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CASE REPORT

Vascular neoplasms of the lower limbs: similar clinical presentation, distinct prognoses

Neoplasias Vasculares dos Membros Inferiores: Clínica Semelhante, Prognósticos Distintos

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Abstract

Angiosarcoma and classic Kaposi sarcoma are vascular neoplasms that may present with similar clinical features but have distinct prognoses and therapeutic implications. We describe two patients presenting with violaceous plaques and nodules on the lower extremities. Histopathological evaluation was essential for the differential diagnosis. In the first case, a high-grade epithelioid endothelial proliferation was observed, showing marked atypia, frequent mitoses, positivity for ETS-related gene, and negativity for human herpesvirus 8 (HHV-8), consistent with cutaneous angiosarcoma. In the second case, a spindle-cell vascular proliferation with irregular, thin-walled vessels and HHV-8 positivity was identified, confirming Kaposi sarcoma. Despite clinical similarity, angiosarcoma exhibits aggressive behavior and carries a poor prognosis, whereas classic Kaposi sarcoma typically follows an indolent course. Early biopsy with immunohistochemical characterization is essential for accurate diagnosis and appropriate therapeutic guidance.

Keywords: Angiosarcoma. Kaposi sarcoma. Vascular neoplasms. Human herpesvirus 8. Immunohistochemistry.

Resumo

O angiossarcoma e o sarcoma de Kaposi clássico são neoplasias vasculares que podem manifestar-se com clínicas semelhantes, mas com prognósticos e implicações terapêuticas distintas. Descrevem-se dois doentes com placas e nódulos violáceos nos membros inferiores. O estudo histopatológico foi determinante para o diagnóstico diferencial. No primeiro caso, observou-se proliferação endotelial epitelióide de alto grau, com atipia marcada, mitoses frequentes, positividade para gene relacionado com a sequência específica de transformação E26 (ERG) e negatividade para herpesvírus humano 8 (HHV-8), compatível com angiossarcoma cutâneo. No segundo caso, identificou-se proliferação vascular fusocelular com vasos irregulares de parede fina e positividade para HHV-8, confirmando sarcoma de Kaposi. Apesar da semelhança clínica, o angiossarcoma apresenta comportamento agressivo e mau prognóstico, enquanto o sarcoma de Kaposi clássico tem evolução geralmente indolente. A biópsia precoce com caracterização imunohistoquímica é fundamental para diagnóstico e orientação terapêutica adequada.

Palavras-chave: Angiossarcoma. Sarcoma de Kaposi. Neoplasias vasculares. HHV-8. Imuno-histoquímica.

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Introduction

Cutaneous violaceous vascular lesions of the lower limbs in elderly patients represent a significant diagnostic challenge in clinical practice. Cutaneous angiosarcoma and classic Kaposi sarcoma, although sharing similar clinical features, are distinct entities with fundamentally different biological behaviors, prognoses, and therapeutic approaches.[1-3]

Cutaneous angiosarcoma is an aggressive malignant neoplasm of vascular endothelial origin, accounting for < 1% of all soft-tissue sarcomas.[2] Although it most commonly arises on the scalp and face, it may also involve the lower extremities.[1,4] Its clinical presentation is often deceptive, frequently mimicking benign conditions such as ecchymoses or hemangiomas, thereby contributing to significant diagnostic delays.[4] Histologically, it is characterized by a proliferation of atypical endothelial cells displaying marked nuclear pleomorphism, prominent nucleoli, and frequent mitotic figures, forming irregular vascular channels that dissect collagen bundles.[1] Immunohistochemically, tumor cells typically express endothelial markers such as cluster of differentiation 31 (CD31), CD34, and E26 transformation-specific–related gene (ERG), the latter being a highly specific marker for endothelial vascular neoplasms.[5] The prognosis remains poor, with 5-year survival rates reported to be as low as 9% in some series.[2]

In contrast, classic Kaposi sarcoma is a low-grade angioproliferative neoplasm associated with infection by human herpesvirus 8 (HHV-8), predominantly affecting elderly men of Mediterranean or Eastern European descent.[3,6] It typically follows an indolent clinical course, with violaceous lesions that progress slowly over years or decades, usually beginning on the lower extremities.[6,7] Immunohistochemical detection of the HHV-8 latent nuclear antigen-1 is pathognomonic and essential for differential diagnosis.[8,9] The prognosis is generally more favorable than that of angiosarcoma, with 5-year survival rates of approximately 75% in Kaposi sarcoma.[7]

In this report, we present two clinical cases that exemplify this diagnostic dilemma. This case report was prepared following the CAse REport guidelines.[10]

Clinical case

Case 1

A 92-year-old man with no history of chronic lymphedema or prior radiotherapy presented with multiple violaceous plaques and nodules, some ulcerated, predominantly on the anterior aspect of the left lower leg. The largest lesion measured approximately 12 cm in diameter. The lesions had progressively increased in number and size over 2 months in a centrifugal pattern (Fig. 1). No palpable lymphadenopathy was detected.

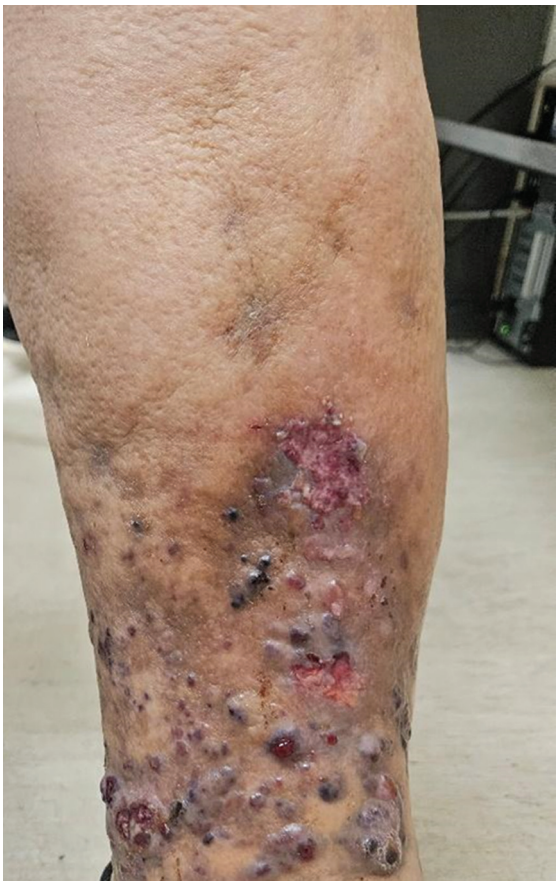

Figure 1. Multiple violaceous papules and nodules with areas of ulceration and hemorrhagic crusting on the lower leg of an elderly patient (cutaneous angiosarcoma).

An incisional biopsy demonstrated a malignant proliferation of epithelioid endothelial cells with marked cytological atypia, vesicular nuclei, prominent nucleoli, and frequent mitotic figures within a hemorrhagic stroma (Fig. 2). Immunohistochemistry showed strong nuclear ERG expression and absence of HHV-8 immunoreactivity (Figs. 3 and 4). These findings established the diagnosis of cutaneous angiosarcoma.

The case was discussed in a multidisciplinary tumor board, and palliative radiotherapy was proposed given the extent of disease and patient age. However, treatment was not initiated, as the patient passed away 1 month after the group's decision.

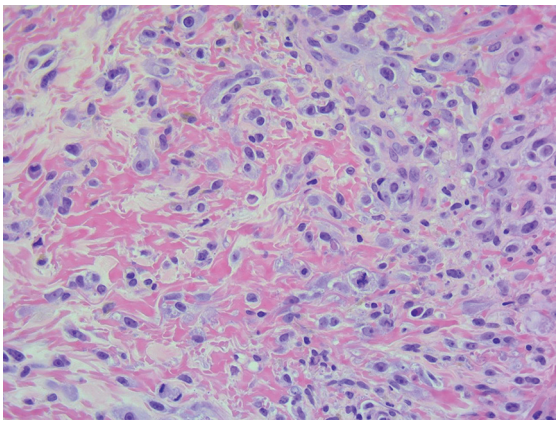

Figure 2. Cutaneous angiosarcoma showing epithelioid endothelial proliferation with marked atypia (H&E stain, original magnification × 400).

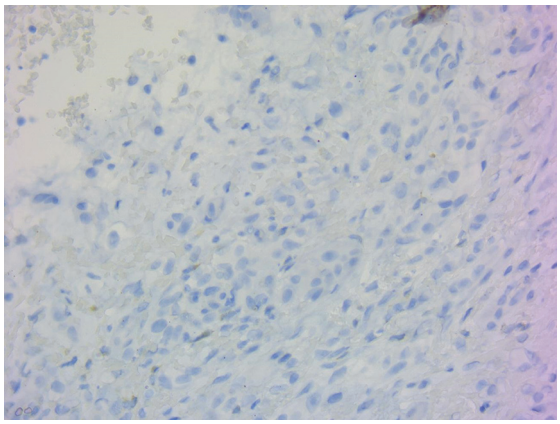

Figure 4. Absence of human herpesvirus 8 immunoreactivity in angiosarcoma (original magnification × 400).

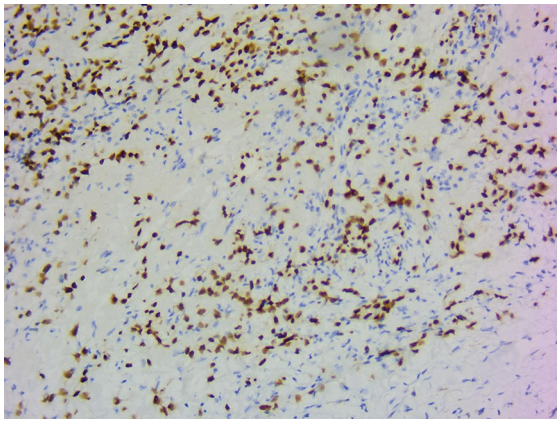

Figure 3. Tumor cells positive for E26 transformation-specific–related gene immunostaining (original magnification × 200).

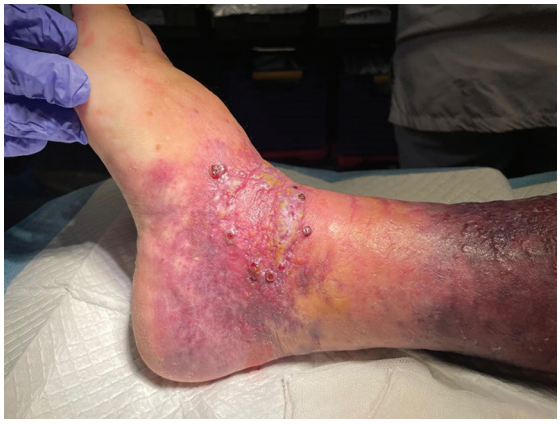

Figure 5. Confluent violaceous plaque with surrounding erythema and multiple papulonodular vascular lesions on the ankle and lower leg, some exhibiting central ulceration and serohemorrhagic crusting (classic Kaposi sarcoma).

Case 2

A 79-year-old Caucasian woman without immunosuppression presented with a 1-month history of progressive violaceous papules and nodules on the anterior aspect of the right lower leg and ankle, some with hemorrhagic crusting (Fig. 5).

Histopathological examination revealed irregular, thin-walled vascular spaces dissecting collagen bundles and a spindle-cell proliferation with mild-to-moderate atypia (Fig. 6). Immunohistochemistry demonstrated positive staining for HHV-8 in neoplastic cells, confirming the diagnosis of classic Kaposi sarcoma (Fig. 7).

While awaiting multidisciplinary evaluation, the patient developed a pulmonary thromboembolism requiring hospital admission and subsequently died. The etiology of the thromboembolic event was not determined.

Discussion

The distinction between these entities is crucial, as it determines radically different therapeutic strategies. Angiosarcoma requires an aggressive multimodal approach. For localized disease, wide surgical excision

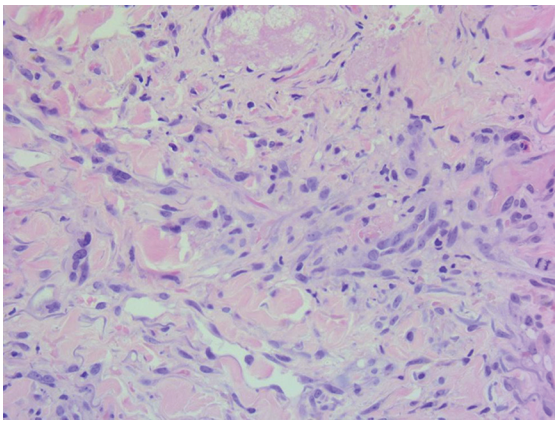

Figure 6. Classic Kaposi sarcoma with irregular thin-walled vessels and spindle-cell proliferation (hematoxylin and eosin stain, original magnification × 400).

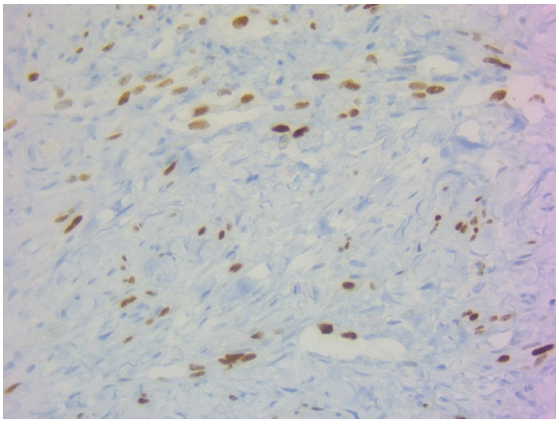

Figure 7. Positive human herpesvirus 8 immunostaining in Kaposi sarcoma (original magnification × 400).

is the primary treatment, with adjuvant radiotherapy recommended to improve local control. For advanced/metastatic disease, systemic chemotherapy (paclitaxel or anthracycline-based regimens) is the mainstay of treatment. Anti-angiogenic agents (e.g., pazopanib) and immune checkpoint inhibitors (e.g., pembrolizumab) represent additional options in select circumstances.[2] In contrast, classic Kaposi sarcoma may be managed with local therapies (options included cryotherapy, laser therapy, radiotherapy, and topical treatments such as 0.1% alitretinoin gel, 5% imiquimod cream, and topical timolol) for limited lesions or systemic agents (options included pegylated liposomal doxorubicin, paclitaxel, or immunomodulatory agents including pomalidomide) in cases of extensive disease.[3]

The cases presented herein clearly illustrate this diagnostic dilemma. In the first case, a 92-year-old man developed multiple ulcerated violaceous plaques and nodules on the left lower leg, with rapid progression over 2 months. Biopsy revealed a proliferation of epithelioid endothelial cells with marked cytological atypia and nuclear ERG expression, without HHV-8 immunoreactivity, confirming the diagnosis of cutaneous angiosarcoma. In the second case, a 79-year-old woman developed progressive violaceous papules and nodules on the right ankle and lower leg over 1 month. Histopathological examination demonstrated irregular vascular spaces dissecting collagen bundles and a spindle-cell proliferation with mild-to-moderate atypia, with positive HHV-8 staining, establishing the diagnosis of Kaposi sarcoma.[8,9]

Conclusion

These cases underscore the importance of early biopsy in atypical vascular lesions of the lower limbs, particularly in elderly patients. Adequate immunohistochemical characterization is essential to establish the correct diagnosis. Accurate differentiation between angiosarcoma and Kaposi sarcoma not only guides appropriate therapeutic decisions but also provides critical prognostic information for proper counseling of patients and their families.

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. The authors have followed their institution's confidentiality protocols, obtained informed consent from all patients, and secured approval from the Ethics Committee. SAGER guidelines have been followed as applicable to the nature of the study.

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.

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