

Disseminated Lymphadenopathic Kaposi's Sarcoma in Zambian Children

*** G.P. BHAGWAT**

*** K.G. NAIK**

**** R. SACHDEVA**

**** VIDYA BHUSHAN**

*** Department of Pathology and Microbiology**

**** Department of Paediatrics,
University Teaching Hospital,
LUSAKA.**

SUMMARY

Two cases of Kaposi's Sarcoma with generalized massive lymphadenopathy and visceral involvement without cutaneous lesions have been presented in young Zambian children. The difficulties in diagnosis and clinical course are discussed. We suggest an increasing awareness of this condition for the detection of more cases in Zambia.

INTRODUCTION

Kaposi's Sarcoma with skin involvement is a common tumour in adult Zambian and African population living South of the Sahara (Naik, 1977). However, it is uncommon amongst Zambian children and constitutes only 5% of all Kaposi's in Zambia (Naik, 1977). We wish to present two cases of Kaposi's Sarcoma in Zambian children in view of unpublished literature containing detailed information, in children, its rarity, diagnostic difficulties and the different natural course of the disease.

CASE REPORTS

Case 1:

C.T. a four year old male Zambian child from Eastern Province was admitted to University Teaching Hospital on 5th February 1973, with the complaints of swelling over the face and neck, fever, chest pain and coughing one week prior to admission. On examination, he was anaemic and found to have bilateral massive cervical lymphadenopathy giving rise to bull neck appearance. He also had bilateral axillary lymphadenopathy. He was dyspnoeic and had an ecchymotic patch on right cheek. The spleen was 6 cm below costal margin and liver 3 cm. There was a blood stained nasal discharge. The laboratory investigations showed Hb 5.1 Gm% and total W.B.C. count 16,000/cu mm with neutrophils 40%, lymphocytes 59% and Eosinophils 1%. Reticulocyte count was 5.0%. Platelet count was 15,000/cu mm X-ray chest showed hilar lymphadenopathy. Clinical diagnosis of tuberculosis or lymphoma was made. The patient

was transfused for anaemia and put on prednisone. Bone marrow at this stage was hypercellular with increase in erythropoiesis. Megakaryocytes were present in good number. The lymphnodes started regressing within a week and were palpable only as tiny nodes. But 15 days later, the patient developed generalised oedema, generalized lymphadenopathy and condition deteriorated. A gland biopsy performed after four weeks of admission confirmed the final diagnosis of Kaposi's Sarcoma. The patient was given cyclophosphamide with fresh blood transfusion. A week later, the patient developed right sided hemothorax, generalized petechiae and died five weeks after admission. The significant autopsy findings include massive haemorrhagic lymphnodes matted together in the cervical, axillary, mediastinal and mesenteric regions; petechiae on the skin, mucous membranes and serosal surfaces; mild degree of splenomegaly with scatter of tiny red brown nodules; reddish brown often blackish nodules (0.2 x 0.2 cms to 1.5 x 1 cms) on mucosal surface of jejunum, ileum, caecum, ascending, transverse and descending colon and massive bilateral haemothorax. Histologically Kaposi's Sarcoma, mixed cell pattern was confirmed in all the lymph nodes, spleen and the intestinal tract.

Case 2:

R.C., a four year old male black child from Northern Province was admitted to the University Teaching Hospital in May, 1978. Two weeks prior to admission he developed difficulty in breathing, mild dry cough and low grade continuous pyrexia. The parents noticed enlargement of cervical nodes. On examination he was malnourished, pale and dyspnoeic. There was no cyanosis. He had cervical, axillary and inguinal lymphadenopathy. Lymph nodes were 1 to 3 cm in size, discrete nontender, rubbery and non-suppurative. There were no skin or conjunctival nodules. The liver was 4 cm below the costal margin, soft and nontender. The spleen was not palpable. Coarse crepitations were heard on both sides of chest. Other systems were unremarkable. Investigations on admission showed haemoglobin of 8 gms % with hypochromic anaemia. Total leucocytic count was 6,000/cu mm with 60% neutrophils, 38% lymphocytes and 2% monocytes. E.S.R. was 30 mm in the first hour (Westergren method). Biochemistry was unremarkable. Heaf test was positive. X-ray chest revealed enlargement of hilar and paratracheal nodes with patchy consolidation in both lung fields. Clinically tuberculosis was diagnosed and a lymphnode

biopsy was performed which showed histology consistent with Kaposi's Sarcoma. The condition of the patient deteriorated with increasing dyspnoea and died on 6th day of admission. Autopsy confirmed the clinical findings and in addition showed marked enlargement of mediastinal lymphnodes pressing on and partially obstructing the tracheal lumen. Para aortic and mesenteric lymphnodes were enlarged measuring from 2-5 cm in diameter. The cut surface of lymphnodes showed brownish black areas. The right lung had small blackish nodules of the size of 2 cm x 1 cm in the upper lobe. Both the lungs were congested. Other organs were normal. Microscopically the diagnosis of Kaposi's Sarcoma (mixed cellularity) was confirmed in the lymphnodes and in the right lung. Liver showed fatty change.

DISCUSSION

The incidence of Kaposi's Sarcoma is much lower in Zambian children compared to East and West African children (Templeton, 1973). This is partly due to underdiagnosis. The marked male preponderance in children throughout Africa (Templeton, 1973) is again confirmed in our study. The reasons for such preponderance are unknown. Hutt (1973) has described four clinical varieties of Kaposi's Sarcoma, viz: lymphadenopathic, nodular, florid and infiltrative. The latter three types are common in adults with prolonged survival for years. Lymphadenopathic type is seen frequently in young children and presents with a generalized lymphadenopathy usually without cutaneous manifestations. The patients deteriorated rapidly with generalised lesions often involving the intestines, abdominal lymph nodes and other viscera. Both our cases have similar presentation and rapid downhill course. However, generalised lymphadenopathy may occur with skin nodules or plaques in older children and young adults. Kyalwazi (1969) noted pleural effusions, gastrointestinal haemorrhage, epistaxis and other haemorrhagic manifestations like our first case in 10% cases with wide spread disease. The reasons for the differences in presentation in children and adults remain obscure. The different hormonal status and defective immune system may be suggested for such presentation.

The response to chemotherapy differs with different clinical and histological types of Kaposi's Sarcoma (Vogel et al 1973; Kyalwazi, 1976). They suggested that patients with skin nodules/plaques and with mixed cell histological pattern are most sensitive to chemotherapy and carry the most favourable

prognosis with many years survival. In contrast, young children with wide spread disease have the poorest prognosis with any form of therapy. Our cases confirmed these observations.

Differential Diagnosis:

The causes of childhood superficial lymphadenopathy commonly include acute and chronic non-specific inflammation, tuberculosis, toxoplasmosis, histoplasmosis, infectious mononucleosis, sinus histiocytosis with massive lymphadenopathy, Kaposi's Sarcoma, acute lymphatic leukaemia, lymphomas, etc. It is often difficult to differentiate all these conditions on clinical grounds alone without laboratory and radiological investigations. Often the patients are treated on clinical diagnosis and disappear after initial treatment. This is not uncommon in Zambia. Lymph node biopsy is most essential in diagnosing the various forms of lymphadenopathy. Kaposi's Sarcoma can only be diagnosed by histopathological examination. Histologically Kaposi's Sarcoma requires differentiation from angiomas, granulation tissue, angiosarcoma, lymphangiosarcoma, mesotheliomas, fibroxanthosarcoma, leiomyosarcoma, rhabdomyosarcoma, lymphoma, undifferentiated carcinoma and sarcoma. The histological features, like interlacing bundles of spindle shaped cells, vascular slits containing red cells, vascular channels of varying sizes and degree of differentiation (Fig. 1), inflammatory cells, recent or old haemorrhage, presence of hyaline bodies; often supplemented by special stains – reticulin, PTAH, Masson's trichrome, phloxine tartarazine – differentiate Kaposi's Sarcoma from the above lesions.

ACKNOWLEDGEMENT

We wish to thank Mr M.A. Ansary of the Medical Illustration Unit of the University Teaching Hospital, Lusaka for the illustration.

REFERENCES

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2. S.K. Kyalwazi (1969); *East African Medical Journal, 46, No. 8, 459.*

Fig. 1:



Kaposi's Sarcoma: Showing spindle shaped cells, vascular slits and vascular channels of varying sizes with in-between red cells and mononuclear cells (Magnification x 666)

3. S.K. Kyalwazi (1976); *East African Medical Journal, 53, No. 12, 670.*
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