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Generalized Kaposi's Sarcoma in an HIV Negative Patient: Case Report

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Abstract

BACKGROUND: Kaposi's Sarcoma is a vascular neoplasm associated with human herpesvirus 8 (HHV-8). It typically manifests with cutaneous lesions but can also involve extracutaneous sites such as mucous membranes, lymph nodes and visceral organs. Kaposi's Sarcoma is classified in 4 major subtypes: classic, epidemic, endemic and iatrogenic. The classic form usually follows a slow course, with progressions of cutaneous lesions over more than 10-15 years. However atypical cases of this form have been described in the literature. We aimed to present an atypical case of generalized Kaposi's Sarcoma in an HIV negative patient.

CASE REPORT: We report an atypical case of Kaposi's Sarcoma (KS) classic type, in a 62-year-old HIV-negative male, notable for its unusual cutaneous presentation, distribution, and rapid progression. The patient presented to the Emergency Department with asymptomatic violet-to-red cutaneous lesions, including both plaques and nodules, distributed across the upper and lower extremities, abdomen, and back. The lesions first appeared approximately eight months prior but demonstrated a fulminant course in the weeks leading up to presentation, with rapid dissemination. Initial home management of the patient included local and oral folk remedies of unknown composition. A skin biopsy performed during hospitalization revealed nodular Kaposi's Sarcoma, and immunohistochemistry (IHC) was recommended for diagnostic confirmation. HIV serology was negative, and imaging, including lymph node ultrasound, revealed no evidence of visceral involvement.

CONCLUSIONS: This case illustrates an atypical presentation of classic Kaposi Sarcoma (KS) in an immunocompetent, HIV-negative patient, characterized by rapid progression and diffuse cutaneous involvement. Such a clinical course is uncommon for the classic subtype, which is typically indolent and slowly progressive. Our findings support the growing body of evidence that subgroups within classic KS may exhibit distinct clinical behaviors, requiring tailored staging and management strategies. Given the absence of any known immunosuppressive conditions or identifiable triggers, further research is needed to elucidate the underlying pathophysiological mechanisms in atypical cases of classic KS.

Introduction

Kaposi Sarcoma (KS) is a painless, angioproliferative spindle cell tumor originating from endothelial and immune cells infected with Human Herpesvirus type 8 (HHV-8) [1].

While it is most commonly known for involving the skin and mucous membranes, KS may also spread via lymphatic routes and affect internal organs, particularly in severe cases, with lesions appearing in the gastrointestinal tract and lungs. On the skin, it typically presents as a purplish-red macule, which may become confluent and elevated into plaques over time [2].

There are 4 subtypes of KS: classic, epidemic, iatrogenic and endemic. Each subtype presents with its own distinct characteristics regarding the morphology of lesions, time of onset, and pattern of progression. The classic form of KS typically follows a slow and indolent clinical course, often extending over 10 to 15 years or more. Lesions tend to appear and expand very gradually, usually beginning in the lower extremities, and the development of new lesions occurs slowly over time.

Case Presentation

A 62-year-old male from Tirana, was admitted to the Dermatology Department of the University Hospital of Tirana, as an emergency case in July 2024, following an 8-month history of disease onset. During this period, he had been treated with local and oral folk remedies of unknown composition. The cutaneous lesions were asymptomatic, appearing as violaceous to reddish in colour. Clinically, some presented as plaques, while others appeared as nodules. The lesions were widespread, involving the lower (Fig. 1) and upper extremities (Fig. 2), abdomen (Fig. 3) and back.

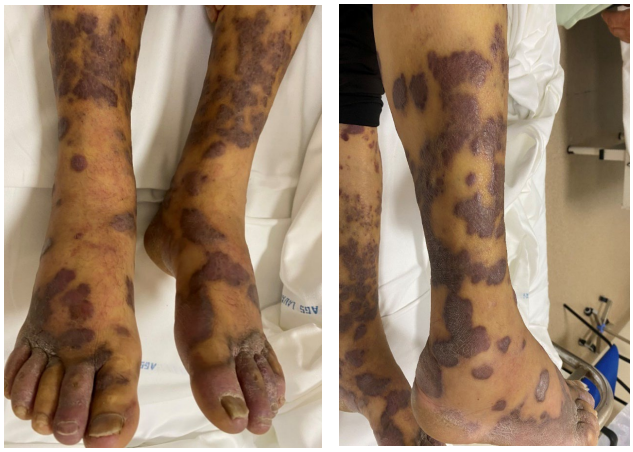

Figure 1: Red to violaceous macules, plaques and nodular lesions located in the lower extremities

The lesions demonstrated a rapid and widespread progression over a short period of time. Based on the clinical features, Kaposi Sarcoma (KS) was suspected, and a skin biopsy was performed for diagnostic confirmation. Histopathological findings included: a dermal spindle-cell proliferation arranged in vascular-like structures, showing moderate atypia and a low mitotic index, presence of mononuclear inflammatory infiltrates and siderophages (Fig. 4).

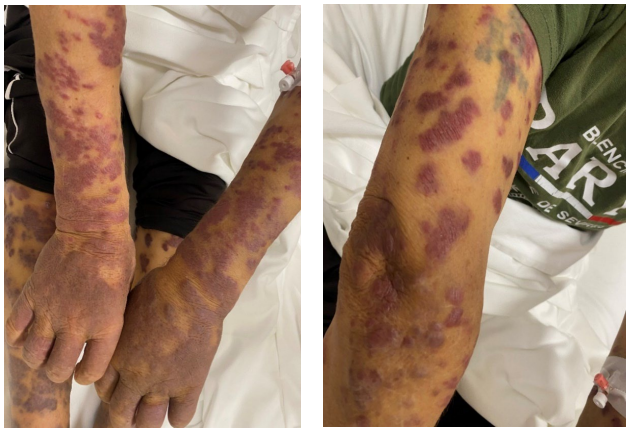

Figure 2: Red to violaceous macules, plaques and nodular lesions located in the upper extremities

This histopathological profile was suggestive of nodular-stage Kaposi Sarcoma. To confirm the diagnosis, immunohistochemical (IHC) analysis was recommended. The IHC results supported the diagnosis of KS and showed the following markers: Vimentin: positive, CD34: positive, CD31: positive, HHV-8: positive in endothelial cells, Ki-67: 10%.

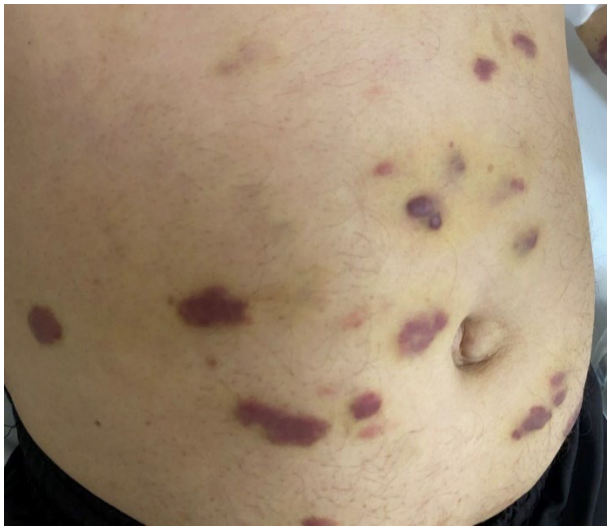

Figure 3: Kaposi lesions located at the abdomen

An ultrasound of the lymph nodes was also performed, revealing the following findings: non-reactive laterocervical lymph node (9 mm), supraclavicular lymph node on the left (8 mm), no paravascular adenopathy, reactive laterocervical (13 mm) and supraclavicular left (12 mm) lymph nodes, no right paravascular adenopathy. Not reactive axillary lymph nodes: left 18 mm, right 14 mm, no inguinal or para-aortic adenopathies bilaterally. Additionally, HIV testing was performed and returned negative. There was no evidence of visceral involvement.

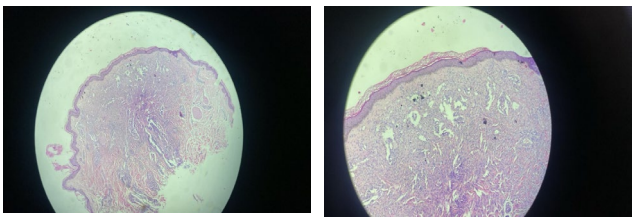

Figure 4: Dermal spindle-cell proliferation arranged in vascular-like structures, showing moderate atypia and a low mitotic index, presence of mononuclear inflammatory infiltrates and siderophages

The patient was referred to the Oncology Department for continuation of treatment. He underwent: 6 cycles of Doxorubicin (50 mg every two weeks), followed by 6 cycles of Paclitaxel (260 mg every two weeks). Compared to the previous CT there were no significant changes in the size of the laterocervical, mediastinal, or peri-aortic lymph nodes. However, infraclavicular left lymph nodes appeared reduced in size. Persistent cutaneous lesions were noted. Based on these findings, the patient was started

on a new treatment regimen with Vinblastine 10 mg, which he is currently continuing.

Discussion

Kaposi Sarcoma (KS) is a vascular neoplasm with both cutaneous and extracutaneous involvement. It is classified into four main clinical-epidemiological subtypes, each with distinct features in terms of lesion morphology, disease progression, and distribution [3], [4]. The classic subtype typically presents with slowly progressing nodules or plaques, most often located on the lower extremities, and primarily affects elderly men of Mediterranean origin. The endemic (African) form affects young adults and children in sub-Saharan Africa and may be locally invasive or involve visceral organs. Iatrogenic subtype occurs in organ transplant recipients receiving immunosuppressive therapy and often regresses after dose reduction or cessation. The epidemic (HIV/AIDS-related) subtype occurs predominantly in young men who have sex with men, with multifocal mucocutaneous lesions, rapid progression, and frequent visceral involvement [8], [9].

Although classic KS generally follows an indolent course over 10–15 years, atypical presentations have been increasingly reported. In our case, the patient demonstrated diffuse and rapidly progressing cutaneous lesions over a short period of time, without visceral involvement and HIV-negative status—a presentation not typical of classic KS. A study from the Department of Dermatology in Bari, conducted by Lospalluto, Mastrolonardo, Loconsole, Conte, and Rantuccio, proposed a classification of classic KS into three clinical subgroups based on cutaneous lesion dissemination. Their findings support the hypothesis that subgroups within classic KS exist, with distinct prognoses and treatment approaches [5]. According to this classification, our case corresponds to the HEG subgroup (High Extension and Growth), due to the rapid dissemination of lesions across multiple cutaneous regions. Another study from the Department of Dermatology in Istanbul, by Altunay et al., reported a rising number of clinically atypical KS cases [6]. For staging purposes, they utilized the Brambilla et al. classification (2003), which helps guide therapeutic decisions in classic KS [6]. Based on this system, our patient would be staged as classic KS, stage IVB.

One case in the Istanbul study involved an unusually aggressive course in a patient with classic KS, mimicking the epidemic (HIV-related) form clinically. This resembles our case as well, in which the diffuse cutaneous spread resembled a HIV-positive KS pattern, despite negative serology. In another case report from a Dermatology Department in Spain, a 52-year-old immunocompetent male with classic KS also presented with lesion characteristics typically seen in younger patients, adding further evidence for atypical variants of classic KS [7]. Notably, this patient reported same-sex sexual activity, a rare feature within the classic KS subtype, but more commonly associated with the epidemic form. This case report highlights the need for further investigation into the pathophysiological mechanisms that contribute to the development and progression of Kaposi Sarcoma in immunocompetent patients [7]. In our patient's case, the mechanisms underlying the rapid and diffuse cutaneous spread remain unclear. No identifiable etiological factor was reported during the clinical history that could explain such an aggressive course. The only possible contributing factor may be the use of non-medically supervised folk remedies, administered both topically and orally, with unknown composition. While a direct causative link cannot be confirmed, this highlights the importance of patient education and early referral to specialized care when skin lesions emerge.

Conclusion

This case illustrates an atypical presentation of classic Kaposi Sarcoma (KS) in an immunocompetent, HIV-negative patient, characterized by rapid progression and diffuse cutaneous involvement. Such a clinical course is uncommon for the classic subtype, which is typically indolent and slowly progressive. Our findings support the growing body of evidence that subgroups within classic KS may exhibit distinct clinical behaviors, requiring tailored staging and management strategies. Given the absence of any known immunosuppressive conditions or identifiable triggers, further research is needed to elucidate the underlying pathophysiological mechanisms in atypical cases of classic KS.

Additionally, this case underscores the importance of early dermatological evaluation and the potential risks associated with unsupervised alternative treatments in delaying diagnosis and appropriate care.

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