

Multisystem disseminated kaposi sarcoma in untreated advanced HIV infection

Abstract

Kaposi Sarcoma (KS) is an AIDS-defining malignancy associated with human herpes virus 8 infection and advanced immunosuppression. Although its incidence has declined with antiretroviral therapy, disseminated disease remains a clinical challenge in untreated Human Immunodeficiency Virus (HIV) infection. We report a case of multisystem disseminated KS in a 28-year-old man with untreated advanced HIV infection presenting with progressive cutaneous lesions, gastrointestinal bleeding, and severe thrombocytopenia. Imaging revealed pulmonary infiltrates, pleural effusions, and widespread lymphadenopathy. The patient developed acute abdomen requiring appendectomy, and histopathology demonstrated acute appendicitis with atypical vascular proliferation suspicious for KS. Despite supportive management and initiation of antiretroviral therapy, the patient developed progressive multiorgan dysfunction and died during hospitalization.

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Received: Sep 05, 2026; **Accepted:** Oct 05, 2026;

Published: Oct 12, 2026

Citation: Mendonça Nicolau LF, Martinho Nicolau ZF, Moura L, Neves Nicolau R, Correa Moreira S, et al. Multisystem disseminated kaposi sarcoma in untreated advanced HIV infection. *Ann Case Rep Med Images*. 2026; 3(2): 1101.

Keywords: Kaposi sarcoma; HIV infections; Acquired immunodeficiency syndrome.

Introduction

Kaposi Sarcoma (KS) is a cytokine-mediated antiproliferative malignancy. Multiple factors have been implicated, and epidemiologic, serologic, and molecular studies support an association with human herpesvirus type 8 (HHV8) infection [1]. Individuals with HIV infection have an increased risk of developing KS, an AIDS-defining malignancy associated with advanced immunosuppression [2].

Although Kaposi sarcoma may be suspected clinically, definitive diagnosis relies on histopathological confirmation, typically characterized by proliferation of spindle-shaped cells [3]. Antiretroviral therapy plays a central role in the management of AIDS-related Kaposi sarcoma by promoting immune reconstitution and suppression of HIV replication, although some patients may experience clinical worsening due to Immune Reconstitution Inflammatory Syndrome (IRIS) [4].

We report a case of untreated advanced HIV infection presenting with disseminated KS and a complex multisystem clinical course characterized by severe cytopenia's, pulmonary involvement, extensive lymphadenopathy, and acute abdomen requiring urgent surgical intervention.

Case presentation

A 28-year-old man with recently diagnosed Human Immunodeficiency Virus (HIV) infection and no prior exposure to Antiretroviral Therapy (ART) presented with a three-month history of progressive violaceous popular skin lesions involving the anterior and posterior trunk as well as the upper and lower extremities. The lesions were non-tender and gradually increased in number and size. During the same period, he developed constitutional symptoms including unintentional weight loss, intermittent evening fever, and productive cough with episodes of hemoptysis. He also reported progressive swelling of the right lower limb.

The patient was admitted after developing gastrointestinal bleeding manifested by melena and hematemesis, associated with recurrent epistaxis. On initial evaluation, he was tachycardic but hemodynamically stable. Laboratory testing revealed anemia, leukocytosis with neutrophilia, elevated inflammatory markers, and marked thrombocytopenia. Previous laboratory evaluation had confirmed advanced HIV infection with a CD4 count of 159 cells/ μ L and a plasma HIV viral load of 167,764 copies/mL.

Physical examination revealed numerous violaceous papules and plaques distributed over the trunk and extremities, diffuse ecchymoses, bilateral subconjunctival hemorrhage, and palpable cervical and inguinal lymphadenopathy. There was marked asymmetric edema of the right lower limb. Doppler ultrasonography excluded deep venous thrombosis but demonstrated enlarged inguinal lymph nodes with reactive features.

Computed Tomography (CT) of the chest demonstrated multifocal areas of consolidation associated with ground-glass opacities predominantly involving the upper lobes and scattered in the lower lobes, suggestive of an inflammatory or infectious pneumopathy. Abdominal CT revealed diffuse abdominal lymphadenopathy and inflammatory changes in the pericaval region, including appendiceal wall thickening and surrounding fat stranding compatible with acute appendicitis. Given the imaging findings and the presence of abdominal pain, the patient underwent urgent appendectomy and was subsequently admitted to the intensive care unit for postoperative monitoring. Histopathological examination of the surgical specimen demonstrated acute transmural appendicitis with edema and vascular congestion. In addition, an atypical vascular proliferation composed of irregular and anastomosing vascular spaces with mildly enlarged endothelial cells was identified within the appendiceal wall. The proximal surgical margin was free of lesion. These findings raised suspicion for vascular neoplastic involvement, and additional immunohistochemical evaluation was recommended.

Histopathological examination of a previously obtained skin biopsy demonstrated dermal vascular proliferation composed of spindle-shaped and epithelioid cells forming irregular vascular channels, findings compatible with Kaposi Sarcoma (KS). A subsequent biopsy of an inguinal lymph node revealed architectural distortion due to proliferation of spindle cells arranged in fascicles and irregular bundles with eosinophilic cytoplasm and elongated nuclei. These cells formed aggregates associated with slit-like vascular spaces containing extravasated erythrocytes. Mild nuclear atypia and occasional mitotic activity were observed, along with areas of fibrosis, hemosiderin deposition, and small vascular thrombi. The morphologic findings were suggestive of KS.

During hospitalization, the clinical course was marked by persistent and severe thrombocytopenia associated with recurrent bleeding manifestations, including epistaxis, hemoptysis, gingival bleeding, and gastrointestinal hemorrhage, requiring repeated transfusions of platelets and packed red blood cells. Extensive microbiological investigation for opportunistic infections was performed. Sputum smear microscopy for acid-fast bacilli and molecular testing for *Mycobacterium tuberculosis* (Expert MTB/RIF) were negative. Subsequent bronchoscopy with bronchoalveolar lavage was likewise negative for acid-fast bacilli, mycobacteria by molecular testing, and fungal pathogens. Serologic testing showed IgG

reactivity for cytomegalovirus, herpes simplex virus, Epstein-Barr virus, parvovirus B19, and *Toxoplasma gondii*, without evidence of acute infection. Tests for *Histoplasma capsulatum*, HTLV-1/2, syphilis, and hepatitis B and C were negative. Upper gastrointestinal endoscopy revealed severe congestive and hemorrhagic pangastritis with moderate congestive bulb duodenitis. During the subsequent hospital course, the patient developed progressive dysphagia and odynophagia associated with oral candidiasis.

Despite supportive management and antimicrobial prophylaxis for opportunistic infections, the patient experienced progressive clinical deterioration. Follow-up imaging demonstrated widespread lymphadenopathy involving mediastinal, axillary, retroperitoneal, mesenteric, and inguinal regions. Chest imaging also revealed bilateral pleural effusions with compressive atelectasis, a small pericardial effusion, and persistent diffuse pulmonary abnormalities characterized by centrilobular micronodules and multifocal ground-glass opacities. Abdominal imaging demonstrated small-volume ascites, diffuse subcutaneous edema, and persistent abdominal lymphadenopathy.

Bone marrow evaluation revealed markedly hypercellular marrow with granulocytic predominance and dysplastic megakaryopoiesis, erythroid hypoplasia, and absence of increased CD34- or CD117-positive precursor cells. Scattered mature lymphocytes with mixed B- and T-cell phenotype and a small population of plasma cells were identified, without evidence of hematologic malignancy.

Antiretroviral therapy was initiated after clinical stabilization. However, the patient subsequently developed progressive anasarca, worsening cytopenia's, and respiratory distress associated with large bilateral pleural effusions. Despite intensive supportive care, his condition evolved to progressive multiorgan dysfunction. Following discussions with the patient and family, goals of care were transitioned to palliative management, and the patient ultimately died during hospitalization.

Discussion

Although the incidence of AIDS-related KS has declined with widespread use of antiretroviral therapy, disseminated disease continues to occur in patients presenting with untreated or advanced HIV infection. Such cases may involve multiple organs simultaneously and present with complex systemic manifestations [5].

Pulmonary abnormalities represented an important component of the clinical presentation in this patient. Chest computed tomography demonstrated multifocal ground-glass opacities, micronodular infiltrates, and pleural effusions. In patients with advanced HIV infection, these findings frequently raise concern for opportunistic infections. However, pulmonary KS is also a well-recognized manifestation of visceral disease. Radiologically, pulmonary KS typically involves the bronchovascular interstitial and pleura, producing findings such as peribronchovascular and interlobular septal thickening, ground-glass attenuation, ill-defined nodules, mediastinal adenopathy's, and pleural effusions [1]. Peribronchovascular infiltration reflects tumor growth, whereas septal thickening and ground-glass opacities may result from edema, tumor infiltration, or alveolar hemorrhage [1]. These patterns may resemble infectious pneumonias, which often complicates the diagnostic evaluation in immunocompromised patients.

In this case, extensive microbiological investigation including bronchoscopy with bronchoalveolar lavage did not identify mycobacterial or fungal pathogens. The coexistence of disseminated cutaneous and nodal KS together with compatible radiologic findings strongly suggests pulmonary involvement by KS.

Gastrointestinal involvement also played a significant role in the clinical course. The gastrointestinal tract is the most common extracutaneous site of disease in AIDS-associated KS and may remain asymptomatic [6]. When symptomatic, patients may present with weight loss, diarrhea, abdominal pain, or gastrointestinal bleeding [7]. Endoscopic findings are heterogeneous and may include erythematous lesions, nodules, polypoid masses, or maculopapular lesions [8]. In this case, upper gastrointestinal endoscopy revealed severe hemorrhagic pangastritis and duodenitis, findings that may reflect mucosal vascular involvement and inflammatory changes related to KS infiltration.

The presence of imaging findings compatible with acute appendicitis represented an additional diagnostic challenge. Although acute appendicitis is common in the general population, gastrointestinal involvement by KS has been reported throughout the digestive tract, including rare cases involving the appendix [9]. In patients with advanced HIV infection and disseminated KS, abdominal inflammatory presentations may therefore reflect infectious, inflammatory, or neoplastic processes. Differentiating these etiologies may be difficult in the acute setting and often requires surgical or histopathological evaluation.

Severe thrombocytopenia and recurrent bleeding manifestations represented another major complication in this patient. HIV-induced immune and inflammatory mechanisms can promote platelet activation and accelerated clearance, contributing to thrombocytopenia and hypercoagulability [10]. In addition, HIV infection may affect the hematopoietic system by impairing bone marrow function through direct effects on hematopoietic stem and progenitor cells as well as alterations in the bone marrow microenvironment. Virus-induced inflammatory mediators and immune responses may further disrupt hematopoiesis, leading to reduced bone marrow output [11]. In the present case, bone marrow examination revealed hypercellular marrow with dysplastic megakaryopoiesis and erythroid hypoplasia.

Another important aspect of this case was the extensive lymphatic involvement observed during the clinical course. The patient developed widespread lymphadenopathy involving cervical, inguinal, mediastinal, and retroperitoneal regions, in addition to progressive lower limb edema and later generalized anasarca associated with pleural and pericardial effusions. Kaposi sarcoma involves proliferation of lymphatic endothelial spindle cells that can replace nodal parenchyma and obstruct lymphatic lumina, impairing lymphatic drainage [12]. Large cutaneous lesions may also externally compress superficial lymphatic vessels. These complications are more frequent in patients with high tumor burden and multicentric disease. Extensive lymphedema, nodal or visceral involvement, and serous effusions are markers of advanced disease and poor prognosis in KS staging [13].

ART represents the cornerstone of treatment for HIV-associated KS, as immune reconstitution is a key component

of disease control [14]. However, KS may recur even at high CD4 T-cell counts [15]. An important consideration is the potential development of IRIS following ART initiation. IRIS arises from rapid expansion of antigen-specific CD4⁺ and CD8⁺ T lymphocytes, accompanied by an imbalance between pro- and anti-inflammatory immune responses [16]. The risk of IRIS is higher in individuals with low CD4 cell counts and elevated viral load, and the occurrence of IRIS has been associated with increased mortality compared with patients who do not develop the syndrome [4].

This case highlights the aggressive and multisystem nature that Kaposi sarcoma may assume in the context of untreated advanced HIV infection. Disseminated disease may involve multiple organs simultaneously and produce overlapping clinical manifestations affecting the respiratory, gastrointestinal, hematologic, and lymphatic systems. Recognition of these severe presentations remains essential for early diagnosis and coordinated multidisciplinary management. In patients with advanced HIV infection and disseminated KS, clinicians should maintain a high index of suspicion for visceral involvement and systemic inflammatory complications.

Declaration of interest: The authors declare that they have no conflicts of interest, including any financial, personal, or professional affiliations that could have influenced the work reported in this case report.

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