



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

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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

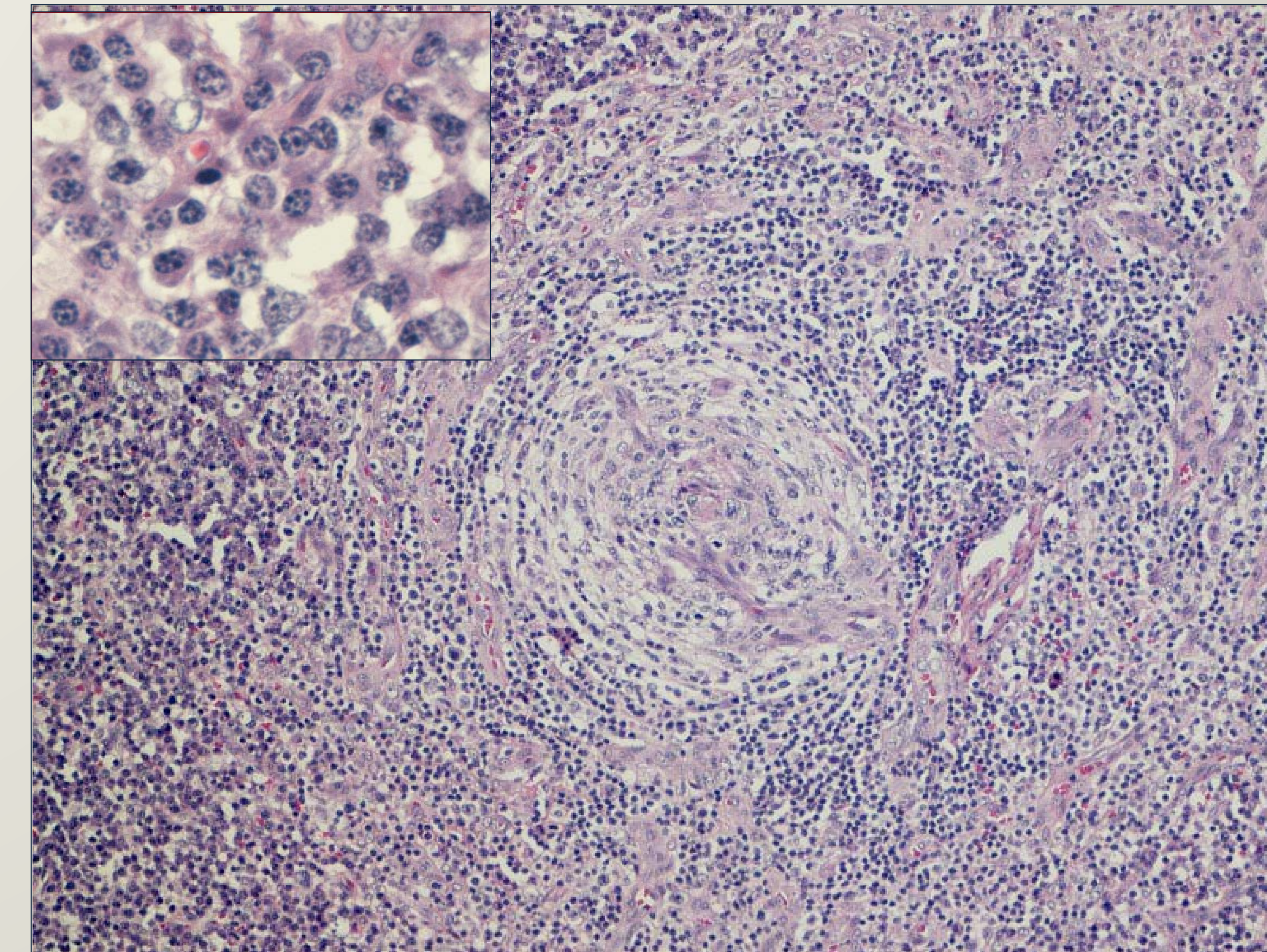


Image 1. Castleman disease, plasma cell variant : H&E showing a germinal center with blood vessels and sheets of mature plasma cells within the interfollicular region (insert)

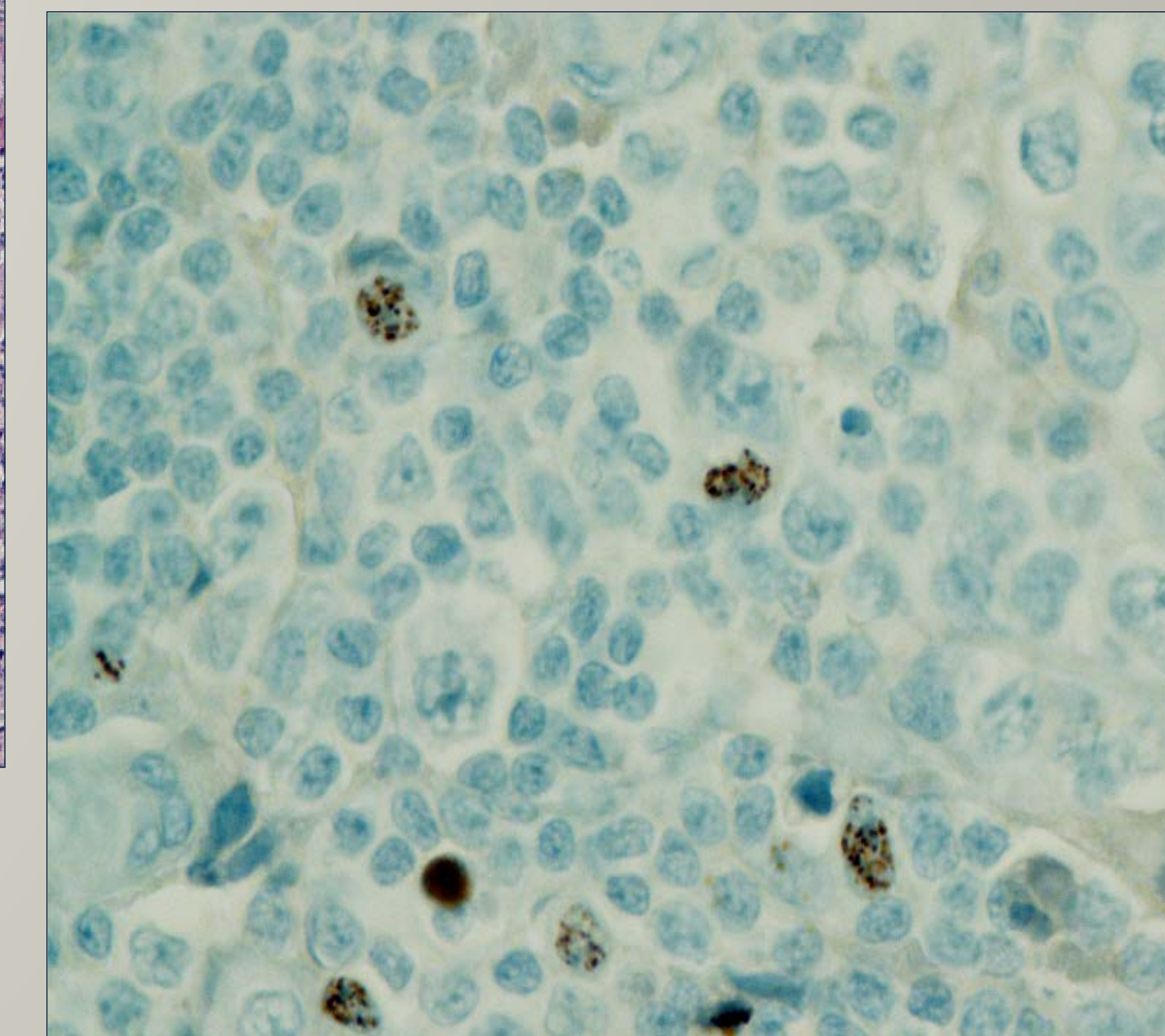


Image 2. Castleman disease, plasma cell variant: HHV-8 immunostain nuclear positivity in lymphocytes

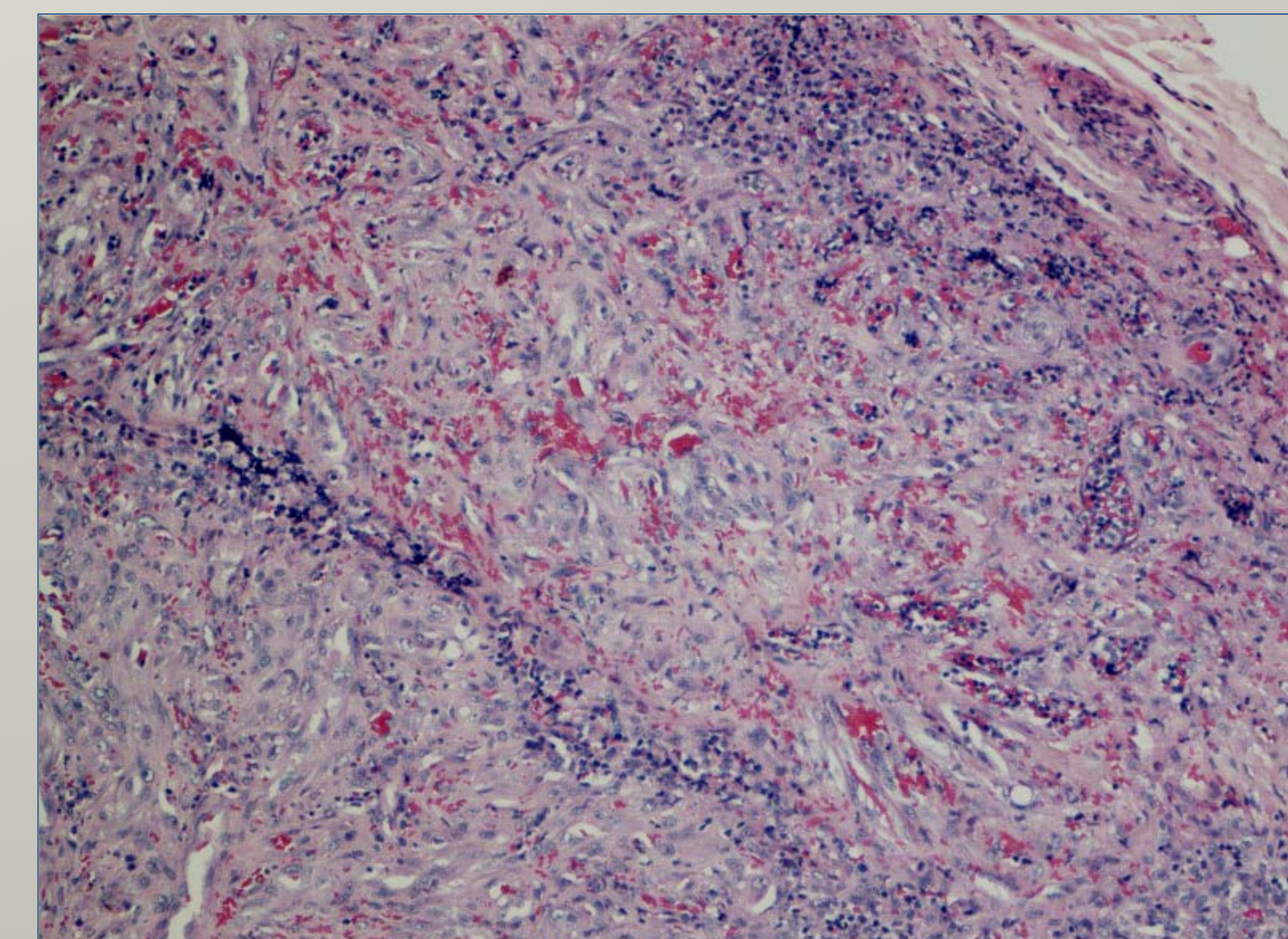


Image 3. Kaposi sarcoma: H&E. Subcapsular proliferation of slit like vascular spaces filled with erythrocytes

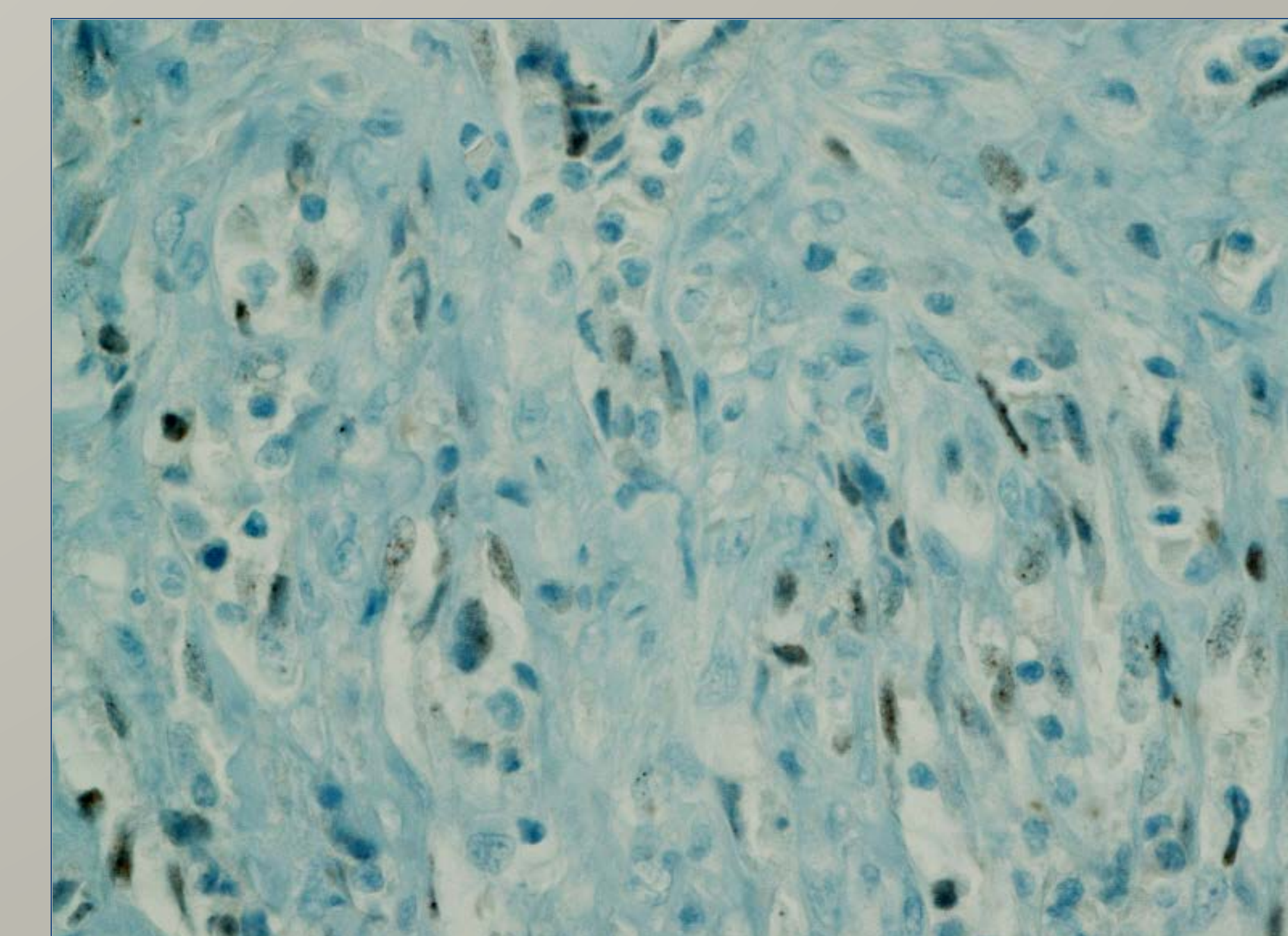


Image 4 . Kaposi sarcoma: HHV-8 Immunostain nuclear positivity in spindle cell area

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.

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 $3.07 \times 10^6 \mu\text{L}$ (reference interval: $4.5-5.9 \times 10^6 \mu\text{L}$), a WBC of $1.7 \times 10^3 \mu\text{L}$ (reference interval: $4.4-11.0 \times 10^3 \mu\text{L}$) and a platelet count of $140,000 \times \mu\text{L}$ (reference interval: $150,000-450,000 \times \mu\text{L}$). An abdominal CT scan revealed hepatosplenomegaly and retroperitoneal lymphadenopathy. A left cervical lymph node was sent 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.

References

1. Ioachim Harry L., Medeiros Jeffrey L., Ioachim's Lymph node pathology. Fourth ed. Lippincott Williams & Wilkins, 2009
2. J. Lamovec J, Knuutila S. World Health Organization, Pathology and Genetics of Tumours of Soft Tissue and Bone IARC Press Lyon, 2002
3. Grandadam M, Dupin N, Calvez V, et al. Exacerbation of clinical symptoms in human immunodeficiency virus type 1- infected patient with multicentric Castleman's disease are associated with a high increased in Kaposi's sarcoma in virus DNA load in peripheral blood mononuclear cells. J. Infect Dis 1997; 175: 1198-1201
4. Amin HM, Medeiros LJ, Manning JT, et al. Disruption of the lymphoid follicle is a feature of the HHV-8 + variant of Plasma cells Castleman disease. Am J Sur Path 2003; 27:91-100
5. Brousset P, Cesarman E, Meggetto F, et al. Colocalization of the viral interleukin-6 with latent nuclear antigen 1 of human herpes virus-8 in endothelial cells and spindle cells of Kaposi sarcoma and lymphoid cells of multicentric Castleman disease. Hum Pathol 2001; 32:95-100
6. Bisceglia M, Amini M, Bosman C. Primary Kaposi sarcoma of the lymph node in children. Cancer 1988; 61:1715-1718
7. Ramos CV, Taylor HB, Hernandez BA, et al. Primary Kaposi sarcoma of lymph node. Am J Clin Pathol, 1976; 66: 998-1003
8. Amazon K, Rywlin AM. Subtle clues to diagnosis by conventional microscopy: lymph node involvement in Kaposi sarcoma. Am J Dermatopathol 1979; 1:173