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CASE REPORT

A rare histologic variant of Kaposi's sarcoma

Uma variante histológica rara de sarcoma de Kaposi

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Abstract

An 80-year-old male presented with a 3-year history of progressive violaceous and edematous plaques on both hands. The remaining physical examination was unremarkable and systemic symptoms were absent. All complementary investigations were negative, including serology testing for human immunodeficiency virus. Skin biopsy revealed a dermal vascular proliferation, composed of angulated vessels dissecting collagen bundles, lined by flattened endothelial cells without atypia. Immunohistochemistry showed positive staining for human herpesvirus 8, CD31, and erythroblast transformation-specific-related gene, confirming the diagnosis of lymphangioma-like Kaposi's sarcoma (LLKS). LLKS represents a rare pathological variant of all Kaposi's sarcoma (KS) subtypes, which is noticeable for the lymphangioma-like spaces that are usually only focally found. We present a case of LLKS as a variant of classic KS to raise awareness among dermatologists for this uncommon morphologic expression of KS and its distinctive histologic pattern.

Keywords: Kaposi sarcoma. Lymphangioma. Human herpesvirus-8. Pathology.

Resumo

Um homem de 80 anos apresentou um quadro progressivo com 3 anos de evolução de placas violáceas e edematosas em ambas as mãos. O restante exame físico não revelou alterações e sintomas sistémicos estavam ausentes. Todos os exames complementares foram negativos, incluindo a serologia para VIH. A biopsia cutânea revelou uma proliferação vascular dérmica composta por vasos angulados, revestidos por células endoteliais achatadas sem atipia, que dissecavam os feixes de colagénio. A imunohistoquímica revelou positividade para HHV-8, CD31 e ERG, confirmando o diagnóstico de sarcoma de Kaposi semelhante a linfangioma (SKSL). O SKSL representa uma variante histológica rara de todos os subtipos de sarcoma de Kaposi (SK), que se destaca pelos espaços semelhantes a linfangiomas, normalmente encontrados apenas de forma focal. Apresentamos um caso de SKSL como uma variante do SK clássico para sensibilizar os dermatologistas para esta expressão morfológica incomum de SK e para o seu padrão histológico distinto.

Palavras-chave: Sarcoma de Kaposi. Linfangioma. Herpesvírus humano 8. Patologia.

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Introduction

Kaposi's sarcoma (KS) is a low-grade vascular neoplasm caused by human herpesvirus 8 (HHV-8) with a broad spectrum of clinicopathological manifestations[1].

For the past decades, multiple histologic variants of KS have been reported, namely the lymphedematous subtypes, which include lymphangioma-like KS (LLKS), lymphangiectatic KS, and bullous KS[2].

LLKS corresponds to < 5% of all reported KS cases and is mostly defined by the presence of ectatic vascular spaces with a labyrinthine architecture that dissects collagen bundles[3].

We hereby present the case of a patient with lymphangioma-like classic KS, reviewing the clinical and pathological features of this rare entity.

Case report

An 80-year-old male, without relevant past medical history, presented to our department with erythematous to violaceous edematous plaques on the dorsum of both hands, especially on his right side (Fig. 1). These were soft and easily compressible lesions which had been slowly growing for 3 years and were now starting to limit his daily activities. On physical examination, no other mucocutaneous lesions were identified and lymph nodes were not palpable.

The patient denied any systemic symptoms, as well as prior malignancy or radiation therapy.

Routine laboratory workup, including complete blood count, was normal and serology testing for human immunodeficiency virus was negative. Additional investigation included a chest X-ray, an abdominal computed tomography scanning, and a fecal occult blood test, which were all negative.

A skin punch biopsy was performed on the dorsum of the right hand and revealed an ill-defined vascular proliferation on the dermis admixed with an inflammatory infiltrate of lymphocytes and rare plasma cells (Fig. 2A). The proliferation was composed of anastomosing angulated spaces lined by flattened endothelial cells, dissecting collagen bundles and surrounding pre-existing adnexal structures and blood vessels (Fig. 2B). No significant cytological atypia or mitoses was observed. There was no relevant hemosiderin deposition. Immunohistochemistry showed diffuse positive staining for HHV-8 (Fig. 3), as well as positive expression of CD31 and erythroblast transformation-specific-related gene (ERG) by endothelial cells.

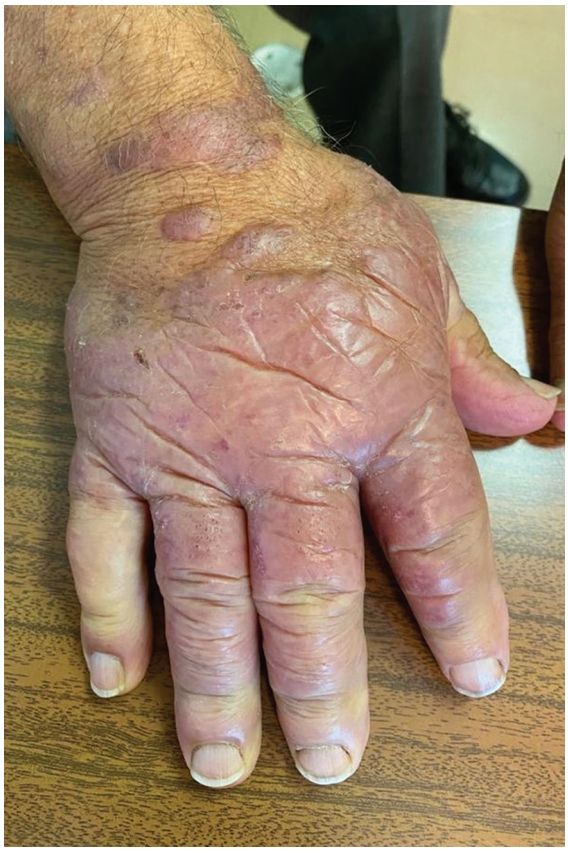

Figure 1. Violaceus and edematous plaques on the dorsum of the right hand and fingers.

A diagnosis of lymphangioma-like Kaposi sarcoma (LLKS) was established. Therapeutic options were discussed but the patient declined any treatment, so a "wait-and-see" approach was adopted. At the last follow-up visit, his lesions remained stable.

Discussion

KS is a multifocal vascular neoplasm with four epidemiological subtypes recognized by the World Health Organization: classic or mediterranean, endemic or African, iatrogenic or post-transplant and epidemic or Acquired immunodeficiency syndrome-associated[4].

Mucocutaneous lesions of KS typically evolve from early macules (patch stage) into plaques (plaque stage) that may later grow into larger nodules (tumor stage)[1]. Extracutaneous locations, most commonly lymph nodes, lungs, or the gastrointestinal tract, may also be affected[1].

Histologic features of KS vary with the stage of the lesion (patch, plaque, or nodule) but generally include ill-defined fascicles of spindle cells associated with

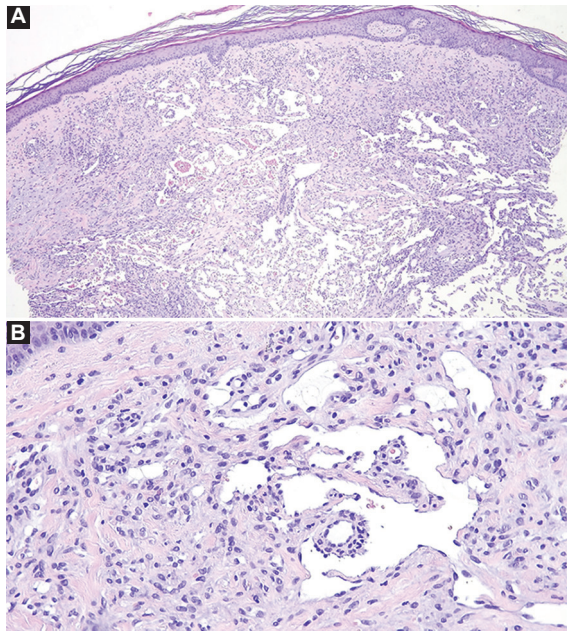

Figure 2. A: vascular proliferation with numerous thin-walled ectatic vessels dissecting collagen bundles and anastomosing lymphangioma-like spaces (hematoxylin and eosin [H&E], ×10); **B:** lymphangioma-like ectatic immature vessels lined by single-layered endothelial cells with isolation of one pre-existing vessel (H&E, ×100).

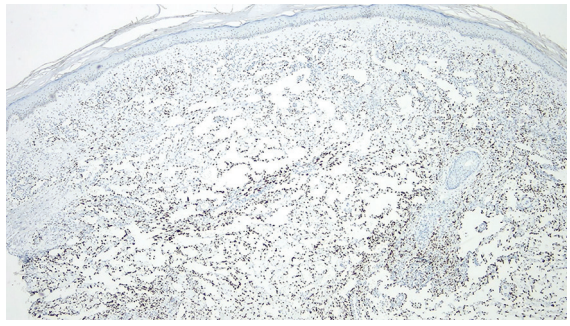

Figure 3. Positive diffuse immunostaining for human herpesvirus 8.

slit-like vascular spaces that dissect the dermis and sometimes circulate around adnexal structures and pre-existing native vessels ("promontory" sign)[5]. A lymphoplasmacytic inflammatory infiltrate, extravasated red blood cells, hemosiderin deposits, and occasional hyaline globules are additional histologic clues[1,5].

Over the years, numerous pathological KS variants have been described such as anaplastic, bullous, telangiectatic, keloidal, verrucous, micronodular, ecchymotic, pyogenic granuloma-like and the lymphedematous subtypes, which comprise lymphangiectatic, lymphangioma-like and bullous KS[1,2].

Lymphangioma-like KS (LLKS) corresponds to < 5% of all reported KS cases and may virtually occur in all epidemiological settings[6], although Ramirez et al. suggest a higher prevalence in patients with classic KS[5].

Previous irradiation and chronic lymphedema have been hypothesized as risk factors for the development of LLKS, but their presence is not mandatory[7,8].

Clinically, LLKS is characterized by the presence of tense vesiculobullous lesions with a serious content[9,10]. However, blisters are not always present and, in some cases, the clinical presentation may be indistinguishable from typical KS lesions[6].

This entity affects mainly the extremities and is more frequent in males, usually above the sixth decade of life[11,12], although a lower mean age at onset (45.1 years) has been reported in 2017[6].

On histology, LLKS stands out for its distinctive anastomosing networks of ectatic vessels in the dermis, which are lined by a layer of flat endothelial cells with no - or very mild - atypia[3,5], resembling the dilated lymphatic channels of a lymphatic tumor, such as a benign lymphangioendothelioma/acquired progressive lymphangioma[2]. By contrast to classic KS, there is no prominent population of spindle cells[13] and red blood cells, lymphocytes or thrombi are usually absent in those spaces[5].

These findings are normally found in up to 50% of the lesional tissue, with adjacent typical KS histologic features that make the diagnosis easier[5]. On the other hand, when lymphangioma-like spaces occupy the entire lesion, differential diagnosis with other vascular tumors such as hemangiomas or hemangioendotheliomas may be challenging[3]. Immunohistochemical studies with positive staining for endothelial cell markers (CD31, CD34, and ERG), lymphatic markers (D2-40) and especially for HHV-8 may be key for confirming the diagnosis[7,14].

The treatment of LLKS is similar to conventional KS, depending on the patient's symptoms, immune status, the number of lesions, and the involvement of extracutaneous sites[6].

Unlike other KS histologic variants[2], LLKS seems to have prognostic significance, with a more indolent course than the traditional classic subtype[6]. This helped to further validate the "wait-and-see" option taken together with the patient in our case.

We believe this case is a good example of lymphangioma-like classic KS, presenting with the typical histologic features but some less common clinical findings as the absence of bullae.

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Conflicts of interest

None.

Ethical disclosures

Protection of human and animal subjects. The authors declare that no experiments were performed on humans or animals for this study.

Confidentiality of data. The authors declare that they have followed the protocols of their work center on the publication of patient data.

Right to privacy and informed consent. The authors have obtained the written informed consent of the patients or subjects mentioned in the article. The corresponding author is in possession of this document.

Use of artificial intelligence for generating text. The authors declare that they have not used any type of generative artificial intelligence for the writing of this manuscript nor for the creation of images, graphics, tables, or their corresponding captions.

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