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**CASE REPORT**

# Lymphangioma-like Kaposi sarcoma: clinical, pathological, and microvascular imaging correlation

## *Sarcoma de Kaposi linfangioma-símile: correlação clínica, patológica e por imagem microvascular*

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## Abstract

Lymphangioma-like Kaposi sarcoma is a rare variant that may clinically mimic lymphatic or bullous lesions. We report the case of a 71-year-old immunocompetent man with progressive violaceous plaques, nodules, bullous-like lesions, and marked edema of the lower limbs. Histopathology showed spindle-cell proliferation with ectatic vascular spaces, and immunohistochemistry was positive for human herpesvirus 8 (HHV-8) confirming Kaposi sarcoma. Serological tests for human immunodeficiency virus and other infections were negative, and imaging studies showed no systemic involvement. High-frequency dermatologic ultrasound with superb microvascular imaging (SMI) demonstrated dermal anechoic and hypoechoic lymphatic-like structures with variable vascular flow. The patient received liposomal doxorubicin, with marked clinical improvement and complete resolution of edema. This case highlights an uncommon presentation of Kaposi sarcoma in an immunocompetent patient and suggests that high-frequency ultrasound may assist in lesion characterization and therapeutic follow-up.

***Keywords:*** Kaposi sarcoma. Human herpesvirus 8. Lymphangioma-like. Dermatologic ultrasound. Superb microvascular imaging.

## Resumo

O sarcoma de Kaposi linfangioma-símile é uma variante rara que pode mimetizar clinicamente lesões linfáticas ou bolhosas. Relatamos o caso de um homem imunocompetente de 71 anos com placas violáceas progressivas, nódulos, lesões pseudobolhosas e edema acentuado dos membros inferiores. O exame histopatológico evidenciou proliferação de células fusiformes com espaços vasculares ectásicos, e a imuno-histoquímica foi positiva para herpesvírus humano 8, confirmando sarcoma de Kaposi. Sorologias para vírus da imunodeficiência humana e outras infecções foram negativas, e os exames de imagem não demonstraram acometimento sistêmico. A ultrassonografia dermatológica de alta frequência com superb microvascular imaging demonstrou estruturas dérmicas anecoicas e hipoecoicas linfático-símiles, com fluxo vascular variável. O paciente recebeu doxorrubicina lipossomal, com melhora clínica acentuada e resolução do edema. Este caso destaca uma apresentação incomum de sarcoma de Kaposi em paciente imunocompetente e sugere que a ultrassonografia de alta frequência pode auxiliar na caracterização e no seguimento das lesões.

***Palavras-chave:*** Sarcoma de Kaposi. Herpesvírus humano 8. Linfangioma-símile. Ultrassonografia dermatológica. Relato de caso.

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## Introduction

Kaposi sarcoma is a multifocal vascular neoplasm associated with human herpesvirus 8 (HHV-8). Although classically associated with immunosuppression, Kaposi sarcoma may occur in immunocompetent individuals, particularly in its classical form. In such cases, atypical presentations may delay diagnosis.[1]

Lymphangioma-like Kaposi sarcoma is an uncommon variant characterized by dilated vascular spaces resembling lymphatic channels. Because it may present with bullous or pseudovesicular lesions, it can be mistaken for lymphatic malformations or other vascular disorders.[2]

Reports describing high-frequency dermatologic ultrasound and superb microvascular imaging (SMI) findings in this variant are scarce. This case describes an immunocompetent patient with lymphangioma-like Kaposi sarcoma and emphasizes the potential value of non-invasive imaging in lesion assessment and follow-up.

## Clinical case

A 71-year-old man, a retired truck driver, presented to a tertiary hospital in December 2023 with pain, edema, erythema, and dark bullous-like lesions affecting the lower limbs. He reported a 4-month period of progressive lesion enlargement, superimposed on a 4-year history of edema, insidious violaceous plaques and pseudobullous lesions on the distal lower limbs, with gradual increase in number and size.

Lower-limb Doppler ultrasound ruled out deep venous thrombosis. He was initially treated for presumed soft-tissue infection and underwent dermatologic evaluation with skin biopsy. The infectious episode resolved after oral antibiotic therapy. In January 2024, histopathological examination revealed spindle cell proliferation with ectatic vascular spaces, and immunohistochemistry was positive for HHV-8, confirming the diagnosis of lymphangioma-like Kaposi sarcoma, and the patient was referred for oncologic evaluation due to extensive bilateral lower-limb involvement.

Review of systems revealed marked bilateral lower-limb edema and violaceous nodules and plaques. He denied oral or genital mucosal lesions, weight loss, fever, hyporexia, and recurrent infections. His medical history included chronic obstructive pulmonary disease, benign prostatic hyperplasia and cardiovascular disease. He was a former heavy smoker and reported social alcohol use.

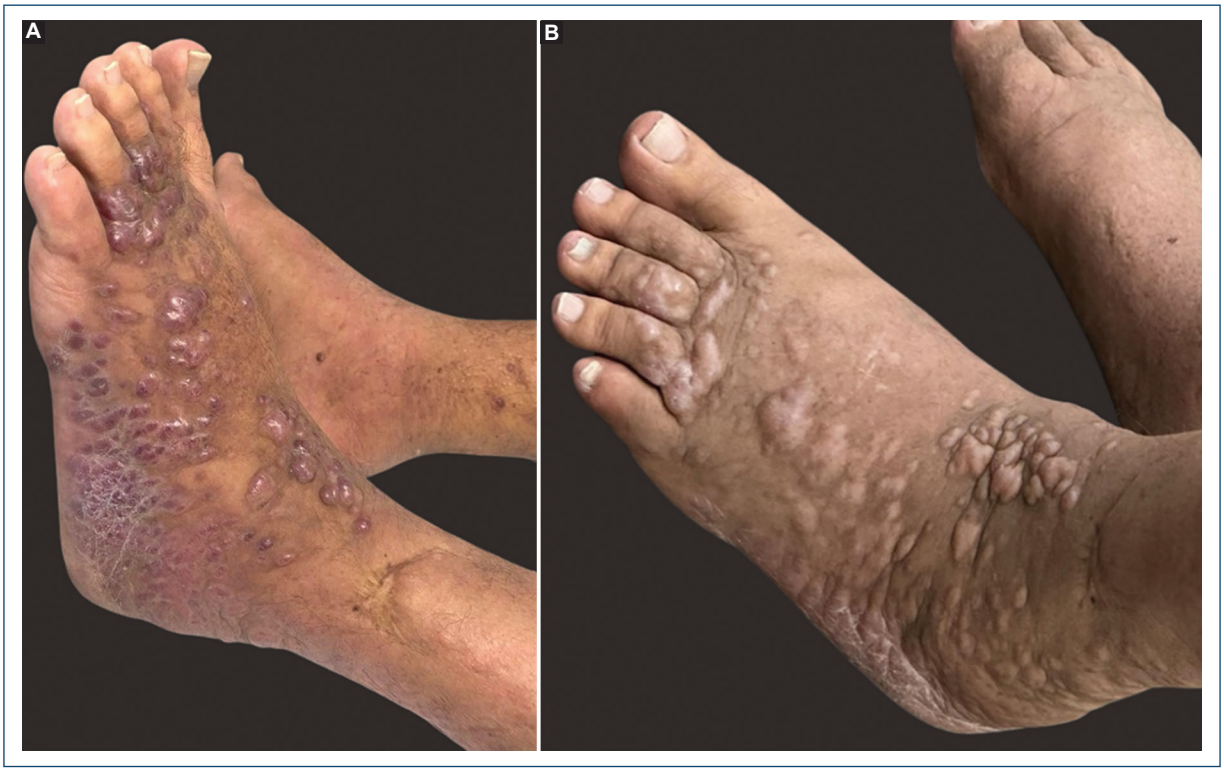


**Figure 1.** Cutaneous lesions involving the lower limbs. **A:** Clinical presentation at baseline showing violaceous plaques, nodules, and bullous-like lesions on the lower limbs **B:** with marked improvement after systemic treatment with liposomal doxorubicin.

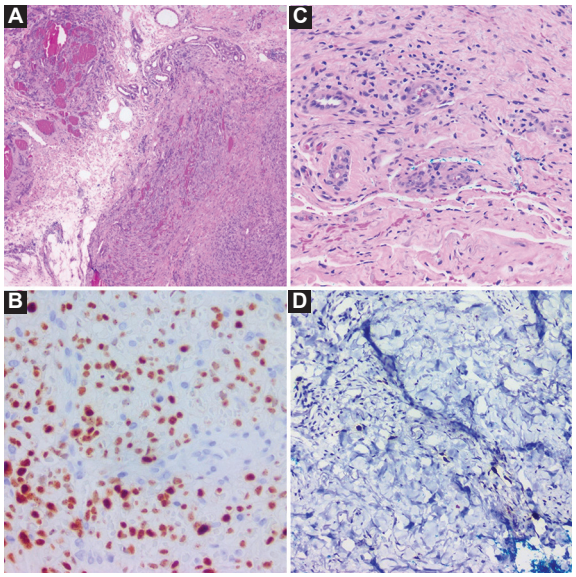


**Figure 2.** Histopathological and immunohistochemical findings before and after systemic treatment. Pre-treatment findings are shown in the left column, with histopathology above **A:** (H&E, ×40) and human herpesvirus 8 (HHV-8) immunohistochemistry below **B:** (×400), demonstrating strong nuclear HHV-8 expression. Post-treatment findings are shown in the right column, with histopathology above **C** (H&E ×200) and HHV-8 immunohistochemistry below **D** (×400), demonstrating residual HHV-8-positive cells. Residual disease on histopathology and ultrasound led to subsequent radiotherapy, with good clinical response.

Dermatologic examination revealed violaceous nodules of soft consistency, some with a bullous-like appearance, associated with erythematous-violaceous plaques. Bilateral painful pitting edema and diffuse erythema of the legs and feet were present (Fig. 1A).

Histopathology demonstrated dermal spindle-cell proliferation arranged in nodular patterns, forming irregular vascular spaces, with the presence of hemosiderin-laden macrophages. Immunohistochemistry was positive for HHV-8 in neoplastic cells (Fig. 2). Direct immunofluorescence for immunoglobulin A, immunoglobulin G, immunoglobulin M, and complement component C3c was negative.

Computed tomography of the chest, abdomen, and pelvis showed no mediastinal, hilar, or retroperitoneal lymphadenopathy and no pleural effusion. Laboratory tests, including renal and hepatic function and lactate dehydrogenase, were within normal limits. Serological tests for hepatitis B and C, human immunodeficiency virus types 1 and 2, syphilis, and human T-cell lymphotropic virus types 1 and 2 were negative. Additional investigations for immunodeficiency were negative.

The patient received systemic chemotherapy with liposomal doxorubicin at a dose of 20 mg/m$^2$. After seven sessions, there was marked improvement of the lower-limb lesions and complete resolution of edema (Fig. 1B). Follow-up dermatologic examination revealed residual asymptomatic soft, compressible, confluent nodules, and scattered erythematous-violaceous macules. High-frequency ultrasound demonstrated dermal anechoic and hypoechoic oval structures compatible with lymphatic-like spaces. SMI showed variable vascular flow, supporting residual lesion activity in selected areas (Figs. 3 and 4). No significant edema was observed; therefore, regular physical activity was recommended, without specific compressive measures. Clinical monitoring was maintained monthly during the first 3 months, followed by quarterly visits, and is currently performed semiannually.

## Discussion

We describe a case of lymphangioma-like Kaposi sarcoma in an immunocompetent patient, with clinicopathological correlation and complementary imaging findings. This is an uncommon histopathological variant of Kaposi sarcoma, characterized by dilated vascular spaces resembling lymphatic channels, which may pose diagnostic challenges, particularly in atypical clinical presentations.[2]

Although typically associated with immunosuppression, classical Kaposi sarcoma may also occur in immunocompetent individuals, particularly in its classical form. In such cases, atypical presentations may delay diagnosis.[1]

The lymphangioma-like variant is rare and characterized by prominent dilation of vascular spaces mimicking lymphatic vessels.[2] This pattern is thought to be related to HHV-8-induced angiogenic and lymphangiogenic pathways, involving VEGF-C- and VEGF-D-mediated pathways, leading to the formation of ectatic vascular channels and contributing to the bullous or pseudovesicular clinical appearance observed in some patients.[2,3]

From a diagnostic perspective, the main challenge lies in distinguishing this variant from other lymphatic or vascular conditions, including acquired lymphangioma, lymphatic malformations, pseudolymphangiomatous lesions, and angiosarcoma. Histopathological examination, supported by HHV- 8 immunohistochemistry, remains essential for definitive diagnosis.[2,4]

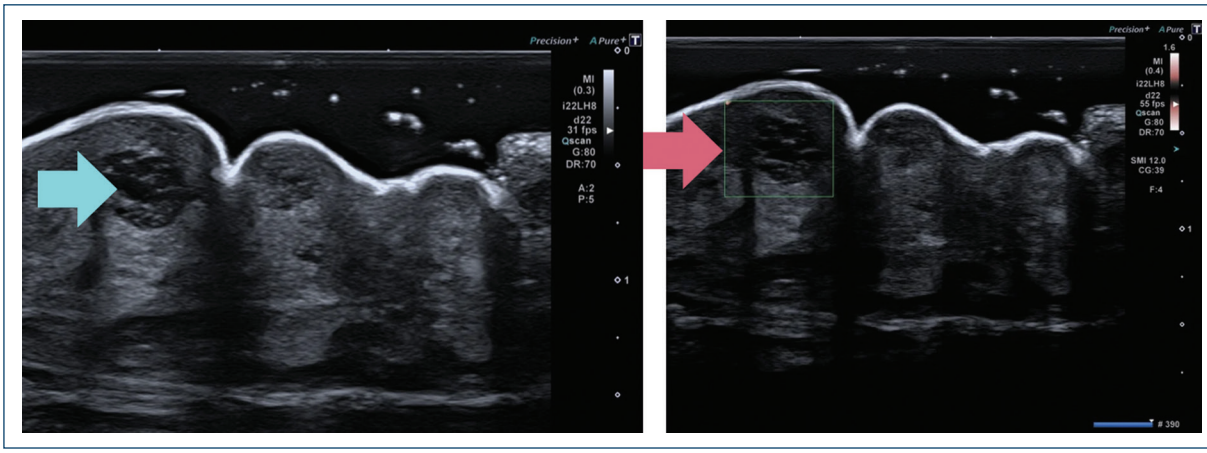

**Figure 3.** High-frequency ultrasound demonstrating anechoic and hypoechoic dermal structures without detectable flow on superb microvascular imaging, compatible with lymphatic-like spaces.

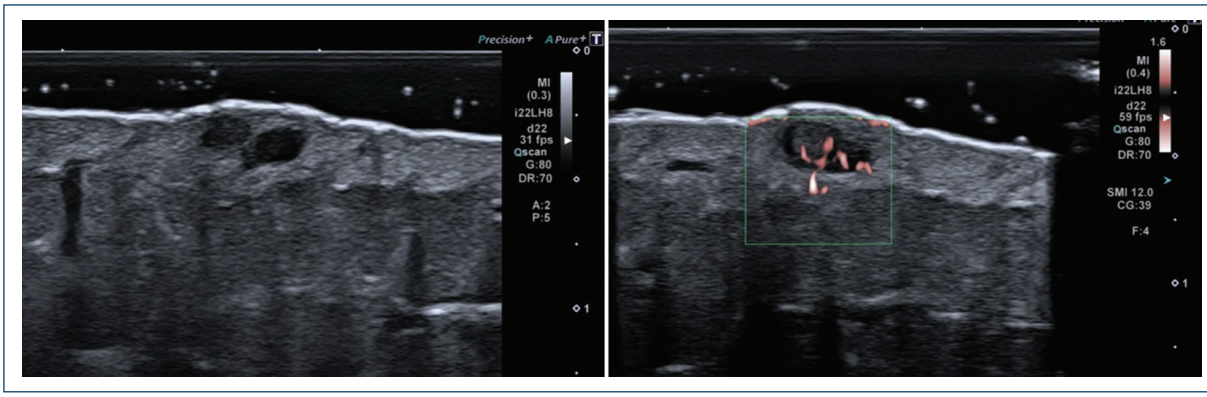

**Figure 4.** High-frequency ultrasound showing dermal lymphatic-like structures with increased flow on superb microvascular imaging, suggesting residual lesion activity.

An important aspect of this case was the use of high-frequency dermatologic ultrasound associated with SMI, which demonstrated anechoic and hypoechoic dermal structures compatible with lymphatic-like channels, with variable vascular flow patterns. Although ultrasound has been increasingly used in dermatology for the assessment of cutaneous tumors, inflammatory diseases, and vascular lesions, its application in specific variants of Kaposi sarcoma remains limited.[5] These findings suggest that ultrasound may serve as a useful adjunctive tool for lesion characterization and therapeutic monitoring.

Treatment of Kaposi sarcoma should be individualized based on disease extent, symptoms, and clinical subtype. In this case, systemic chemotherapy with liposomal doxorubicin resulted in significant clinical improvement, reinforcing its role in extensive disease, even in the absence of immunosuppression.[6]

## Conclusion

This case highlights a rare presentation of Kaposi sarcoma in an immunocompetent patient and underscores the importance of recognizing uncommon histopathological variants of Kaposi sarcoma. In addition, it supports the potential role of high-frequency ultrasound as a complementary non-invasive tool.


## Funding

None.

## Conflicts of interest

None.

## Ethical considerations

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

**Confidentiality, informed consent, and ethical approval.** This study does not involve personal patient data, medical records, or biological samples, and does not require ethical approval. SAGER guidelines do not apply.

**Declaration on the use of artificial intelligence.** The authors declare that no generative artificial intelligence was used in the writing or creation of the content of this manuscript.

## References

1. Antman K, Chang Y. Kaposi's sarcoma. N Engl J Med. 2000;342:1027-38.
2. Davis DA, Scott DM. Lymphangioma-like Kaposi's sarcoma: etiology and literature review. J Am Acad Dermatol. 2000;43:123-7.
3. Dittmer DP, Damania B. Kaposi sarcoma associated herpesvirus pathogenesis(KSHV) --an update. Curr Opin Virol. 2013;3:238-44.
4. Cesarman E, Chang Y, Moore PS, Said JW, Knowles DM. Kaposi's sarcoma-associated herpesvirus-like DNA sequences in AIDS-related body-cavity-based lymphomas. N Engl J Med. 1995;332:1186-91.
5. Hladik W, Dollard SC. Kaposi's sarcoma-associated herpesvirus seroprevalence in sub-Saharan Africa: a review. Infect Agent Cancer. 2013;8:22.
6. Montaner LJ, Celentano DD. Prevention and treatment of Kaposi's sarcoma in the era of HIV/AIDS. Clin Infect Dis. 1998;26:994-1003.