Kaposi Sarcoma Associated with Castleman Disease in an HIV Positive Patient: A Case Report

Maria J. Bertoli-Avella MD, R. Velez MD, Ramiro Perez, MD.

Department of Pathology and Laboratory Medicine
School of Medicine. University of Puerto Rico.

Introduction

Kaposi sarcoma (KS) is a locally aggressive endothelial tumor that typically presents with cutaneous lesions in the form of multiple patches, plaques or nodules but may also involve mucosal sites, lymph nodes and visceral organs. The disease is uniformly associated with human herpes virus 8 (HHV-8) infection. KS occurs in sporadic, iatrogenic, endemic and epidemic forms. Castleman disease (CD) is a form of lymph node hyperplasia; it was first described by Castleman et al in 1956 as a benign asymptomatic mass involving mediastinal lymph nodes. Now CD is defined as a heterogeneous and most likely represents multiple distinct diseases with different etiologies that share common histologic reaction patterns. Both entities are associated with HIV infection and rarely may coexist within the same organ.

Case Report

We present a case of a 41 year old man with history of HIV infection who presented with generalized lymphadenopathy and pancytopenia; a CBC revealed anemia with a hematocrit of 25.6% (reference interval: 41.1-50.4%), a hemoglobin of 9.1 g/dL (reference interval: 14.0-17.5 g/dL), a RBC of 3.07 x10^6 µL (reference interval: 4.5-5.9 x10^6 µL), a WBC of 1.7 x10^3µL (reference interval:4.4-11.0 x10^3µL) and a platelet count of 140,000 x µL (reference interval: 150,000-450,000 x µL). An abdominal CT scan revealed hepatosplenomegaly and retroperitoneal lymphadenopathy. A left cervical lymph node was send for biopsy. Microscopic examination was consistent with Castleman disease, plasma cell variant, and a partial involvement a spindle cell vascular proliferation with features of Kaposi sarcoma. Immunostains for HHV-8 were positive in both lesions.

Discussion

There are two variants of CD: localized, hyaline-vascular (HV) and plasma cell (PC) variant (Image 1), unicentric or multicentric. The causes of CD are mostly unknown. As stated above, one known etiology is HHV-8, (Image 2), also known as Kaposi sarcoma herpes virus. HHV-8 can be present in approximately 50% of cases of PC variant and in most cases arising in patients with human immunodeficiency virus (HIV) infection. Patients with CD are prone to developing malignant tumors. In the HV variant, a follicular dendritic cell sarcoma has been reported and in the PC variant, particularly those with HIV infection, Kaposi sarcoma (Image 3) and lymphomas may coexist. Even though these two entities are not infrequently seen in HIV+ patients, the simultaneous presence of both lesions in a lymph node is not frequent. This uncommon finding is another piece of evidence of the close link among HHV-8 (Image 4), Kaposi sarcoma, and multicentric Castleman disease.

References

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