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Case Report

A Case of Kaposi Sarcoma and Castleman Disease in a Young Male with New-Onset HIV

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Abstract

Kaposi sarcoma is an AIDS-defining illness associated with HHV-8 infection that is caused by the malignant proliferation of endothelial cells in blood and lymphatic vessels. Castleman disease is a rare, lymphoproliferative disease identified by specific histopathologic features of the lymph nodes following biopsy. An association between Castleman disease and Kaposi sarcoma is poorly described, and there are a limited number of cases in the literature-presenting patients with both conditions simultaneously. Thus, we present a case of a young male with newly diagnosed HIV that tested positive for HHV-8 and presented with both Kaposi sarcoma and multicentric Castleman disease.

A 22-year-old African American male presented to the ED with a one-week history of sore throat, fever, fatigue, diffuse cervical lymph node enlargement, and night sweats. Initial workup indicated that the patient was positive for Epstein-Barr virus and HIV, with an initial CD4 count of 141. CT of the neck revealed extensive cervical adenopathy and abdominal CT revealed splenomegaly. These findings raised suspicion for lymphoma, and bone marrow and lymph node biopsies were ordered. Surgical pathology of the lymph node confirmed the diagnosis of Kaposi sarcoma and multicentric Castleman disease.

This case highlights a rare manifestation of HIV and HHV-8 that is even more rare given the young age of the patient. The patient also lacked the classical skin changes associated with Kaposi Sarcoma. This case emphasizes the importance of a thorough workup upon the initial diagnosis of HIV to ensure proper identification and treatment of more uncommon associated malignancies/diseases.

Keywords: HIV; Multicentric Castleman disease; Kaposi Sarcoma; HHV-8

Abbreviations

HHV-8: Human Herpesvirus-8

Introduction

Kaposi sarcoma is an AIDS-defining illness associated with human herpesvirus-8 infection that is caused by the malignant proliferation of endothelial cells in blood and lymphatic vessels [1,2]. Kaposi sarcoma consists of four different subtypes, with the HIV-associated subtype, also known as the epidemic form, being the most common form in the world [1,3]. The annual incidence of Kaposi sarcoma in America is six cases per one million Americans, and it very rarely presents in younger adults [4].

Castleman disease is a rare, lymphoproliferative disease that presents with fever and weight loss and is confirmed by identifying specific histopathologic features of the lymph nodes following biopsy [5]. Castleman disease can be sub classified as unicentric or multicentric depending upon the number of lymph nodes involved [5]. Multicentric Castleman disease, which affects multiple lymph nodes throughout the body, is frequently caused by Human Herpesvirus-8 (HHV-8) infection [6]. The epidemiology of multicentric Castleman disease is largely unknown, but a review of records of patients with multicentric Castleman disease at two large United States treatment facilities found that the mean age of these patients was 53, and the estimated US 10-year prevalence was 2.4 cases per million [7].

While multicentric Castleman disease and Kaposi sarcoma are both strongly associated with HIV and HHV-8 infections, the association between Castleman disease and Kaposi sarcoma is poorly described, and there are a limited number of cases in the literature presenting patients with both conditions simultaneously [8,9]. Thus, we present a case of a 22-year-old male with newly diagnosed HIV that tested positive for HHV-8 and presented with both Kaposi sarcoma and multicentric Castleman disease.

Case Presentation

This case report describes a 22-year-old African American MSM with no reported past medical history who presented to the ED with a one-week history of sore throat, fever, fatigue, diffuse cervical lymph node enlargement, and night sweats. The patient denied shortness of breath, chest pain, diarrhea, skin changes, and neurological symptoms. He did not take any medications and had no reported family history. Social history was significant for mild alcohol use and sexual relationships with multiple male partners. Physical exam showed significant lymphadenopathy, but there was no evidence of any skin lesions, blotches, or tumors.

On admission, he had a platelet count of 10K and a hemoglobin of 4.9. Further workup indicated that the patient was positive for

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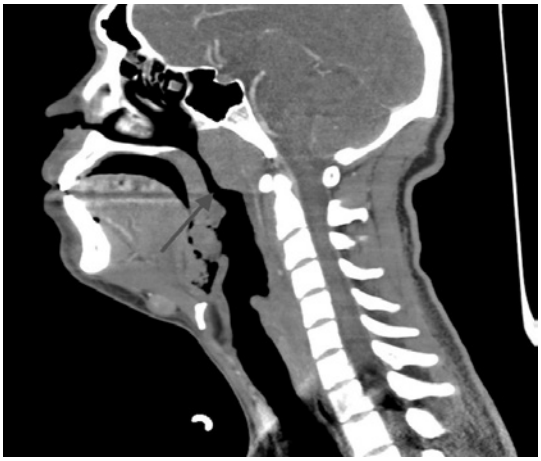

Figure 1: CT scan of the neck showed tonsillar enlargement and extensive adenopathy.

Epstein-Barr virus and HIV, with an initial CD4 count of 141. The patient was started on the antiretroviral Biktarvy in addition to atovaquone for PJP prophylaxis. CT of the neck soft tissue with IV contrast revealed enlargement of the palatine and lingual tonsils, in addition to extensive adenopathy in the anterior and posterior cervical regions and supraclavicular regions (Figure 1). CT of the chest, abdomen, and pelvis revealed splenomegaly at 16.7cm (Figure 2). These findings raised suspicion for lymphoma, and bone marrow and lymph node biopsies were ordered. The bone marrow biopsy and flow cytometry were not significant for any leukemias or lymphomas. Flow cytometry of the left axillary lymph node was negative for any monoclonal B or T-cell proliferations. Surgical pathology of the lymph node confirmed the diagnosis of Kaposi sarcoma.

The patient had an outpatient follow-up appointment two weeks later with Oncology. At this visit, he had a hemoglobin of 7.4 and platelet count of 7,000. He was re-admitted to the hospital to receive two units of platelets and one unit of blood. During this admission, he had an abnormal lymph node biopsy and was diagnosed with multicentric Castleman disease. He was started on doxorubicin and rituximab for outpatient management. He continues to follow-up with oncology and infectious disease to monitor for symptom recurrence and abnormal labs.

Discussion

This case describes a rare combination of pathologies in an atypical demographic given the patient's age. HIV-associated Kaposi sarcoma has a peak incidence in individuals between 30 and 39 years old [10]. Also, as mentioned above, preliminary review of records indicate that the mean age of patients with multicentric Castleman disease is 53 years old [7]. Thus, this patient's presentation of both diseases at 22 years old is unique and much younger than the typical demographic.

The patient also lacked many of the classical skin changes associated with Kaposi Sarcoma, which made the diagnosis of Kaposi sarcoma less apparent in the differential. However, comprehensive testing following a positive HIV test included HHV-8 testing and revealed the diagnosis. While the incidence of Kaposi sarcoma in the United States has significantly decreased with the advent of HAART and earlier diagnosis of HIV, it is still present and should be considered in the differential [4].

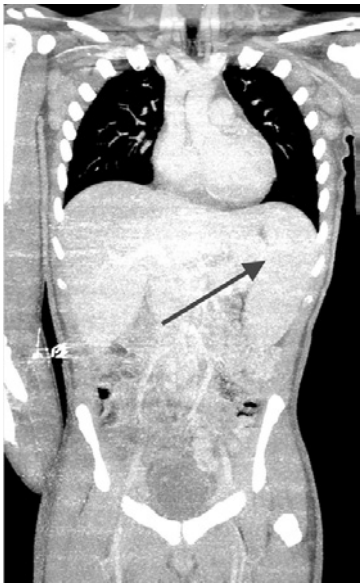

Figure 2: CT scan of the abdomen revealed splenomegaly.

The patient was diagnosed with multicentric Castleman disease following identification of an atypical pattern on lymph node biopsy, which was indicated due to his widespread lymphadenopathy with HIV. Multicentric Castleman disease is more common in individuals with HIV and almost always presents with concurrent HHV-8 infection [11]. Given that this patient tested positive for HIV and HHV-8 and presented with widespread lymphadenopathy, multicentric Castleman disease should have remained high on the differential. Although the condition is rare, patients with HIV and positive HHV-8 testing could potentially be considered for lymph node biopsy in order to evaluate for multicentric Castleman disease.

Conclusion

We present a case of a HIV patient with HHV-8 who had simultaneous multicentric Castleman disease and Kaposi sarcoma, a combination that has rarely been seen in the literature. In addition, the patient's young age at presentation and lack of typical skin lesions associated with Kaposi sarcoma make the case even more unique. Overall, this case demonstrates the importance of a thorough workup following diagnosis of HIV to ensure proper identification and treatment of more uncommon associated diseases, and physicians should remain cognizant of both multicentric Castleman disease and Kaposi sarcoma on the differential diagnosis when treating a new-onset HIV patient that is positive for HHV-8 and presents with symptoms suggesting lymphoma.

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