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# Kaposi's sarcoma in an HIV-negative chronic lymphocytic leukemia patient without immunosuppressive therapy: A case report

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## Abstract

Kaposi's sarcoma is a neoplasm of endothelial cells. That vascular tumor is usually limited to the skin, but it may involve mucous membranes, visceral organs, and lymph nodes. Serological evidence has shown that human herpesvirus 8 infection is required for the development of Kaposi's sarcoma. Chronic lymphocytic leukemia is the most common leukemia all over the world. Increased skin cancer risk has been reported for patients with chronic lymphocytic leukemia. The relation between these two pathologies has not yet been clarified. We report a case of Kaposi's sarcoma along with chronic lymphocytic leukemia in a patient who did not receive therapy for chronic lymphocytic leukemia.

## Keywords

Chronic lymphocytic leukemia, HIV-negative, Kaposi's sarcoma

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## Introduction

Kaposi's sarcoma (KS) is a low-grade malignant angioproliferative tumor, typically manifesting multifocal lesions in mucocutaneous sites like the skin of the lower extremities, trunk, genitalia, gastrointestinal, and respiratory tract. It was first described in five male patients by Moritz Kaposi,<sup>1</sup> one of the leading dermatologists of that time, in 1872. Only after more than a century, Yuan Chang et al.<sup>2</sup> found a new DNA virus in the tissue of KS. Today, it is well-known that KS is associated with gamma human herpesvirus 8 (HHV-8) or Kaposi sarcoma-associated herpesvirus (KSHV) infection of HIV-positive or otherwise immunocompromised patients.<sup>3</sup>

Chronic lymphocytic leukemia (CLL) presents with an increased number of dysfunctional lymphocytes of monoclonal origin located in the bone marrow, peripheral blood, and the reticuloendothelial system.<sup>4</sup> Herein, we present a case of a disseminated KS in a non-transplant and HIV-negative patient, who developed a secondary neoplasm during the course of CLL.

## Case report

A 78-year-old male patient was admitted complaining of redness, itchiness, straining sensation, and swelling in the lower legs. He described that the affected area spread from

the lower legs to the trunk in the last 30 days. Dermatological examination revealed multiple livid papules on the feet and lower legs medially converging with grayish-black plaques (Figure 1). Single papules were visible on the upper trunk and proximal part of the upper extremities (Figure 2). The patient's past medical history showed that he was diagnosed with CLL 7 years ago. In bone marrow biopsy, there was increased cellularity characterized by small lymphocytes. Positivity for CD5 and CD23 indicated the diagnosis of CLL. CLL was staged as Rai 0. The disease was regularly monitored by the hematologist and did not require treatment. Laboratory findings at admission were as follows: hemoglobin 118 g/L, platelet  $212 \times 10^9/L$ , leukocyte

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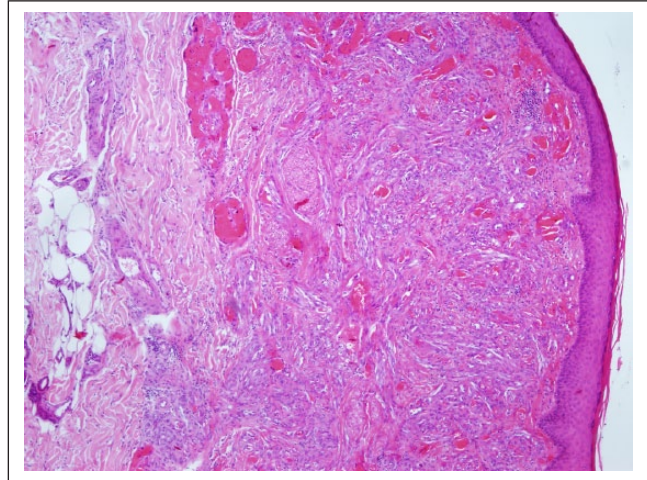


**Figure 1.** Purplish hyperkeratotic plaque.

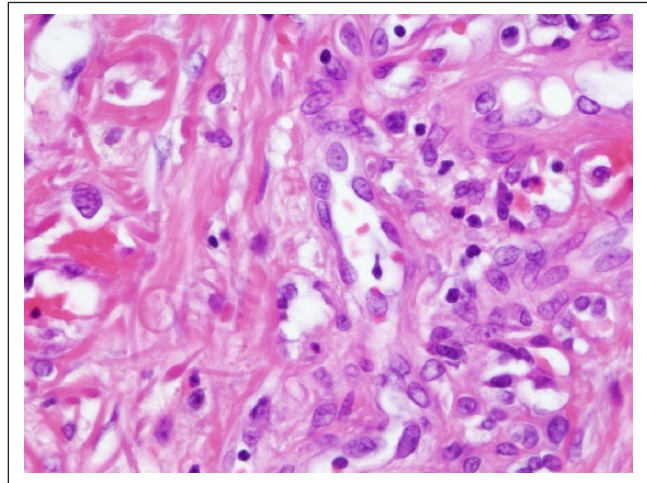


**Figure 2.** Bluish and indurate lesions on upper trunk.

$38.6 \times 10^9/L$ , and lymphocyte  $32.4 \times 10^9/L$  with Gumprecht shadows present. Abdominal ultrasonography did not show a large spleen or enlarged lymph nodes. The patient was seronegative for HIV and hepatitis B and C. Regular hematological checkup was indicated. Systemic therapy with antihistamines and corticosteroids led to major regression of dermatological symptoms. Multiple papules persisted. Initial differential diagnosis included angioma, pigmented purpuric dermatosis, hemangioendothelioma, and KS. Two papules from the lower leg and trunk were excised and sent



**Figure 3.** Skin biopsy specimen: vascular tumor in dermis (HE 40 $\times$ ).



**Figure 4.** Lesion tissue composed of vascular spaces with atypical endothelium, among which are spindle cells and extravasated erythrocytes (HE 100 $\times$ ).

for a biopsy. Microscopic examination revealed the presence of spindle-cell nodules in the dermis separated by irregular vascular spaces coated with atypical endothelium. Erythrocytic extravasation was present (Figures 3 and 4). DNA isolated from the biopsy tested by polymerase chain reaction (PCR) for HHV-8 yielded a positive result. Obtained findings all fit in the diagnosis of disseminated KS of the nodular type. After oncology consultation, radiotherapy was recommended. However, the patient refused to continue the treatment.

## Discussion

Diagnosis of KS is generally based on clinical and histopathological features, but in some difficult cases, demonstration of HHV-8 in situ can confirm the diagnosis. Clinically,

the lesions can include patches, papules, and plaques and can be red, brown, or bluish in color. The histopathology varies with the type of lesion: macules, papules, nodules, or plaques. However, it is typically the presence of slit like spaces, spindle cells with frequent mitoses which demonstrate endothelial differentiation by immunohistochemistry. Today, KS can be subdivided into four clinical and epidemiological types. The classic or sporadic type observed mostly in elderly men of Mediterranean or Eastern European origin, starts mostly on the skin of the lower extremities. The endemic type is observed frequently in East and Central Africa and commonly seen in adults but also in children, and it is more aggressive. Furthermore, the iatrogenic type is observed frequently in patients after solid organ transplant (especially renal) or patients under long-term immunosuppressive treatment for other diseases. The AIDS-associated KS is very aggressive, frequently with oral lesions, and develops in the immunosuppressive phase when there is a clear decline of CD4 count. After the introduction of the HAART therapy this tumor became rarer.<sup>5</sup> In 1994, a new DNA virus in KS was found. It was a virus of the genus rhadinovirus. Some of its genes are homologous to human genes important in the regulation of cell proliferation, angiogenesis, and apoptosis. It was found later that the virus was present in all types of KS.<sup>6</sup> New studies have revealed some aspects of the pathogenesis: the virus enters various cells (B lymphocytes, dendritic cells, monocytes, and endothelial cells) through endocytosis and causes a lifelong infection—latency that evades the immune recognition by the host.<sup>3</sup> The viral latency-associated nuclear antigen (LANA) plays an important role in that process. Only the presence of co-factors such as immunosuppression, co-infections, inflammation, anoxia, and perhaps quinine can lead to activation and neoplastic transformation (lytic phase) with exposure of many genes. This virus was also found in multicentric Castleman disease and primary effusion lymphoma.<sup>5</sup> CLL is characterized by progressive defects in both cell-mediated and humoral-mediated immunity. Development of second nonlymphoid neoplasms in CLL patients has been linked to abnormalities in T-cell function. Cytokine production abnormalities were found in these patients. Moreover, T cells in B-CLL may be unable to complete an immune response to the antigens presented by malignant B cells and other antigens.<sup>7–9</sup> The relationship between immunosuppression and KS is well documented in the literature, but other co-factors must also be considered.

## Conclusion

We describe a case of KS in an HIV-negative patient with CLL without immunosuppressive therapy. A few similar Kaposi sarcoma cases are reported in the literature, but most patients in the reports have had immunosuppression factors.<sup>10–12</sup> Sometimes, it is difficult to differentiate the disease from other lesions such as acroangiodermatitis, hemangioendothelioma, angiosarcoma, pyogenic granuloma, angiohistiocytoma, melanoma, and others.

The physician must be vigilant to detect early neoplastic lesions, especially KS, even in a CLL patient without active therapy or HIV infection.

## Declaration of conflicting interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

## Ethical approval

Our institution does not require ethical approval for reporting individual cases or case series.

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## Informed consent

Written informed consent was obtained from the patient(s) for their anonymized information to be published in this article.

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