Two Rare Variants of Kaposi Sarcoma: Case Report

Abbas Darjani[1], Seyyede Zeinab Azimi[1], Seyed Alireza Mesbah[2]

[1] *Department of Dermatology, Skin Research Center, Razi Hospital, Guilan University of Medical Sciences, Rasht, Iran*

[2] *Department of Pathology, Razi Pathobiology Laboratory, Rasht, Iran*

Received: 05 Jan. 2018; Accepted: 19 Nov. 2018

Abstract- We reported two cases of immunocompetent patients with a rare form of AIDS-associated Kaposi sarcoma (KS), without visceral involvement, presenting with an unusual clinical and histopathological picture called telangiectatic and lymphangioma-like KS, respectively. Dermatologists and pathologists need to be aware of this uncommon described variant to avoid the potential for misdiagnosis.

Acta Med Iran 2019;57(3):205-207.

Keywords: Kaposi sarcoma; Pathology; Histological variants; HHV-8; AIDS

Introduction

Kaposi Sarcoma (KS), a distinctive multicentric disorder of endothelial origin, most commonly affects the skin and gastrointestinal tract tissues (1).

The cutaneous spectrum of KS includes a patch, plaque, and nodular stages. Expanded spectrum of histomorphological variants consists of lymphadenopathic, exophytic, infiltrative, ecchymotic, telangiectatic, keloidal, and cavernous or lymphangioma-like lesions (2).

Herein, we report two unusual variants of KS in two immunocompetent patients.

Case Report

Case one

A 78-year-old woman was referred to our dermatology department in August 2017 for the treatment of her nodules and bullous of lower legs. On physical examination, she was noted to have multiple flesh-colored, and erythematous nodules on her right toes and violaceous plaque and nodules on her left lower leg since one year ago; in addition, she had a pink color patch on her right third finger from two months ago (Figure 1).

There was no hepatosplenomegaly or lymphadenopathy. Also, the remainder of her physical examination findings was unremarkable. She had diabetes mellitus and hypertension. No history of immunosuppressive drug administration was detected. A histological examination of a purple colored nodule from the left lower leg showed a dermal lesion that consisted of a proliferation of small vascular infiltrates lined with plump endothelial cells and groups of spindle-shaped pleomorphic cells with prominent nuclei arranged in short, somehow haphazard fascicles. Red blood cell (RBC) extravasations were prominent. The histology diagnosis was compatible with Kaposi's sarcoma (conventional). The concurrent biopsy of one of the nodular lesions on her right toes demonstrated large, congested ectatic vascular spaces lined by endothelial cells positive for CD34 and CD31 admixed with areas of nodular- stage KS, characteristic of telangiectatic KS (Figure 2).

Both specimens were positive for human herpesvirus 8 (HHV8). The patient was referred to an oncologist, and her evaluations were all normal. She was under observation and close follow up.

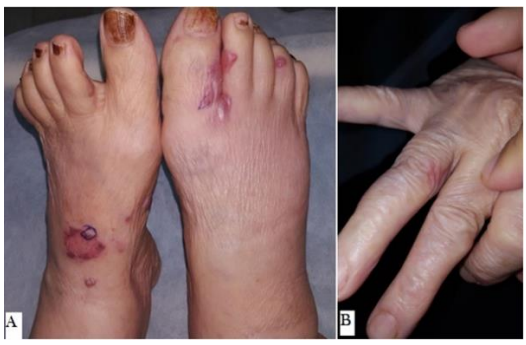

Figure 1. A: Multiple flesh-colored and erythematous nodules on the right toes and violaceous plaque and nodules on the left lower leg since one year ago; B: A pink color patch on the right third finger from two months ago

Corresponding Author: S.Z. Azimi
Department of Dermatology, Skin Research Center, Razi Hospital, Guilan University of Medical Sciences, Rasht, Iran
Tel: +98 933 7362300, Fax: +98 414 3714713, E-mail address: sz.azimi@yahoo.com

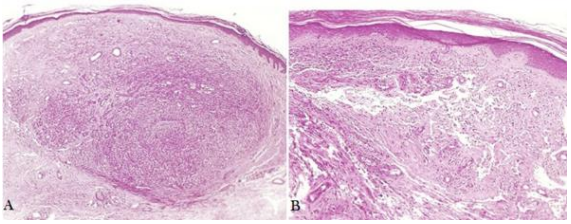

Figure 2. A: (H and E) Dermal lesion consisted of a proliferation of small vascular infiltrates lined with plump endothelial cells and groups of spindle-shaped pleomorphic cells with prominent nuclei arranged in short, somehow haphazard fascicles (Original magnification: ×40). B: (H and E): large, congested ectatic vascular spaces lined by endothelial cells admixed with areas of nodular stage KS, characteristic of telangiectatic KS (Original magnification ×40)

Case two

A 75-year-old man was visited because of purple nodular lesions and hemorrhagic bullous lesions on his lower limbs for two years (Figure 3).

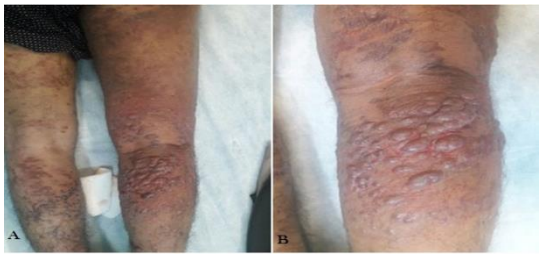

Figure 3. Clinical picture of the second case: (A and B) Purple nodular lesions and hemorrhagic bullous on the lower limbs of a 75-year-old man

He experienced cerebrovascular accident eight years ago, and since then he has taken clopidogrel and aspirin. Also, he had benign prostatic hyperplasia. He developed unilateral lymphedema from six years ago on his left lower extremity. He has never taken immunosuppressant. We performed a skin biopsy from both nodular and bullous lesions on the lower extremities. Histopathologic examination revealed vascular dilatation in the dermis and perivascular proliferation of spindle cells with the fascicular pattern. Some slit-like vascular spaces were filled by RBCs and foci with lymphangiectatic appearance were evident. Promontory sign was also evident. Immunohistochemistry study reported positive findings on CD31 and CD34 (Figure 4).

HHV-8 test using polymerase chain reaction was positive as well. So, we diagnosed as mixed conventional and lymphangiectatic (Lymphangioma-like) KS. Human immunodeficiency virus (HIV) was negative. He had no systemic involvement.

We reported two cases of immunocompetent patients with a rare form of AIDS-associated (acquired immune deficiency syndrome) KS, without visceral involvement, presenting with an unusual clinical and histopathological picture called telangiectatic and lymphangioma-like KS respectively.

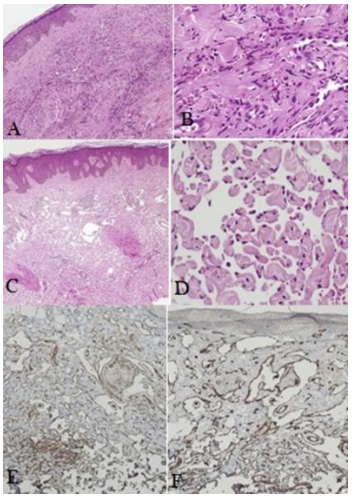

Figure 4. (H and E) (A and B) vascular dilatation in the dermis and perivascular proliferation of spindle cells with the fascicular pattern. Some slit-like vascular spaces were filled by RBCs presenting a conventional KS (Original magnification: A×40, B×100). (H and E) (C and D) foci of lymphangiectatic appearance with promontory sign were evident (Original magnification: C×40, D×100). Immunohistochemistry study reported positive findings on CD31 (E) and CD34 (F)

Discussion

KS is a low-grade malignant vascular lesion which is categorized to four different epidemiological forms including classic, endemic, iatrogenic, and AIDS-associated etiology containing HHV-8 (3). Classic type is the most common epidemiological type. Different clinical pictures and histopathological features make numerous morphologic variants of KS lesions (4).

In classic KS, the disease is typically limited to the lower extremities; however, it may be more extensive. In immunodeficient patients, such as patients with AIDS or those with a solid organ transplant, KS presents as a multifocal systemic disease (5).

Telangiectatic KS which is an uncommon variant of KS, displays erythematous translucent nodules with prominent telangiectasia. KS usually presents with blue or violaceous hue. Erythematous and flesh-colored nodules are uncommon (6).

Due to the dilated ectatic vascular spaces, telangiectatic KS could be misdiagnosed as sinusoidal hemangioma which is an uncommon acquired form of

cavernous hemangioma, and aneurysmal fibrous histiocytoma, although HHV8 is only positive in KS (1).

Clinical and histological lymphedematous forms of AIDS-associated KS have been described before. These include variants of ectatic lymphatics (also called lymphangioma-like and lymphangiectatic KS) and variants with subepidermal and intraepidermal edema (so-called as bullous KS). Lymphangiectatic and lymphangioma-like KS are presented with ectatic lymphatics. However, in lymphangiectatic KS there were large dilated intratumoral and peritumoral thin-walled lymphatic vessels (2).

We could find two cases of telangiectatic KS which had been reported before. A man with thymoma and myasthenia gravis receiving long-term immunosuppressive therapy has been reported before (6). The second case was a 36-year-old South African man who presented with a multinodular mass in his posterior auricular area. The clinical manifestation provided dark hemorrhagic, firm to the spongy cut surface lesion. He was also HIV positive (7).

In HIV patients, KS is one of the indications for starting anti-retroviral treatment. It has considerably diminished the morbidity and mortality associated with KS (9). Local treatments are useful if cutaneous or mucosal lesions are few in association with no systemic involvement. Cryotherapy, intralesional administration of vinblastine, laser and irradiation can be used in such cases. Internal involvement, extensive lymphedema associated with KS, widespread and rapidly progressive cutaneous KS and failure to local treatment are indications for systemic treatment. Doxorubicin and daunorubicin paclitaxel are drugs of choice. Interferon-alpha and combination chemotherapies are other systemic modalities (9).

To the best of our knowledge, this is the first report of unusual variants of cutaneous KS (telangiectatic and Lymphangiectatic variants respectively) in immunocompetent patients, reported from the Middle East. The clinical significance of these unusual variants discussed in this report has yet to be determined. Finally, pathologists need to be aware of this uncommon described variant to avoid the potential for misdiagnosis.

References

1. Ramdial PK, Miles E, Pillay B. The Emerging Histopathologic Diagnostic Challenges of Kaposi Sarcoma in the Acquired Immunodeficiency Syndrome Era. Austin J HIV/AIDS Res 2016;3:1020.
2. Grayson W, Pantanowitz L. Histological variants of cutaneous Kaposi sarcoma. Diagn Pathol 2008;25;3:31.
3. Weiss SW, Goldblum JR. In: Enzinger and Weiss's soft tissue tumors. 4th ed. Strauss M, Gery L, editor. Philadelphia: Mosby, 2001:938-47.
4. Kandemir NO, Barut F, Gün BD, Tekin NS, Keser SH, Özdamar ŞO. Histopathological analysis of vesicular and bullous lesions in Kaposi sarcoma. Diagn Pathol 2012;7:101.
5. Santos M, Vilasboas V, Mendes L, Talhari C, Talhari S. Lymphangiectatic Kaposi's sarcoma in a patient with AIDS. Anais Bras Dermatol 2013;88:276-8.
6. Snyder RA, Schwartz RA. Telangiectatic Kaposi's sarcoma. Occurrence in a patient with thymoma and myasthenia gravis receiving long-term immunosuppressive therapy. Arch Dermatol 1982;118:1020-1.
7. O'Donnell PJ, Pantanowitz L, Grayson W Unique histologic variants of cutaneous Kaposi sarcoma. Am J Dermatopathol 2010;32:244-50.
8. Schwartz RA. Kaposi's sarcoma: an update. J Surg Oncol 2004;87:146-51.
9. Dongre A, Montaldo C. Kaposi's sarcoma in an HIV-positive person successfully treated with paclitaxel. Indian J Dermatol Venereol Leprol 2009;75:290-2.