

# Kaposi's Sarcoma With a Non-Hodgkin's Lymphoma

## Its Association in a Male Homosexual With Human T-Cell Lymphotropic Virus Type III Infection

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• **Combined tumor syndromes, specifically reticuloendothelial malignancies and Kaposi's sarcoma, have long been recognized. With the recognition of the acquired immunodeficiency syndrome (AIDS), several patients with concurrent non-Hodgkin's lymphoma and Kaposi's sarcoma have been reported at high risk for developing AIDS. The present Centers for Disease Control definition of AIDS excludes these patients on the assumption that one tumor is affecting the cellular immunity, allowing for the development of the second malignancy. In evaluating such a patient who had serologic evidence of human T-cell lymphotropic virus type III infection, the probable cause of AIDS, we have reviewed reports of patients with similar concurrent malignancies before and since the onset of the AIDS epidemic. We conclude that patients in high-risk groups for AIDS who develop similar combined tumor syndromes should be classified as having AIDS.**

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With the recognition of the acquired immunodeficiency syndrome, a number of authors have documented the association of reticuloendothelial neoplasms associated with Kaposi's sarcoma in homosexual men. We present a case of non-Hodgkin's lymphoma and Kaposi's sarcoma in a homosexual man with serologic evidence of human T-cell lymphotropic virus type III (HTLV-III) infection and argue for the inclusion of these patients in the present Centers for Disease Control surveillance definition.

### REPORT OF A CASE

The patient was a 26-year-old male homosexual who had noted the onset of generalized lymphadenopathy two years prior to the time of hospital admission. An occipital node biopsy specimen revealed nonspecific changes. His lymphadenopathy persisted. In June 1983, he noted weight loss, polydipsia, polyuria, night sweats, and fever. By mid-July the patient had developed hoarseness, dysphagia associated with a cough productive of blood-tinged saliva, and weakness and dyspnea on exertion. He was admitted to the Ochsner Foundation Hospital, New Orleans, on July 20, 1983.

Physical examination revealed a cachectic man with a blood pressure of 104/50 mm Hg, a pulse rate 120 beats per minute, an oral temperature of 36.8 °C, and a respiratory rate of 18/min. He had a massive submucosal hemorrhage of his posterior pharynx. Tachycardia, with an S4 gallop, a grade 1/6 systolic ejection murmur, decreased breath sounds in both posterior lung fields, and hepatosplenomegaly were present. Firm, matted, nontender, lymph nodes measuring up to 2 × 3 cm were noted bilaterally in the occipital, cervical, supraclavicular, axillary, and inguinal areas.

His white blood cell count was 27,800/cu mm, with 52% polymorphonuclear cells, 22% band forms, 1% metamyelocytes, 2% myelocytes, 18% lymphocytes, 4% monocytes, and 1% basophils. The hemoglobin level was 4.6 g/dL, and the platelet count was 23,000/cu mm. Reticulocyte count was 9%, direct and indirect Coombs' tests were positive, and the haptoglobin value was decreased. The prothrombin time and the partial thromboplastin time were normal, but his bleeding time was greater than 15 minutes. T-cell studies revealed a decreased T-helper/inducer to suppressor/cytotoxic cell ratio of 0.71 resulting primarily from decreased helper/inducer T-lymphocytes. The rapid plasma reagin was reactive at a dilution of 1:16. Immunoelectrophoresis revealed an elevated IgG level in a polyclonal pattern. A chest roentgenogram revealed bilateral pleural effusions.

Bone marrow aspirate and a biopsy specimen revealed a hyper-

Patients With Concurrent Malignancies at Risk for Developing AIDS\*

Source	Patient Age, yr	Tumor Types	Longevity From Date of Diagnosis of Lymphoma
Ciobanu et al <sup>2</sup>	33	Kaposi's sarcoma; preleukemic stage or a lymphoblastic neoplasia	1 mo
Perlow et al <sup>3</sup>	31	Kaposi's sarcoma; unclassified malignant lymphoproliferative disorder	2 days
Chaganti et al <sup>4</sup>	39	Kaposi's sarcoma; diffuse malignant Burkitt's lymphoma	...
Ziegler et al <sup>5</sup>	38	Kaposi's sarcoma; undifferentiated lymphoma	4 mo
	39	Kaposi's sarcoma; undifferentiated lymphoma	Autopsy diagnosis
	37	Kaposi's sarcoma; immunoblastic sarcoma	3 mo
	40	Kaposi's sarcoma; immunoblastic sarcoma of the brain	Autopsy diagnosis
	31	Kaposi's sarcoma; diffuse mixed lymphoma	...
Figlin et al <sup>6</sup>	31	Kaposi's sarcoma; large-cell, immunoblastic lymphoma	Approximately 3 mo
Present study	26	Kaposi's sarcoma; large-cell, B-cell lymphoma	2½ wk

\*AIDS indicates acquired immunodeficiency syndrome. All patients were homosexual men.

cellularity with adequate to increased megakaryocytes. The stroma had a washed-out, fibrinous background with an absence of fat in some areas. All myeloid elements were well represented. Granulocytic hyperplasia with a predominance of immature forms was present. There was an increase in reticulin, with a slight amount of mild reticulin fibrosis. Special stains identified no acid-fast or fungal organisms and none grew from culture. An echovirus<sup>1</sup> was eventually isolated from culture of the bone marrow.

Cytologic study of the pleural fluid was positive for class 5 malignant lymphoma. Appropriate stains and cultures for mycobacteria and bacteria were negative on both thoracentesis and cerebrospinal fluids. *Candida albicans* subsequently grew from the pleural fluid and sputum. *Mycobacterium avium intracellulare* was also isolated from sputum.

Because of progressive thrombocytopenia and bleeding unresponsive to transfusion and prednisone therapy, a splenectomy was performed on July 26. At surgery, both the liver and intra-abdominal lymph nodes were noted to be enlarged.

Histologically the spleen had two distinct lesions present. There were multiple foci of nodular aggregates of large, noncleaved lymphoma cells, many with relatively abundant eosinophilic cytoplasm. Aggregates of Kaposi's sarcoma were also present.

After an initial rise in the platelet count following splenectomy and 2 mg of vincristine sulfate, it fell to preoperative levels. Cyclophosphamide, intravenous  $\gamma$ -immunoglobulin, vincristine-loaded platelets, and danazol (Danocrine) were subsequently tried but without response. The patient died on Aug 10, 1983.

An autopsy revealed extensive Kaposi's sarcoma and diffuse histiocytic lymphoma involving widespread lymph nodes, the gastrointestinal tract, and the mediastinal and retroperitoneal connective tissue. Immunoperoxidase staining of the spleen revealed the lymphoma to be of B-cell origin, expressing monoclonality for IgM and  $\gamma$  light chains. Serum sent for HTLV-III serologic study was 3+ by the latex agglutination method.

#### COMMENT

The presence of a coexistent tumor excludes patients from the present AIDS surveillance definition on the assumption that one tumor has affected the patient's immunity in such a manner to allow for the development of a second malignancy.<sup>1</sup> Recently, however, a number of authors have reported simultaneously occurring reticuloendothelial neoplasms and Kaposi's sarcoma in patients at risk for developing AIDS (Table). With the isolation of HTLV-III<sup>7</sup> as the probable cause for AIDS, serologic markers became available. The demonstration of serologic evidence of this infection in our patient, we believe, confirms the diagnosis of this syndrome. We conclude that patients with simultaneously occurring reticuloendothelial neoplasms and Kaposi's sarcoma who are at risk for developing AIDS should be included in the present definition of AIDS for surveillance purposes.

Since this report was submitted, the Centers for Disease Control has adopted in the case definition of AIDS, non-Hodgkin's lymphoma of high-grade pathologic type (diffuse, undifferentiated) and of B-cell or unknown immunologic phenotype, diagnosed by biopsy, if the patient has a positive serologic or virologic test for HTLV-III.<sup>8</sup>

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## Life-Threatening Cat-Scratch Disease in an Immunocompromised Host

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• We describe a renal allograft recipient with cat-scratch disease in whom refractory hypotension, severe metabolic acidosis, pulmonary infiltrates, and encephalopathy developed. The patient first presented with a history of cat bites and scratches, fever, headache, and arthralgias. Four weeks later, the clinical presentation of septic shock suddenly developed in the patient. Cat-scratch disease was documented clinically and by finding delicate pleomorphic bacilli in Warthin-Starry silver stains of biopsy specimens taken from the primary inoculation site and regional lymph node. The administration of intravenous sulfamethoxazole and trimethoprim, erythromycin lactobionate, and tobramycin sulfate therapy correlated with recovery. Although cat-scratch disease is usually a benign, self-limited illness, this article illustrates its systemic nature, its potential for devastating complications in the immunocompromised host, and its possible response to vigorous antibiotic therapy.

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Cat-scratch disease (CSD) is the most common cause of chronic regional lymphadenitis in children.<sup>1</sup> It usually produces a mild, self-limited illness. Typically, a cat scratch or bite is followed in seven to 12 days (normal range, three to 50 days) by a papule or pustule at the site of the injury. Regional lymphadenopathy, most commonly in the head, neck, and/or axilla, appears within six or seven weeks. These enlarged lymph nodes are tender in 80% of patients. They may suppurate, but generally regress within two to four months. Constitutional symptoms such as fever and malaise are found in about one third of patients.<sup>1</sup> Unusual manifestations of CSD include encephalitis,<sup>2</sup> osteolytic lesion,<sup>3</sup> mesenteric adenitis,<sup>4</sup> pneumonitis,<sup>5</sup> nonthrombocyto-

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The views expressed herein are those of the authors and are not to be construed as official, or as reflecting the views of the US Air Force, US Army, or the Department of Defense.

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