ORIGINAL RESEARCH PAPER
Volume - 12 | Issue - 10 | October - 2023 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

KAPOSI'S SARCOMA MIMICKING PYODERMA GANGRENOSUM: A RARE CAUSE OF CHRONIC LEG ULCER LEADING TO THE DIAGNOSIS OF HIV

Dermatology

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ABSTRACT

Leg ulcers are the most common presentation of chronic wounds, usually resulting from venous disease and/or arterial disease. Pyoderma gangrenosum and malignancies may be rare causes of leg ulcers. Kaposi's sarcomas are vascular tumors associated with the human herpesvirus 8 (HHV-8). When affecting the skin, they usually present as small asymptomatic papules that vary in color from pink to purple. In this manuscript we report a case of a Kaposi Sarcoma that presented as a chronic leg ulcer, mimicking pyoderma gangrenosum and leading to the diagnosis of HIV.

KEYWORDS

leg ulcer, Kaposi's Sarcoma, Pyoderma Gangrenosum

INTRODUCTION

Leg ulcers are the most common presentation of chronic wounds.[1] As they may be caused many diseases, establishing their etiology is sometimes difficult. The majority of leg ulcers results from venous disease and/or arterial disease. Less frequent etiologies include pyoderma gangrenosum (PG) and malignant neoplasms, accounting, respectively, for 3% and 1,2% of chronic leg ulcers.[1]

Kaposi's sarcomas (KS) are vascular tumors associated with the human herpesvirus 8 (HHV-8).[2] They are the most common neoplasm in people living with HIV.[2] When affecting the skin, they usually present as small asymptomatic papules that vary in color from pink to purple.[2] In later stages, they may present as larger plaques and, occasionally, as exophytic tumors or ulcerated nodules. Presentations such as large chronic ulcers of the lower limbs are rare.[2,3] We report a case of a KS that presented as a chronic leg ulcer, mimicking pyoderma gangrenosum and leading to the diagnosis of HIV.

Case Study

A 34-year-old male sought medical attention due to a five-month painful ulcer in the left shin. It started as an asymptomatic nodule that evolved with pain and ulceration, progressing to a 10cm ulcer, with a necrotic bed and purulent discharge. Its borders were violet-colored, infiltrated, irregular, and undermined (Figure 1). He reported a sudden growth of the lesion after a surgical debridement performed a month prior to his admission.

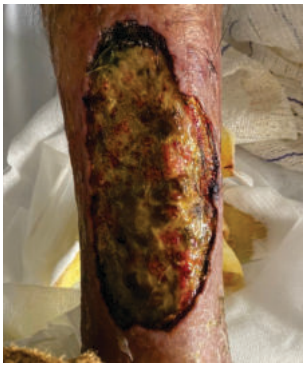

Figure 1. Ulcer in the lower left leg

He also reported a loss of 5kg (7% of his body weight) in the past 3 months. Blood tests revealed anemia (hemoglobin 9,2), leucopenia (1630 cells/mm^{3}), and lymphopenia (228 cells/mm^{3}). Although a violet-colored papule resembling KS (Figure 2) was noted on his left foot, the main hypothesis for the ulcer was pyoderma gangrenosum associated with myelodysplastic syndrome, due to his hematological abnormalities.

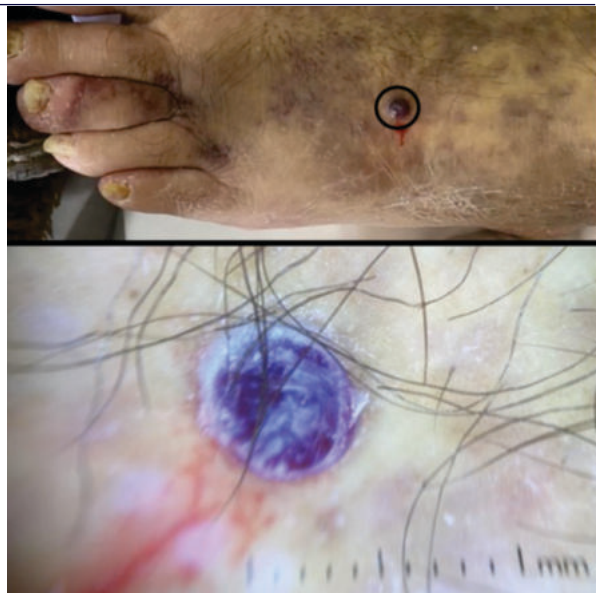

Figure 2. Macroscopic picture and dermoscopy of the papule on the left foot. Dermoscopy reveals a violet papule with white structures.

Anatomopathological examination of the ulcer revealed a vascular endothelial proliferation with ulceration and areas of necrosis with hemosiderin deposits. Immunohistochemistry analysis of the sample was positive for HHV-8, CD 34, and CD 31. There was no clinical evidence of opportunist infections, and cultures for mycobacteria, bacteria, and fungi were negative. The result was compatible with Kaposi's sarcoma. The biopsy of the papule in the left foot also confirmed KS.

The patient tested positive for anti-HIV (viral load of 1,656,046 copies/mL and CD4 count of 4 cells/mm^{3}). Total body imaging ruled out systemic involvement of KS, and the leg ulcer healed after the patient started antiretroviral therapy.

DISCUSSION

Pyoderma gangrenosum is a rare inflammatory skin disease. Its classic form presents as a painful nodule that evolves to an ulcer with an inflammatory undermined border and is frequently associated with the pathergy phenomenon (the development or aggravation of lesions after minor trauma).[4] The clinical appearance of the ulcer and the patient's report of worsening after debridement initially led to a diagnosis of PG. Ulcers are not common presentations of HIV-associated KS.[2] It usually presents in the skin as red papules and plaques that may progress to

larger plaques, often following the skin creases.[2,3] Only occasionally, it can develop exophytic, ulcerated or bleeding lesions.[2] KS lesions are typically painless, non-pruritic and do not produce necrosis in the overlying skin.[2,3] In our patient, the rapid progression to a painful ulcerated lesion was an atypical presentation, rarely described in patients with of HIV-associated KS. The correlation between the debridement and the reported worsening of the ulcer wasn't clear.

The purpose of this manuscript is to demonstrate that KS may also have atypical presentations, such as exuberant ulcers resembling a PG, and to reinforce that ulcerations should be promptly submitted to histological analysis when inflammatory conditions (e.g., PG, vasculitis, panniculitis), malignancies, or infections are in the differential diagnosis, especially in the context of HIV infection.

REFERENCES

1. Körber A, Klode J, Al-Benna S, et al. Etiology of chronic leg ulcers in 31,619 patients in Germany analyzed by an expert survey. J Dtsch Dermatol Ges. 2011;9(2):116-121. doi:10.1111/j.1610-0387.2010.07535.x
2. Cesarman E, Damania B, Krown SE, Martin J, Bower M, Whitby D. Kaposi sarcoma. Nat Rev Dis Primers. 2019 Jan 31;5(1):9. doi: 10.1038/s41572-019-0060-9. PMID: 30705286; PMCID: PMC6685213.
3. Schneider JW, Dittmer DP. Diagnosis and Treatment of Kaposi Sarcoma. Am J Clin Dermatol. 2017 Aug;18(4):529-539. doi: 10.1007/s40257-017-0270-4. PMID: 28324233; PMCID: PMC5509489.
4. Ruocco E, Sangiuliano S, Gravina AG, Miranda A, Nicoletti G. Pyoderma gangrenosum: an updated review. *J Eur Acad Dermatol Venereol*. 2009;23(9):1008-1017. doi:10.1111/j.1468-3083.2009.03199.x5. Amerson E, Woodruff CM, Forrestel A, Wenger M, McCalmont T, LeBoit P, Maurer T, Laker-Oketta M, Muyindike W, Bwana M, Buziba N, Busakhala N, Wools-Kaloustian K, Martin J. Accuracy of Clinical Suspicion and Pathologic Diagnosis of Kaposi Sarcoma in East Africa. J Acquir Immune Defic Syndr. 2016 Mar 1;71(3):295-301. doi: 10.1097/QAI.0000000000000862. PMID: 26452066; PMCID: PMC4770348.