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AGGRESSIVE KAPOSI'S SARCOMA IN CHILDREN: A CASE REPORT*

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In 1872 Morictz Kaposi described an independent entity of "idiopathic multiple pigment sarcoma of the skin" in patients seen in the dermatology department[1]. The tumor was characterized by the development of skin nodules or plaques usually starting on the hands or feet but occasionally involving the viscera[2].

It has been recently reported in the United States that immunosuppressed patients[3-4], homosexual men[5], intravenous drug abusers. Haitian immigrants, hemophiliacs[6], and prision inmates[7] were especially at risk of developing Kaposi's sarcoma (KS). Recent reports of acquired immunodeficiency syndrome (AIDS) have called attention to KS in children[8]. In this article, we present a three year-old boy with aggressive KS.

Case Report

A three-year-old boy was admitted to the Dr. Sami Ulus Children's Hospital in October 1986, presenting with periorbital ecchymoses, palpebral edema, dyspnea, and fever of eight days' duration. There were no significant abnormalities pertaining to his prenatal, natal or early postnatal periods. His physical and motor development was normal. It was ascertained that BCG, DPT, and polio immunizations had been given to the patient at the appropriate intervals, and that he had not contracted mumps, measles, varicella or pertussis. The child, however, received various drugs for frequent upper respiratory tract infections and recurrent episodes of otitis media. His mother, aged 20, is a housewife in good health, and his father, aged 24, is a professional driver suffering from peptic ulcer. The parents had never been abroad, had never been recipients of blood

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transfusions, and were neither drug users nor homosexuals. The patient had a healthy seven-month-old sister. There was no history of abortion, stillbirth or death of a sibling in the family.

Physical examination revealed a child with a temperature of 39°C, dyspnea, and a respiration rate of 45 per minute. He had facial edema, periorbital ecchymoses and subconjunctival hemorrhages. There was a bilateral, fixed, hard mass measuring 7x7 cm in the mandibular area. Bilateral axillar cervical and inguinal nodes, the largest of which was 2x2 cm, were also palpated.

Ascultation of the chest revealed fine bilateral rales. The liver was palpable five cm and the spleen six cm below the arcus costarum in the midclavicular line.

Laboratory findings revealed a hemoglobin of 5.5 g/dl, white blood cell count 17200/mm^3, platelet count 75000/mm^3; the peripheral blood smear showed polychromasia, hypochromia, enisopoikilocytosis, 34 % segmented neutrophils, 28 % lymphocytes, 10 % monocytes, 1 % basophils, 20 % atypical lymphocytes, 7 % atypical monocytes. Examination of the bone marrow aspirate showed hypercellularity, slight megaloblastic changes in erythroid cells and an increase in promyelocytes. The BUN was 30 mg/dl, serum creatinine 0.7 mg/dl, total serum protein 5.5. g/dl, albumin 3.25 g/dl, total bilirubin 0.6 mg/dl, total lipids 830 mg/dl; total cholesterol 170 mg/dl, alkaline phosphatase 4.6 BU; SGOT 15 U; SGPT 30 U. The erythrocyte sedimentation rate was 50 mm/hr, PPD test negative, direct Coombs test negative, immunoelectrophoresis: IgG 1310 mg/dl (N: 929±228), IgM 138 mg/dl (N :56±18), IgA 62.9 mg/dl (N :93±27). Hemoglobin electrophoresis and the throat culture were within normal limits. The chest roentgenogram revealed a mediastinal mass and bilateral parenchymal infiltration. IVP and roentgenograms of the bones were normal.

The patient was transfused 350 ml of blood and combination therapy of ampicillin and gentamicin was initiated intravenously. A biopsy of the right inguinal node was obtained. The lymph node was pink and elastic and measured 1.2x0.5x0.4 cm. Microscopically, the structure of the lymph node was largely distorted and a malignant tumor was noticed. The tumor consisted of atypical fusiform endothelial cells which also formed vascular formations. Malignant tumoral infiltration of varying size and location was observed in the marginal sinuses which was defined as the metastasis of KS. In the patient's sera HTLV-III antibody was found to be negative by the ELISA method. The child died of progressive pulmonary insufficiency on the tenth day of hospitalization.

The following findings were obtained at postmortem examination: Microscopically, examination of the heart showed no significant findings except for left ventricular hypertrophy. Mucous membranes starting in the subepiglottic region and extending to where the main bronchi enter the lungs were hyperemic and

rough. Many irregular lesions of varying size, the largest, 1 cm in diameter, were observed on the visceral pleural sides of both lungs. There were many red lesions on the stomach and on the ascending colon including the cecum, the largest being 1.5 cm and 0.5 cm in diameter, respectively. Tumoral infiltrations ranging from 0.1 to 0.5 cm in diameter were detected in the lymph nodes of the cervical, mediastinal, paraaortic, celiac, mesenteric and inguinal regions.

Microscopically, examination of the epiglottis, trachea, bronchi, lungs, palatine tonsils, submandibular glands, stomach, liver, small intestine and bowel, thymus, spleen, and cervical, mediastinal, abdominal, and inguinal lymph nodes showed malignant tumoral infiltrations. Vascular structures composed of atypical endothelial cells were observed between the mucosa and submucosa. These cells varied in size, shape and staining characteristics. They were either round or notched, varied in diameter and some had red blood cells within their lumens. The presence of vascular structures was demonstrated by using silver and the Massoni trichome staining techniques (Figs. 1-3). On examination no significant pathological findings were detected in the radix linguae, testes, diaphragm, thyroid gland, skeletal muscles, skin, pancreas, medulla spinalis, bladder or surrenal glands. Histological evidence of *pneumocystis carinii* and cytomegalovirus infections was not found.

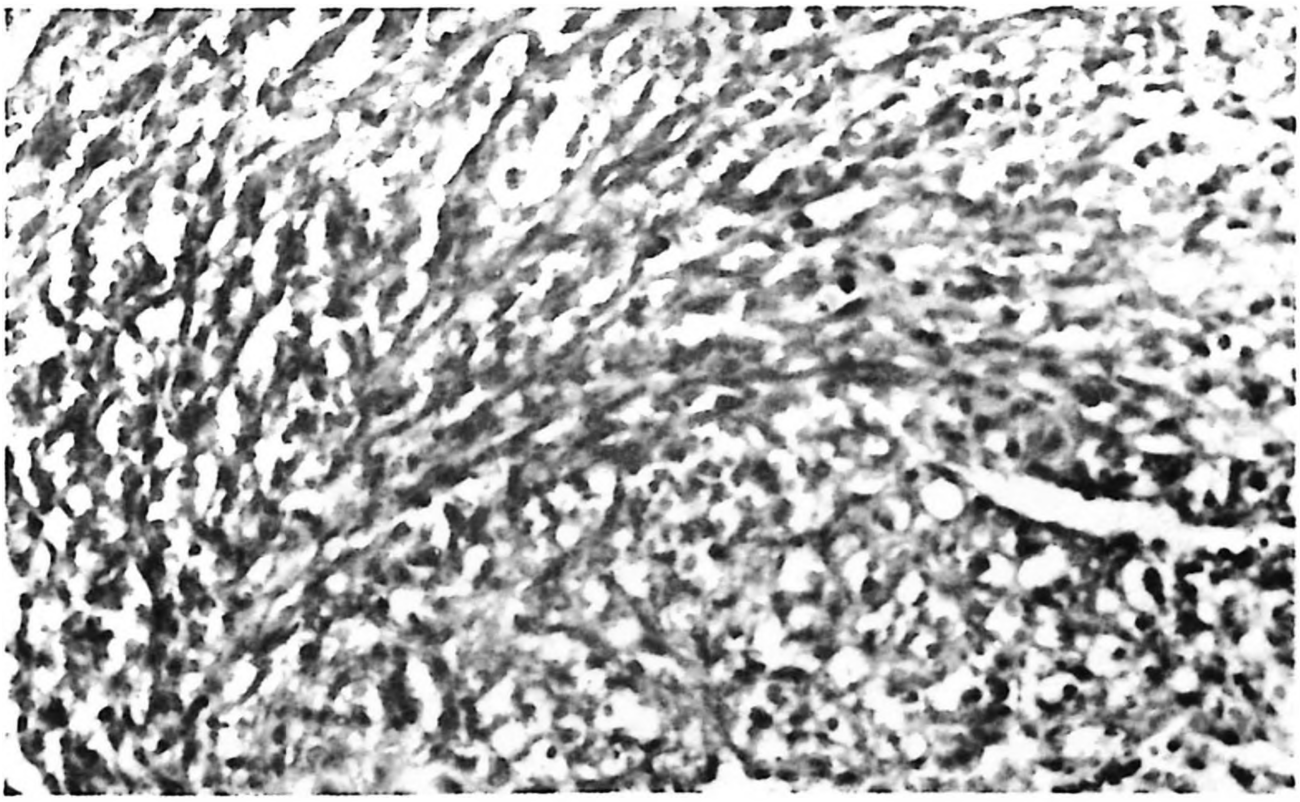

Fig. 1 a: Kaposi's sarcoma metastasis in the lymph nodes; atypical endothelial cells varying in size and shape. Hematoxylin and eosin (x 200).

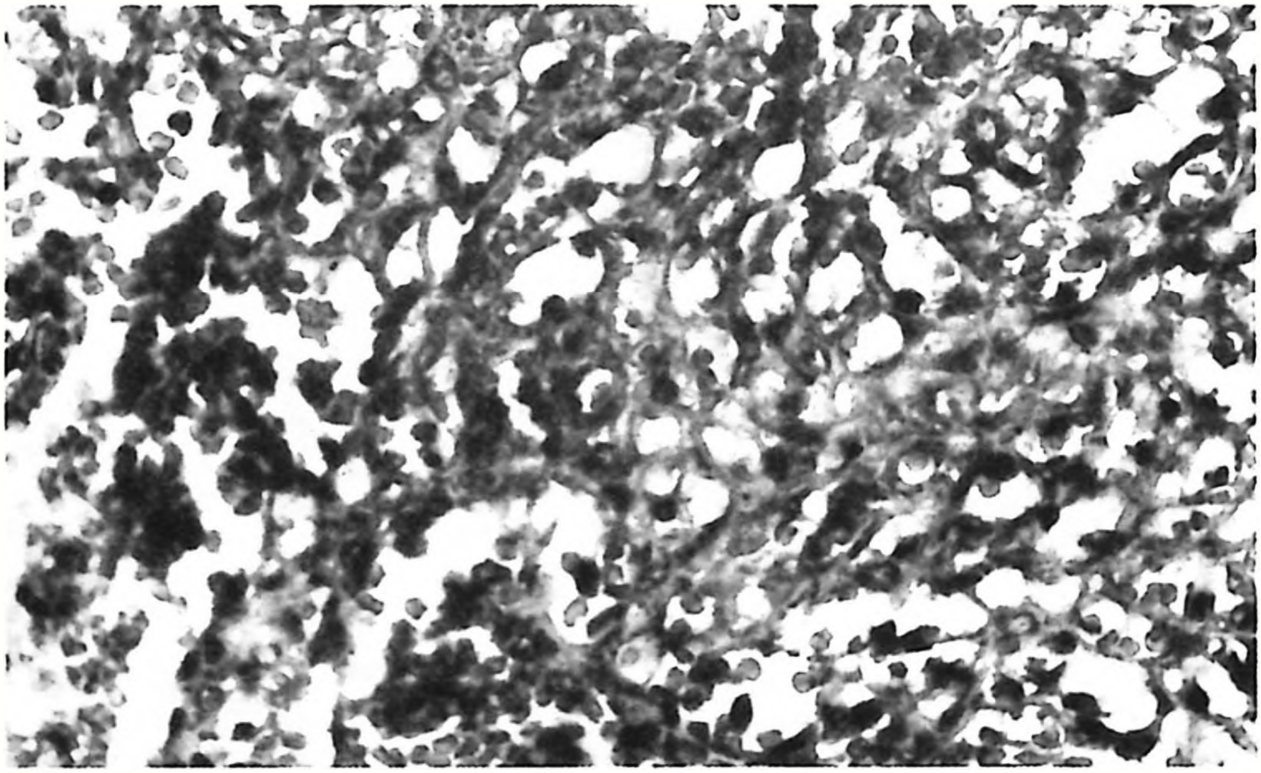

Fig. 1 b: Kaposi's sarcoma metastasis in the lymph nodes; atypical endothelial cells varying in size and shape. Hematoxylin and eosin (x 400).

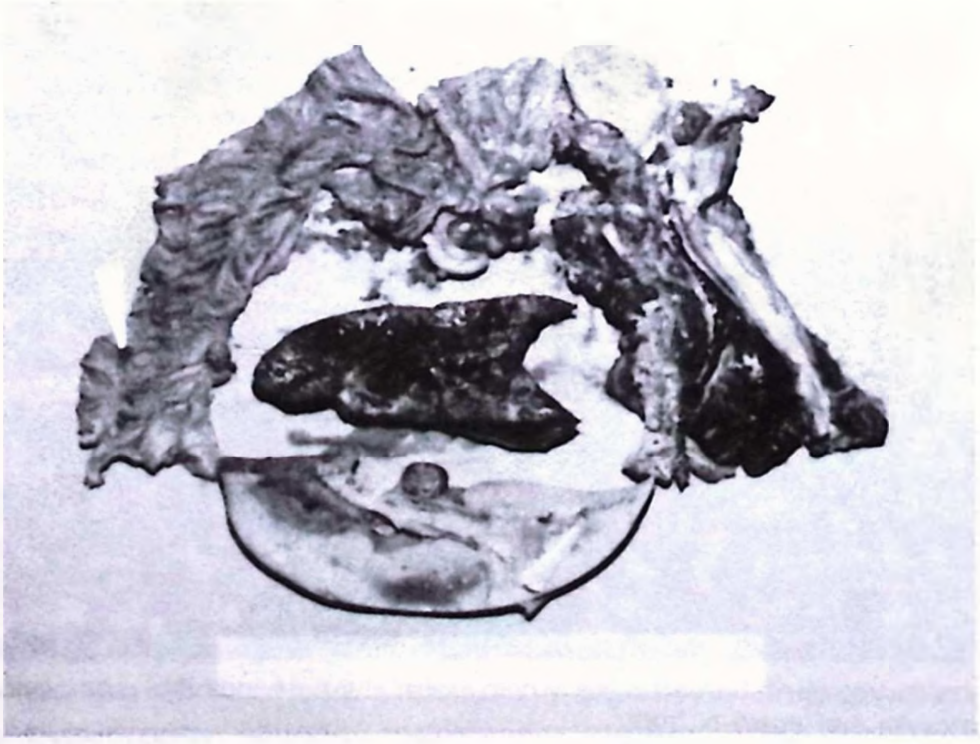

Fig. 2: Thymus, palatine tonsils, mediastinal and paratracheal lymph nodes, lungs, liver, and colon of the patient.

Fig. 3: Tumoral infiltration in the lung (macroscopical).

Discussion

Kaposi's sarcoma, a very rarely encountered tumor in children[8,9] has been reported in central east Africa, Europe and America.[10-12] Especially in Black African children the tumor is usually aggressive and pursues a fulminant course. Black African children with KS often have prominent lymph node enlargement. Visceral involvement develops in up to 20 percent of cases during the course of the disease. The lymph nodes and gastrointestinal tract are the most common sites of deep-seated disease but pulmonary involvement is also seen in disseminated cases.[13]

Recently KS has been regarded as an epiphenomenon of AIDS[14]. Skin or lymph node involvement may occur in KS associated with AIDS but patients suffering from progressive disease usually have multiple organ involvement particularly of the gastrointestinal tract and pulmonary parenchyma. Most of these patients have positive serologic evidence for HTLV-III antibody. However, two cases of AIDS with negative HTLV-III antibodies but positive HTLV-III antigens have been reported, one of these patients had KS[15]. Asymptomatic individuals at risk of developing AIDS who carried HTLV-III in their blood and/or saliva without detectable antibodies have been[16] previously reported. Although in endemic areas KS is thought to be independent of HTLV-III infection, in the United States a diagnosis of KS is usually thought to be synonymous with the diagnosis of AIDS but some cases of primary visceral KS unrelated to AIDS have also been reported[17].

Our patient had generalized Kaposi's sarcoma. When questioned, the family denied having had any contacts either at home or abroad with individuals at risk of developing AIDS. They also disclosed that they had not been recipients of blood transfusions and had not taken intravenous injections of drugs. Postmortem examination did not reveal an opportunistic infectious agent which was also a finding that did not correlate with AIDS. Therefore, we diagnosed our patient as having aggressive primary KS.

Summary

A three-year-old-boy with generalized Kaposi's sarcoma (KS) is presented. The child died of progressive pulmonary insufficiency on the eighteenth day of the course of his illness, the tenth hospital day. On postmortem examination diffuse KS infiltration was observed in the respiratory and gastrointestinal tracts, lymph nodes, liver, spleen and thymus. The patient was considered to be a case of KS unrelated to AIDS because of his negative HTLV-III antibody and epidemiologic characteristics, and therefore was believed to have primary aggressive KS.

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