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A 19 YEAR OLD MALE WITH HIV PRESENTS WITH DIFFUSE LYMPHADENOPATHY

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Background

In 1872, Moritz Kaposi first described “an idiopathic multiple pigmented sarcoma of the skin,” now identified as Kaposi’s sarcoma (KS).¹ While multiple forms of KS exist, over 95% of the cases diagnosed in the US since 1981 are of the AIDS associated variety.² Kaposi originally described KS as skin lesions that can progress to visceral involvement. However, in a small number of cases, KS can appear in the viscera without skin involvement. These alternate presentations of KS are difficult to diagnose; therefore, it is critical to recognize them when considering differential diagnoses, particularly in patients with HIV.

Case Presentation

An 18-year-old African American male with a history of HIV presented with progressive worsening of diffuse and painful lymphadenopathy for five weeks prior to admission. The patient was diagnosed with HIV in 2010 and due to insurance issues, was never treated with highly active antiretroviral therapy (HAART). His last CD4 count (approximately two weeks prior to admission) was 411 and he had no history of opportunistic infections. He first noticed swelling in his neck, under his armpits and in his groin five weeks prior, which had become progressively more painful. The patient denied fevers, chills or weight loss, but did report significant night sweats and episodes of hemoptysis with clots. He denied shortness of breath or chest pain. He also denied recent travel, history of incarceration, homelessness or exposure to active tuberculosis infection.



Figure 1. CT of the chest with IV contrast demonstrates ground glass opacity in the right upper lobe and bulky enhancing adenopathy in the bilateral axilla.

On admission, the patient’s physical exam revealed numerous palpable, mobile and tender lymph nodes in the cervical, axillary and inguinal regions bilaterally. There was no overlying erythema and no skin lesions noted elsewhere. His lungs were clear to auscultation bilaterally. The remainder of his physical exam was within normal limits. His labs upon admission were notable for a hemoglobin of 9.4 and a platelet count of 9,000. The remainder of his labs were within normal limits. A chest x-ray demonstrated no consolidation or infiltrates.

Hospital Course

The differential diagnosis for the HIV patient presenting with diffuse lymphadenopathy includes lymphoma, mycobacterium avium intracellular infection, tuberculosis with extrapulmonary involvement, fungal infection, Castleman’s disease, Kaposi’s Sarcoma, leukemia, infectious mononucleosis secondary to EBV or CMV, Bartonella infection (cat scratch disease), toxoplasmosis and secondary syphilis. Malignancy is a significant concern with the co-existing anemia and thrombocytopenia. He underwent a CT of the chest (Figure 1), abdomen and pelvis (Figures 2 and 3). Given his complaints of hemoptysis, the patient was placed on respiratory isolation and TB was ruled out by sputum acid fast bacilli stain and culture.



Figure 2. CT of the abdomen with IV contrast shows bulky enhancing intra- abdominal and para-aortic lymph nodes.

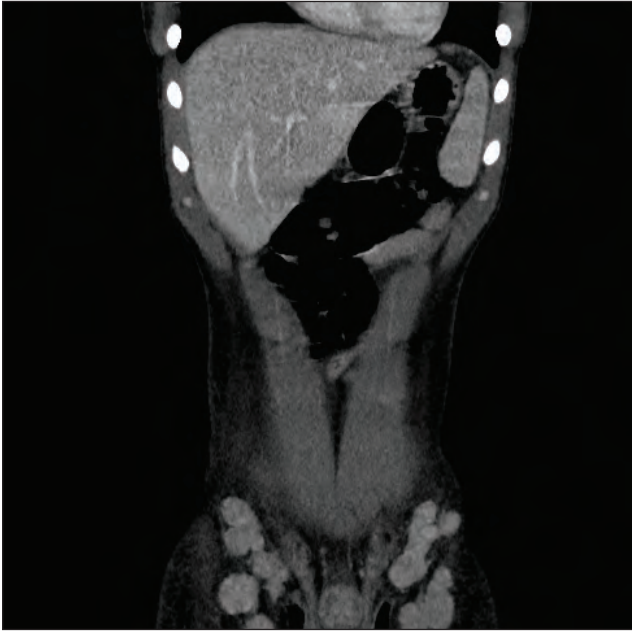


Figure 3. CT of the abdomen and pelvis demonstrates bilateral bulky enhancing inguinal lymph nodes.

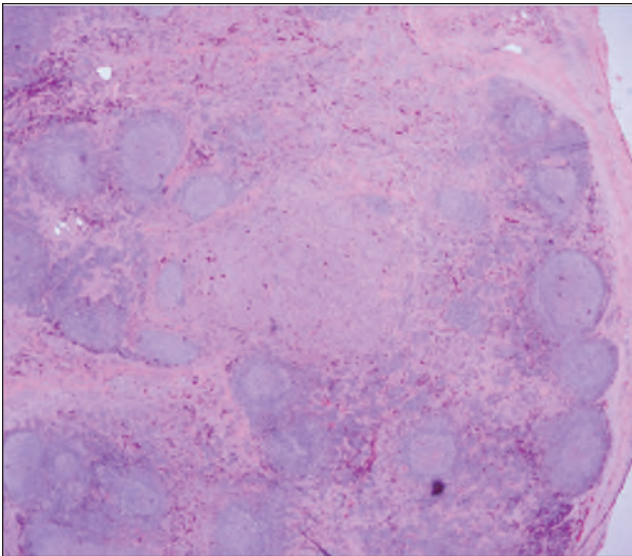


Figure 4. Low power H&E stain of excised left inguinal lymph node demonstrates distortion of the normal lymph node architecture.

Rapid plasma reagin and monospot tests were also negative and repeat CD4 count was 194. The history, laboratory results and radiological findings required excisional lymph node biopsy for definitive diagnosis, but this was limited by the patient's severe thrombocytopenia. Hematology was consulted and determined the thrombocytopenia likely represented HIV associated ITP,

for which he was treated with IVIG and started on HAART. He was started on romiplostim to stimulate platelet production.

On day 4 of his admission, the patient underwent a left inguinal lymph node excisional biopsy. Pathology was consistent with KS (Figures 4-7). It was assumed that his hemoptysis represented pulmonary involvement, although this could not be confirmed by bronchoscopy in the setting of thrombocytopenia. Although first line treatment for HIV-associated KS is HAART, the patient's extensive and symptomatic disease required additional therapy. He received radiation for his bulky cervical adenopathy; however, he developed stridor during treatment and was intubated for airway maintenance. His ICU course was complicated by ventilator acquired pneumonia and he required a tracheostomy for continued airway maintenance. He was eventually weaned from the ventilator and started on liposomal doxorubicin on the general floor. After a nearly two month hospitalization, he was deemed stable for discharge home with a plan to continue HAART, weekly romiplostim, and chemotherapy as an outpatient.

The patient was seen for follow up three weeks after discharge. His hemoptysis had resolved and his lymphadenopathy had decreased in size and was less painful. He continued to receive doxorubicin every other week and he required romiplostim for persistent thrombocytopenia. He no longer required supplemental oxygen and ENT planned to decannulate his tracheostomy.

Discussion

The Hungarian dermatologist Moritz Kaposi first described classic KS in 1872, as a rare, slow growing cutaneous tumor that mainly affects 50 to 70-year-old Jewish Mediterranean and Eastern European males. Since then, other varieties have been described including the African or endemic, and the immunosuppressed forms.³ In 1981, a fulminant and disseminated form of the disease appeared alongside HIV/AIDS. In fact, early in the epidemic, 48% of AIDS patients in the US presented with KS.² KS typically begins with cutaneous lesions, which are usually dark patches, papules, plaques or nodules. Diagnosis requires biopsy with histopathology illustrating a multicentric angioproliferative spindle cell tumor that stains positive for the endothelial markers CD31 and CD34. Positive stain for HHV-8 is also necessary but not sufficient for diagnosis, as the virus is also associated with multicentric Castlemans disease and primary effusion lymphoma in HIV patients.

Although the vast majority of patients with KS present with skin lesions, the disease can involve the viscera, most commonly the oral mucosa, lung, liver, spleen, lymph nodes and the GI tract. Patients in later stages of disease can also experience fevers, weight loss and night sweats. Visceral involvement usually progresses from cutaneous disease, but in a small number of cases, it can be the primary site of involvement. This is seen

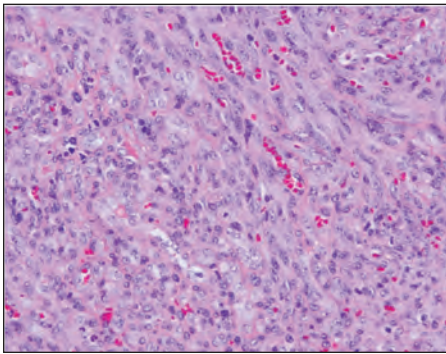


Figure 5. High power H&E stain of excised left inguinal lymph node with infiltration by angioproliferative spindle cells

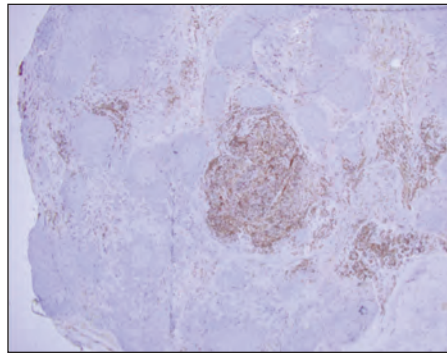


Figure 6. Low power CD34 positive stain of excised left inguinal lymph node. Positive stain for CD 34 indicates vascular endothelium. Normal lymphatics do not stain positive for CD34.

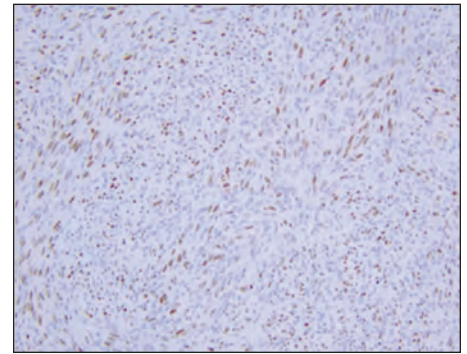


Figure 7. HHV8 positive nuclear stain of excised left inguinal lymph node. Positive HHV8 stain is necessary for the diagnosis of Kaposi Sarcoma

in KS cases involving the lymph nodes. Direct cutaneous invasion into nodes does not appear to worsen prognosis, but KS presenting solely with generalized lymphadenopathy is recognized as a disease more common in children and young adults that is associated with a worse prognosis.⁴

Regardless of the stage of KS, first line treatment is HAART. This has been shown in several trials, including one in which 80% of patients naive to HAART with cutaneous disease showed regression with HAART alone.⁵ The utility of HAART extends to prevention as well, as demonstrated by the sharp decrease in AIDS and KS incidence with anti-retroviral therapy introduction in the 1990s. In fact, the incidence of KS decreased from 30/1000 patient-years prior to 1995 to 0.03/1000 patient-years in 2001.²

Other modalities of treatment include radiation therapy and cytotoxic drugs. Radiation therapy for local disease can be an excellent option, with some series reporting 68-90% resolution of lesions, although the response in the epidemic form is less durable than in other forms of KS.^{6,7} Patients with more disseminated disease can be treated with systemic chemotherapy. Liposomal doxorubicin has been reported to have significantly higher response rates and possibly fewer side effects than other regimens including doxorubicin, bleomycin and vincristine.^{8,9} Based on these studies, liposomal doxorubicin is now considered first-line treatment for advanced KS. Paclitaxel has also shown promising results and is considered second-line therapy.⁹

Conclusion

As KS is a heterogeneous disease, it must always be considered in the workup of patients with HIV/AIDS. As demonstrated by our case, patients can present with normal CD4 counts, lack

skin lesions, and present with symptoms secondary to visceral involvement, including adenopathy, hemoptysis, gastrointestinal bleeding, and systemic symptoms. While KS is rarely a cause of death, it is not curable and can be disabling. Current management options are often able to symptomatically alleviate and improve quality of life.

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