands of the patient

were also seen (**Figure 2**). HHV-8 staining was positive.

A diagnosis of LLKS was made and the patient was proposed for radiotherapy however she failed to follow up.

Discussion

KS is a multicentric vascular neoplasm associated with Kaposi's sarcoma associated herpes virüs (KSHV). The KSHV is also known as the HHV-8 [**4**]. There are four types of KS seen clinically. These include classic variant KS, endemic KS, transplant-associated KS and AIDS-associated KS [**5**].

LLKS, a rare variant of KS, was first described in 1957 by *Ronchese* and *Kern* [**6**]. Since then only 27 cases have been published in the English literature. LLKS can occur in each of the four KS variants and is less than 5% of all KS cases [**2**]. LLKS can present as typical lesions of KS such as blue/purple solid lesions but also has a unique clinical appearence as bullae and bullae-like lesions [**3,7**]. Compressible bulla-like vascular lesions and lymphedema, soft tissue swelling are considered to be specific for this variant [**3,8,9**]. Bulla-like lesions are the most common clinical feature seen in LLKS. Also LLKS usually presents in the lower extremities as our patient [**7**].

The mechanism of bulla-like lesion formation is not clearly understood. Chronic lymphedema, unique lower limb hemodynamics and previous electron beam therapy have been proposed as predisposing factors [**3**]. It is believed that the factors that block the lymphatic drainage, such as tumor tissue, lymphaticovenous shunts formed on neoplastic vascular canals, lymphovascular invasion of tumor cells, the involvement of regional lymph nodes and/or cytokine release, can cause the development of intraepidermal/ subepidermal bulla secondary to absorption of lymphatic liquid by the epidermis [**10**].

The diagnosis of LLKS can be made with the typical clinical appearence and the histological evaluation of the lesions. In histological examination of LLKS lesions, lymphangioma-like spaces is characteristic. Also patterns of other KS variants can be seen on histologic examination such as spindle and endothelial cell proliferation, red blood cell extravasation, hemosiderin-laden macrophages and other signs of inflammatory reaction [**11**].

Differential diagnosis of KS with other vascular tumors may be difficult, especially when typical findings of KS are absent. Benign lymphangioendothelioma, low-grade angiosarcoma, spindle cell hemangioendothelioma, retiform hemangiendothelioma and hobnail hemangioma are the tumors that can mimic KS [**2,8,9**]. The evidence of positive staining for HHV-8 in cutaneous lesions may help to distinguish KS from its mimics [**5**].

Treatment of KS varies from simple excision of a single lesion to combination therapies of surgery, radiation and chemotherapy for multiple and metastized KS [**1**].

In conclusion, LLKS is a rare histological variant of KS that occurs most often in the skin of the lower extremities. It can present as a typical-appearing KS but a bulla-like lesion

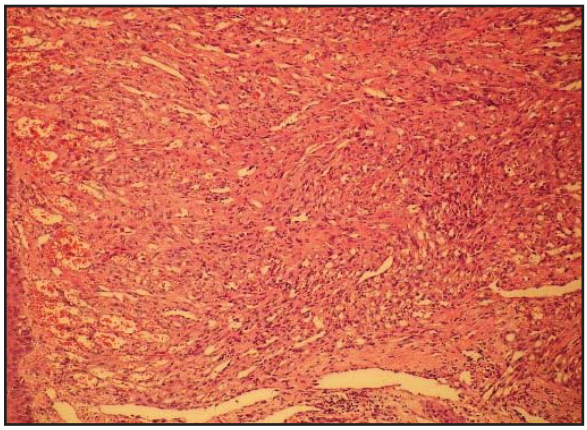


Figure 2. Histologic examination of a bulla on the dorsum of the hand: angiomatous proliferation composed of dilated vascular channels lined with banal- appearing endothelia in the upper dermiş, monomorphic spindle cells without nuclear atypia with a moderate inflammatory infiltrate and extravasated erytrocytes surrounding capillaries (HE,x100).

may be indicative of this histopathological pattern.

References

1. Friedman ER, Farquharson L, Warsch J, et al. Lymphangioma-Like Kaposi's sarcoma presenting as gangrene. Case Rep Oncol Med 2013; 2013: 839613. PMID: 23781364
2. Garcia PC, Garcia-Cruz A, Garcia-Doval I, et al. Lymphangioma-like Kaposi sarcoma, Dermatology Online J 2009; 15: 13. PMID: 19931000
3. Davis DA, Scott DM. Lymphangioma-like Kaposi's sarcoma: etiology and literature review. J Am Acad Dermatol 2000; 43: 123-127. PMID: 10863237
4. Jham BC, Ma T, Hu J, et al. Amplification of the angiogenic signal through the actiivation of the TSC/mTOR/HIF axis by the KSHV Vgpcr İN Kaposi's sarcoma. PLoS One 2011; 6: 4. PMID: 21559457
5. Dittmer DP, Richards KL, Damania B. Treatment of Kaposi sarcoma associated herpes virüs associated cancers. Front Microbiol 2012; 3: 141. PMID: 22529843
6. Ronchase F, Kern AB. Lymphangioma-like tumors in Kaposi's sarcoma. AMA Arch Derm 1979; 100: 418-427. PMID: 13402216
7. Mohanna S, Sanchez J, Ferrufino JC, et al. Lymphangioma-like Kaposi's sarcoma: report of four cases and review. J Eur Acad Dermatol Venereol 2006; 20: 1010-1011. PMID: 16922960
8. Cossu S, Satta R, Cottoni F, Masarelli G. Lymphangioma-like variant of Kaposi's sarcoma: clinicopathologic study of seven cases with review of the literature. Am J Dermatopathol 1997; 19: 16-22. PMID: 9056649
9. Ramirez JA, Laskin WB, Guitart J, Lymphangioma-like Kaposi sarcoma. J Cutan Pathol 2005; 32: 286-292. PMID: 15769278
10. Recht B, Nickoloff BJ, Wood GS. A bullous variant of Kaposi's sarcoma in an elderly female. J Dermatol Surg Oncol 1986; 12: 1192-1197. PMID: 3095406
11. Fauci AS, Lane HC. Human immunodeficiency virüs disease: AIDS and related disorders. In: Fauci AS, Braunwald E, Kasper DL et al. eds. Harrison's Principles of Internal Medicine. New York: McGraw Hill; 2008: 1086-1087.