Case Report

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Atypical presentation of Kaposi sarcoma in an HIV-negative patient: a case report and comprehensive literature review

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Introduction: Kaposi's sarcoma (KS) is a systemic disease that is marked by the presence of neoplastic lesions caused by human herpesvirus-8 (HHV-8) infections. KS usually impacts people with weakened immune systems, although there have been a few cases of it occurring in individuals with normal immune function. Medical records and histopathological slides of the case were retrospectively reviewed. This work has been reported based on SCARE criteria.

Case Presentation: A 51-year-old man from Palestine came in with a single, painless, purple growth on his left forearm that had been growing quickly for 6 months. The patient did not have a history of using immunosuppressants, HIV infection, or engaging in unconventional sexual practices. Histopathological examination confirmed nodular-stage KS, with positive HHV-8 immunostain. The lesion was excised without complications, and the patient remains under periodic follow-up.

Discussion: KS typically manifests with multiple lesions in individuals with weakened immune systems. This case showcases a unique presentation in a patient with a strong immune system and no notable risk factors. Histopathological confirmation is necessary to differentiate between benign and infectious vascular lesions when considering the diagnosis of KS. The treatment approaches can differ depending on the extent of the lesion and the condition of the patient.

Conclusion: This case emphasizes the significance of considering KS as a potential cause for solitary vascular lesions, even in people who have a healthy immune system. It also emphasizes the need for a comprehensive diagnostic evaluation and personalized management.

Keywords: case report, human herpesvirus-8, immunocompetent, Kaposi sarcoma, solitary lesion

Introduction

Kaposi's sarcoma (KS) is a systemic disease characterized by the development of cutaneous and potentially visceral neoplastic lesions associated with human herpesvirus-8 (HHV-8) infections[1]. There are four primary subtypes of KS that have been recognized: classic (predominantly located in the lower extremities of elderly men from Mediterranean areas); endemic (in young Africans with frequent local invasive and/or visceral involvement); iatrogenic (associated with immunosuppressive drug therapy, typically reported in renal allograft recipients); and HIV-associated (epidemic)[2]. Recent literature has expanded this classification to include a non-epidemic form predominantly affecting HIV-negative men who have sex with men (MSM) and have no identifiable immunodeficiency[3,4]. While these subtypes encompass the majority of KS cases, atypical presentations have been documented; here, we display a new case of a solitary fore arm lesion in an immunocompetent adult male, a rare clinical manifestation according to existing literature.

This work has been reported based on SCARE criteria[5].

HIGHLIGHTS

- *Atypical presentation:* KS in a 51-year-old HIV-negative, immunocompetent male with a solitary forearm lesion.
- *Thorough diagnostics:* Histopathological and immunohistochemical analysis confirmed KS.
- *Effective treatment:* Successful lesion excision with no recurrence during follow-up.

Case presentation

A 51-year-old diabetic male patient from Palestine presented to our hospital with a 6-month history of a solitary, painless, and non-pruritic purple cutaneous mass lesion over the volar aspect of his left forearm. The lesion had rapidly progressed in size, measuring 1.8×1.5 cm in diameter at presentation. Notably, it began as a small nodule, not a patch, and grew rapidly (Fig. 1A).

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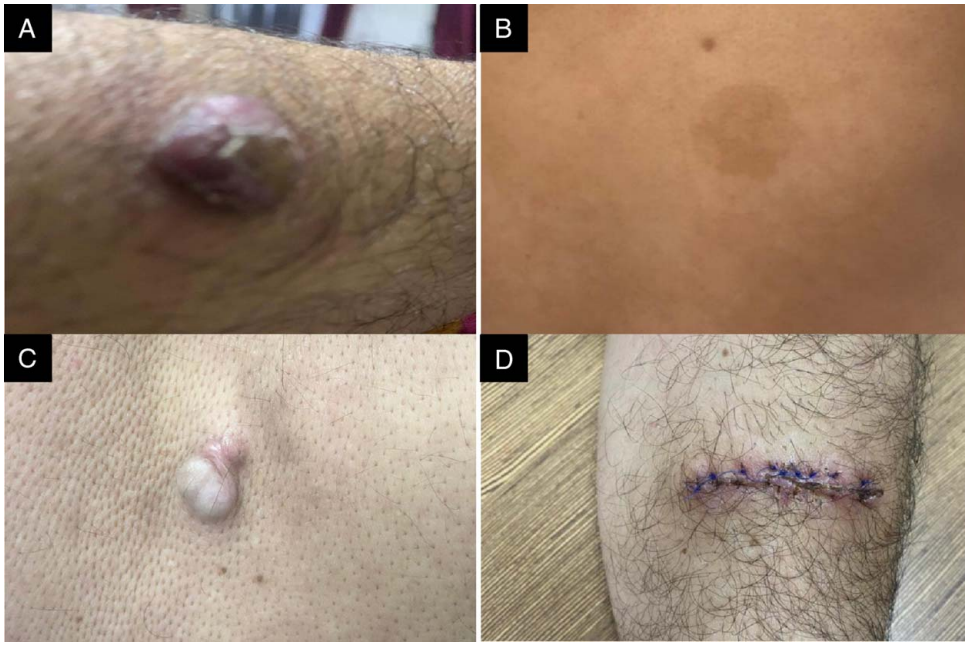

Figure 1. (A) Clinical photograph showing a purple nodular mass lesion over the volar aspect of the left forearm, size measuring 1.8×1.5 cm in diameter. (B) Clinical photograph showing a large hyperpigmented patch over the upper back. (C) Clinical photograph showing a pink, lobulated nodule over the upper back. (D) Clinical photograph showing the site of lesion after complete excision.

On examination, the patient was hemodynamically stable with a normal systemic examination. No lymph node enlargement was noted. A large hyperpigmented patch and a pink, lobulated nodule were observed on his upper back (Fig. 1B, C). He also had several small warts around his neck and back. There was no evidence of mucosal or visceral involvement. The patient denied using immunosuppressant medications, smoking, or illicit drugs. Additionally, he reported no family history of similar skin lesions and denied any unusual sexual practices, such as having sex with other men. The patient was generally well with no history of systemic illness or organ transplantation. Diagnostic workup, including chest, abdominal, and pelvic CT scans, was unremarkable. Blood tests, including differential blood count and B and T cell subcounts, were normal. Serological tests for HIV, hepatitis B surface antigen (HBsAg), and hepatitis C virus (HCV) were negative, while HHV-8 immunostain was strongly positive (Fig. 1D).

The patient reported a previous similar lesion 2 years ago on the left middle finger, which was excised and sent for histopathological evaluation. The results revealed features of pyogenic granuloma without evidence of malignancy. One year prior to the current presentation, a colonoscopy showed multiple polyps, including a large sigmoidal pedunculated polyp and two ascending colon polyps. All polyps were removed by hot snare and sent for pathology, which was unremarkable.

After discussing the lesion and considering its rapid growth, an excisional skin biopsy was performed. Sections revealed a well-circumscribed, dermal-based neoplasm composed of sheets of uniform spindle cells lacking nuclear pleomorphism with poorly formed slit-like vascular spaces. Foci of necrosis and high mitotic activity were observed. Extravasated red blood cells and intracytoplasmic hyaline globules were also present (Fig. 2A). The tumor cells were diffusely positive for CD31 immunostain (Fig. 2B). Based on clinical and histological findings, the diagnosis of nodular-stage Kaposi sarcoma was established. The patient underwent complete excision of the lesion without complications and is currently under periodic follow-up (Fig. 2C).

Discussion

Kaposi sarcoma is a vascular neoplasm characterized by nodular lesions primarily affecting the skin, with less frequent involvement of visceral organs. It represents the most common malignancy observed in patients infected with human immunodeficiency virus (HIV). The pathological basis of KS involves angiosarcomatous transformation within epithelial and mucosal connective tissues at various sites, including the face, neck, upper torso, lower extremities, genitalia, and lymph nodes. Initially described by Hungarian dermatologist Moritz Kaposi[1]. KS exhibits a marked male predominance with an approximate male-to-female ratio of 3:1. Notably, cases in individuals younger than 50 years are relatively uncommon[2].

The typical presentation for KS is characterized by the presence of multiple, pink-purple, angiomatous, rounded, well-defined, infiltrated maculopapules, which do not blanch or bleed on palpation. These lesions are painless, non-pruritic, have different sizes (0.5–2 cm) and are located symmetrically along the lines of skin tension[3]. However, our case deviates from the manifestation that has been described. We present here a new case of a middle-aged man who is HHV-8 positive, HIV negative, with no associated immunodeficiency and has no sexual practice with men. Who complained of a purple, solitary left forearm nodule.

The lesion presents initially as a patch stage that may gradually progress to a plaque stage, with a hyperpigmented appearance due to hemosiderin deposits, then to a nodular stage[6]. However, in this case, the patient reported an atypical presentation, with the

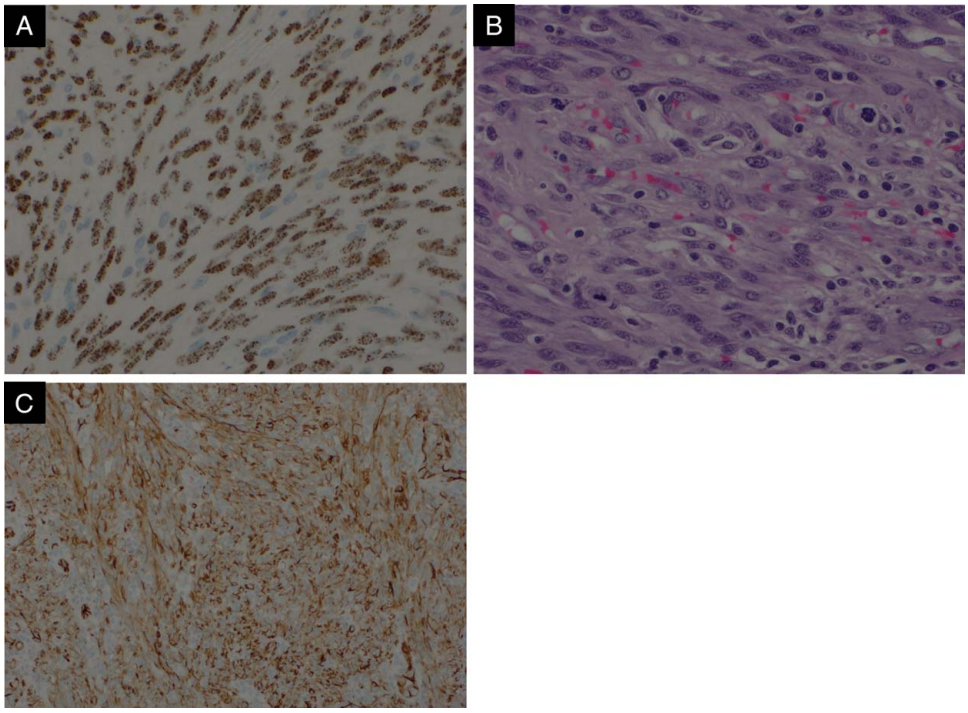

Figure 2. (A) Histopathological features showing sections of dermal-based neoplasm composed of monomorphic spindle cells separated by slit-like spaces with extraverted RBCs, intracytoplasmic hyaline globules, and high mitotic activity (H&E, 40×). (B) Histopathological features showing positive CD31 immunostain in the tumor cells (20×). (C) Histopathological features showing positive HHV-8 immunostain in the tumor cells (40×).

lesion directly progressing from a small nodule without an antecedent patch stage.

Differential diagnosis of cutaneous KS can be challenging due to overlapping features with benign and infectious vascular proliferative skin lesions. Bacillary angiomatosis, prevalent in certain regions, is a notable example of a condition that can mimic KS both visually and histologically. When resources allow, a skin biopsy with histochemical and immunohistochemical analysis remains the gold standard for definitive diagnosis.

The patch stage of Kaposi sarcoma (KS) is characterized by dilated, leaky vessels predominantly located in the reticular dermis, accompanied by a perivascular inflammatory infiltrate composed of lymphocytes and plasma cells. As the disease progresses to the plaque stage, there is diffuse dermal proliferation of spindle cells, forming irregular vascular spaces within a collagenous stroma. Extravasated red blood cells and hemosiderin deposition are common findings. The nodular stage is defined by a dense, well-circumscribed infiltrate of spindle cells replacing dermal collagen, with the formation of pseudo-vascular spaces containing red blood cells[7]. The definitive diagnosis of KS relies on the identification of human herpesvirus-8 (HHV-8), although additional vascular markers such as CD31, CD34, and D2-40 can be supportive[8]. In the present case, the patient was HHV-8 positive, HIV negative, and CD31 positive. Histopathological examination of the excisional biopsy confirmed a diagnosis of nodular-stage KS.

The management of Kaposi sarcoma (KS) encompasses a variety of therapeutic approaches, including conventional surgery, cryotherapy, laser therapy, intralesional injection of vinblastine or bleomycin, interferon-alpha, radiotherapy using a multiple-fraction regimen, and systemic chemotherapy with liposomal daunorubicin or paclitaxel[9]. In the present case, the patient has just one nodule that is completely excised when a biopsy has been taken, no additional treatment was required, and there has been no evidence of recurrence to date.

A review of the literature identified 12 published cases of isolated upper extremity Kaposi sarcoma, a rare presentation of the disease. The majority of these cases (7/12) involved a single lesion, and males were predominantly affected, with a male-to-female ratio of 3:1. Regarding HIV status, upper limb KS patients were HIV-negative (7/12), HIV-positive patients comprised (5/12) cases. The age of patients ranged from 26 to 74 years, with a mean age of 54.1 years (Table 1).

Conclusion

This case underscores the importance of considering Kaposi sarcoma (KS) in the differential diagnosis of solitary vascular lesions, even in immunocompetent patients. The unusual presentation of KS in an immunocompetent, HIV-negative patient with a lesion on the forearm – a rare site for KS – highlights the need for a thorough diagnostic evaluation, including histopathological and immunohistochemical analysis, to confirm the diagnosis. The successful excision of the lesion without complications emphasizes the potential for effective management in similar cases. Continued follow-up is essential to monitor for recurrence or the development of additional lesions. This case contributes to the growing body of literature on atypical presentations of KS and reinforces the necessity of awareness in diagnosing and managing rare manifestations of common diseases.

Table 1

Published cases who developed isolated upper limbs Kaposi lesions.

Case	Age	Sex	HIV status	Site	Number
Josefa Sánchez-López *et al.*[10]	52	M	Negative	Left forearm	Single
Dong Heon Lee *et al.*[11]	46	M	Positive	Right forearm	Single
Kiran Alam *et al.*[12]	60	M	Negative	Right forearm	Single
Elena Campione *et al.*[13]	61	M	Negative	Left forearm	Multiple
E. Dervis *et al.*[14]	54	W	Negative	Upper limb	Multiple
Irina Mamisoa Ranaivo *et al.*[15]	58	M	Positive	Right upper limb	Multiple
Christophoros Kosmidis *et al.*[16]	68	W	Negative	Left hand	Single
María Belén Castaño *et al.*[17]	31	M	Positive	Left forearm	Single
M.R. Ozbek *et al.*[18]	??	??	Positive	Hand palm	Single
John E. Keith Jr *et al.*[19]	26	M	Positive	Left small finger\left arm	Multiple
J.D. Witt *et al.*[20]	66	M	Negative	Palmar surface of the left ring finger\spread to involve the dorsum of the hand	Multiple
Allison L. Limmer *et al.*[21]	74	W	Negative	Right posterior upper arm	Single

Patient perspective

I am a 51-year-old man from Palestine with diabetes. Six months ago, I noticed a small, painless, purple bump on my left forearm that grew quickly. Tests showed it was not HIV, hepatitis B, or C, but positive for HHV-8, linked to Kaposi sarcoma (KS). The doctors confirmed it was nodular-stage KS. They removed the lesion without complications, and now I have regular check-ups. This experience taught me the importance of checking unusual skin changes, even if they seem minor. I'm thankful for the medical team's thorough care and effective treatment.

Ethical approval

Medical records of the case were retrospectively reviewed.

Consent

Written informed consent was obtained from the patient's parents/legal guardian for publication and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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Author contribution

N.E.M.J.: supervision; M.A.N.: data curation; M.J. and H.H.: review and editing; Y.Q. and R.M.: writing – original draft and data curation; H.A.S.: writing – original draft and review and editing.

Conflicts of interest disclosure

The authors declare no conflicts of interest.

Research registration unique identifying number (UIN)

None.

Guarantor

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Data availability statement

The datasets used in this report are available from the corresponding author on reasonable request.

Provenance and peer review

Not commissioned, externally peer-reviewed.

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