Tropical Medicine and International Health

VOLUME 5 NO 2 PP 85–87 FEBRUARY 2000

Aggressive Kaposi's sarcoma in a 6-month-old African infant: case report and review of the literature

K. P. Manji[1], H. Amir[2] and I. Z. Maduhu[1]

1 *Department of Paediatrics and Child Health, Muhimbili University College of Health Sciences, Dar-es-Salaam, Tanzania*
2 *Department of Surgery, Muhimbili University College of Health Sciences, Dar-es-Salaam, Tanzania*

Summary Kaposi's sarcoma (KS), known to exist in Africa for a century now, was rare in children and unknown in the newborn. With the onset of the HIV/AIDS epidemic, a more aggressive, disseminated type of KS (AKS) was recognized. Recently KS was diagnosed in a 6-month-old infant in Tanzania. Data support the notion that HHSV8 infectivity can be potentiated with HIV infection and thus produce multiple lesions in different anatomical sites early in life. Furthermore, the available evidence would suggest a nonsexual route of HHSV8 infection, possibly from mother to fetus.

keywords Kaposi's sarcoma, infant, African, transmission route

correspondence Dr K. P. Manji, Department of Paediatrics and Child Health, MUCHS, P.O.Box 65001, Dar-es-Salaam, Tanzania. E-mail: kmanji@muchs.ac.tz

Introduction

Kaposi's sarcoma (KS) has been known in Africa since the beginning of the century. It presented as an indolent disease of the skin and lower limbs and was predominantly observed in elderly men and infrequently among females (Oettle 1962; Amir *et al*. 1997). KS in children was rare, with a narrow gender ratio, and presented with distinctive characteristics affecting mainly the lymph nodes, rarely the skin (Olweny *et al*. 1976; Connor *et al*. 1990).

With the advent of the HIV/AIDS pandemic, KS has been reported in children worldwide (Malekzadeh *et al*. 1987; Baum & Vinters 1989; Arico *et al*. 1991; Porta *et al*. 1991), presenting predominantly as a muco-cutaneous disease (Ziegler & Katongole-Mbidde 1996). KS has been observed in different age groups among children but has rarely been documented in infants aged up to 6 months in sub-Saharan African countries (Patil *et al*. 1992; Ziegler & Katongole-Mbidde 1996).

We report an HIV-associated KS affecting multiple anatomical sites in an infant with onset of the disease at 6 months of life. The findings are discussed in reference to the role of HIV as a cofactor to HHSV8 in KS pathogenesis and its mode of transmission in infants.

Case report

UN was admitted at the age of 30 months with the chief complaints of multiple swellings on the skin and in the mouth from the age of 6 months or earlier, fever, cough, diarrhoea and failure to thrive for the past 18 months. The symptoms were recurrent and the maternal and child health (MCH) card indicated growth faltering. The infant had received all primary immunization.

The patient had fever, generalized nonpitting oedema and shortness of breath. There were generalized nodular lesions on the skin and multiple oral lesions in the buccal cavity and tongue, of varying sizes (0.5–1 cm). These dark purple nodules were firm in consistency, indurated and nonulcerated (Figure 1). With this there was generalized lymphadenopathy and hepatosplenomegaly. Examination of the chest revealed a respiratory rate of 56 per minute with decreased air entry on the right hemithorax. There were also scattered rales on all lung fields.

The patient was born with a birthweight of 3.1 kg and was doing well up to the age of 6 months or earlier, when the first cutaneous nodular lesions on the scrotum and the scalp began to appear. The child is the second sibling. Each child has a different father. The mother has had 3 first trimester

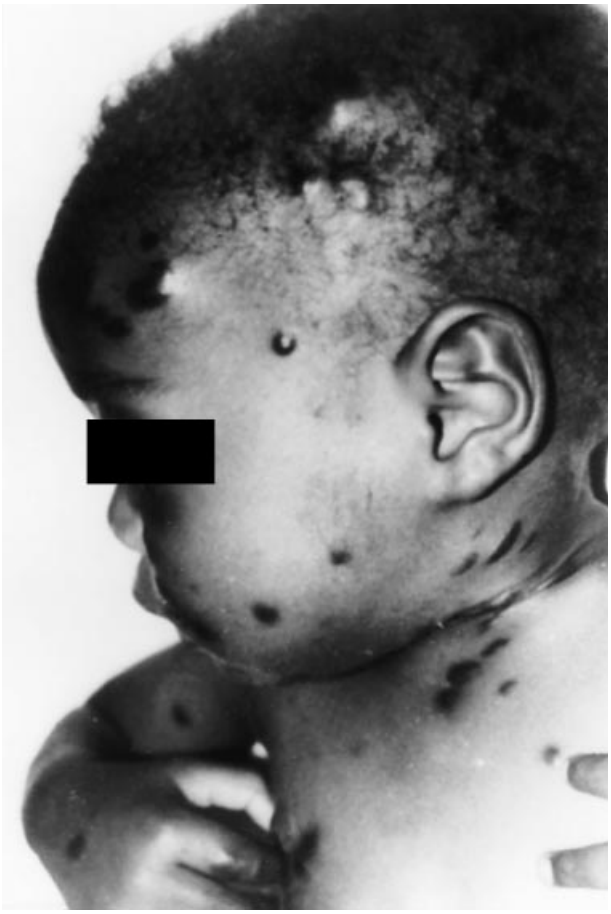

Figure 1 Disseminated cutaneous lesions of Kaposi's Sarcoma.

abortions before completing this index pregnancy. The father of this child has a history of multiple sexual partners. The mother was treated for tuberculosis before this pregnancy and severely wasted with generalized skin disease.

Biopsy of the skin nodules was taken and the child treated according to the standard protocol for pneumonia. Subsequently, the child succumbed after 4 weeks of admission.

Investigations

Biopsy of the skin lesions indicated nodular proliferation of spindle shaped cells with many vascular slits and extravasation of red blood cells accompanying mononuclear inflammatory cell infiltrates consistent with nodular Kaposi's sarcoma. The complete blood count revealed haemoglobin of 10.0 g/dl and a platelet count of 126×10^9/l (low). HIV antibody testing with ELISA was positive and confirmed by Western Blot technique. The CD4 count was 140, the CD8 count 2910 with a CD4: CD8 ratio of 0.05. A CT scan of the chest showed a widened mediastinum, pleural effusion on the right hemithorax, diffuse echogenic attenuation around the hilum and bilateral pleural thickening (Figure 2). A CT scan of the abdomen revealed thickened bowel loops, para-aortic and mesenteric nodal enlargement (Figure 3).

Discussion

Kaposi's sarcoma was infrequently seen among children before the AIDS pandemic. This disease predominantly affected males aged 1–5 years and involved the lymph nodes (Patil *et al*. 1992; Ziegler & Katongole-Mbidde 1996). With the advent of the HIV epidemic in sub-Saharan Africa in the early 1980s, the incidence of childhood KS has increased and the tumour distribution is mainly oro-facial (Ziegler & Katongole-Mbidde 1996).

Recently, Human Herpes Simplex Type 8 virus (HHSV8) was shown to be associated with all types of Tanzanian KS (Schalling *et al*. 1995). The transmission of HHSV8 is primarily associated with sexual transmission in adults and a nonsexual route in children (Lennette *et al*. 1996). Since the KS anatomical site in children is commonly oro-facial during the HIV epidemic, it would suggest a mucosal transmission of the KS agent (Ziegler & Kotongole-Mbidde 1996). Presence of HHSV8 in blood and body secretions (Whitby *et al*. 1995; Levy 1997) would suggest that HHSV8 could be transmitted from mother to fetus during birth or early in life through breastfeeding (Ziegler & Kotongole-Mbidde 1996).

The maternal-fetal transmission could be further substantiated by the development of KS in a 6-day-old neonate (Gutierrez-Ortega *et al*. 1989) and also in the 6-month-old infant documented here. Interestingly, both patients were born to mothers with HIV/AIDS. It has been suggested that HHSV8 is endemic in sub-Saharan Africa and that its infec-

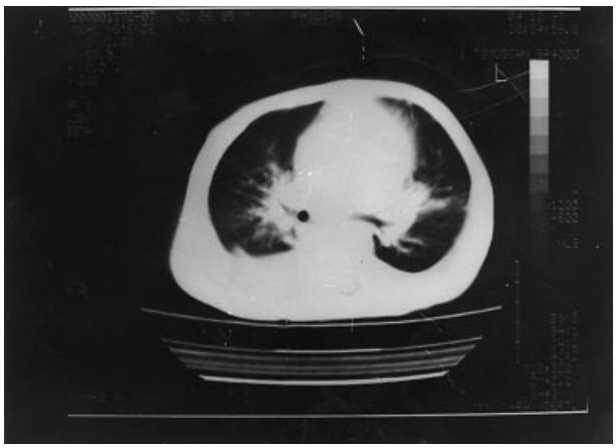

Figure 2 CT scan of the chest showing hilar echogenic (attenuation), right sided pleural effusion and thickened pleura.

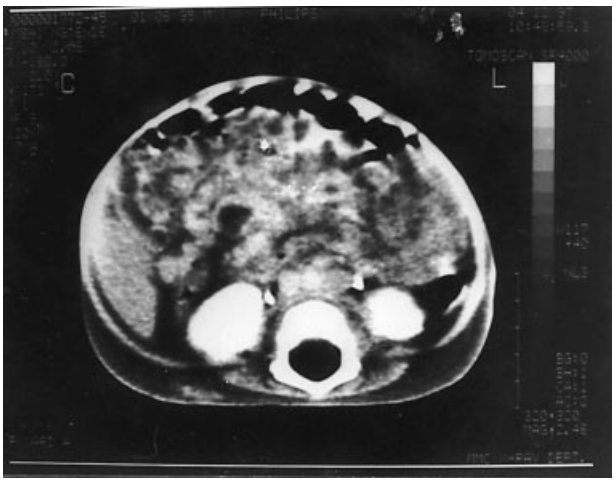

Figure 3 CT scan showing thickened bowel loops, mesenteric and para-aortic lymphadenopathy.

tivity can be potentiated with HIV infection (Ziegler *et al*. 1997). HIV infection continues to increase unabated. With this, KS could be an important cause of further increase in infant mortality. Therefore there is a need not only to prevent HIV transmission, but also HHSV8 infection.

References

Amir H, Kaaya EE, Kwesigabo G, Kitinya JN & Biberfeld P (1997) Kaposi's sarcoma before and during the HIV epidemic in Tanzania: a study of the cancer registry data 1968–95. *International Journal of Oncology* **11**, 1363–1366.

Arico M, Caselli D, D'Argenio P *et al*. (1991) Malignancies in children with human immunodeficiency virus type 1 infection. The Italian multicentre study on human immunodeficiency virus infection in children. *Cancer* **68**, 2473–2477.

Baum LG & Vinters HV (1989) Lymphadenopathic Kaposi's sarcoma in a pediatric patient with acquired immune deficiency syndrome. *Pediatric Pathology* **9**, 459–465.

Connor E, Boccon-Gibod L, Joshi V *et al*. (1990) Cutaneous acquired immunodeficiency syndrome-associated Kaposi's sarcoma in pediatric patients. *Archives of Dermatology* **126**, 791–793.

Gutierrez-Ortega P, Hierro-Orocoz S, Sanchez-Cisneros R, Cuevas F & Montano LF (1989) Kaposi's sarcoma in a 6 day old infant with HIV. *Archives of Dermatology* **125**, 432–433.

Lennette ET, Blackbourn DJ & Levy JA (1996) Antibodies to human herpesvirus type 8 in the general population and in Kaposi's sarcoma patients. *Lancet* **348**, 858–861.

Levy JA (1997) Three new human herpesviruses (HHV 6,7 and 8). *Lancet* **349**, 558–563.

Malekzadeh MH, Church JA, Siegel SE, Mitchell WG, Opas L & Lieberman E (1987) Human immunodeficieny virus-associated Kaposi's sarcoma in a peadiatric renal transplant recipient. *Nephron* **47**, 62–65.

Oettle AG (1962) Geographical and racial differences in the frequency of Kaposi's sarcoma as evidence of environmental or genetic causes. *Acta Unio Int Contra Cancrum* **18**, 330–363.

Olweny CLM, Kaddumukasa A, Atine I, Owor R, Margrath I & Ziegler JL (1976) Childhood Kaposi's Sarcoma: clinical features and therapy. *British Journal of Cancer* **33**, 122–135.

Patil PS, Elem B, Gwavava NJ & Urban MI (1992) The pattern of pediatric malignancy in Zambia (1980–89): a hospital-based histopathological study. *Journal of Tropical Medicine and Hygiene* **95**, 124–127.

Porta F, Bongiorno M, Locatelli F *et al*. (1991) Kaposi's sarcoma in a child after autologous bone marrow transplantation for non-Hodgkin's lymphoma. *Cancer* **68**, 1361–1364.

Schalling M, Ekman M, Kaaya EE, Linde A & Biberfeld P (1995) A role for a new herpes virus (KSHV) in different forms of Kaposi's sarcoma. *Nature Medicine* **1**, 707–708.

Whitby D, Howard MR, Tenant-Flowers M *et al*. (1995) Detection of Kaposi sarcoma associated herpes virus in peripheral blood of HIV infected individuals and progression to Kaposi's sarcoma. *Lancet* **346**, 799–802.

Ziegler JL & Katongole-Mbidde E (1996) Kaposi's sarcoma in childhood: an analysis of 100 cases from Uganda and relationship to HIV infection. *International Journal of Cancer* **65**, 200–203.

Ziegler JL, Newton R, Katongole-Mbidde E *et al*. (1997) Risk factors for Kaposi's sarcoma in HIV-positive subjects in Uganda. *AIDS* **11**, 1619–1626.