HISTOPATHOLOGICAL STUDY OF KAPOSI’S SARCOMA IN JOS: A 16-YEAR REVIEW

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Abstract

Background/objective: To study the pathology of Kaposi’s sarcoma and review relevant literature on this condition.

Method: A retrospective analysis of histologically confirmed cases of Kaposi’s sarcoma over a period of 16 years was undertaken. Fresh sections of slides were reviewed independently by two pathologists.

Results: A total of 65 cases of Kaposi’s sarcoma were documented, accounting for 1.37% of all cancers seen within the period. Forty five were males and 20 females giving a ratio of 1:0.25. Leg and foot lesions accounted for 58.5% of all the lesions. Sixteen (24.16%) cases were HIV positive. The peak age at presentation was 26-45 years (range 12 - 60 years). The predominant histological pattern was spindle cell formation with fibroblastic background and vascular slits lined by plump anaplastic endothelial lining filled red blood cells. Patients with single lesion and multiple lesions confined to restricted areas showed regression after a combination of alpha interferon and cytotoxic drugs administration.

Conclusion: The prevalence of Kaposi’s sarcoma seems to be on the increase in our environment. The control measure being put on forward by various agencies may help to slow down the emergence of epidemic Kaposi’s sarcoma.

Key words: Kaposi’s sarcoma, anatomical sites, HIV

Introduction

Kaposi’s sarcoma was reported first by the Hungarian dermatologist Moritz Kaposi in 1872, and was thought to occur only in Eastern Europe and the Mediterranean. However, Kaposi’s sarcoma was first reported in Africa in the late 50s and 60s as endemic form occurring in young Africans. The classical description of this disease in central, eastern and southern Africa showed that Kaposi’s sarcoma accounted for 3-9% of all cancers.

Kaposi’s sarcoma, though a disease observed in Europe, its appearance in tropical Africa was believed to be associated with occupational exposure of the lower extremities. It is now associated with decreased immune surveillance. It has been described as endemic, sporadic or epidemic, depending on the epidemiological description.

Since the emergence of human immunodeficiency virus (HIV) infection, there has been a steady increase in the prevalence of Kaposi’s sarcoma world wide. All ages are affected, but lymph node involvement is more frequent in children and adolescents. The disease predominantly affects men (M: F of 12:1 - 3:1). The lower limbs are reported to be the predominant site affected in Africans.

The pathogenesis of Kaposi’s sarcoma is largely obscure and has been a subject of debate. The tumour is vascular in origin and has been described as multiple, haemorrhagic, pigmented angiosarcoma. The lesion characteristically consists of thin walled endovascular formation, extravasated red cells, inflammatory lymphocytes and proliferating spindle cells. The spindle cells appear to be the primary neoplastic component. Electron microscopic studies showed that the spindle cells or the plump cells appear to arise from pericytes, endothelial or smooth muscle fibres. However special reticulin stain has shown inconsistent findings. Extracts of spindle cells from Kaposi’s sarcoma lesion when injected into subcutaneous tissues of rats invoked sarcomatous changes.

In 1994 Chang et al identified DNA fragments of a previously unrecognized herpes virus (which has been called Kaposi’s sarcoma associated herpes virus or KSHV, also known as HHV8), in Kaposi’s sarcoma skin lesions from patients with acquired immune deficiency syndrome (AIDS). Subsequent studies have showed that in over 95% of Kaposi’s sarcoma, KSHV is present regardless of HIV status. It has now been established that KSHV is the primary and necessary factor in the development of Kaposi’s sarcoma.

In the past it was believed that Kaposi’s sarcoma is a unifocal lesion with potential of metastasis. However recent studies have shown that the disease is
multifocal and is vascular in origin.\textsuperscript{5-9}

The aim of this study was to examine the pathology of Kaposi’s sarcoma, mode of presentation, sex distribution, and have a better understanding of its biological behaviour.

**Materials and Methods**

This is a retrospective study of histologically confirmed cases of Kaposi’s sarcoma seen at the Department of Pathology of Jos University Teaching Hospital, Jos, Nigeria, over a period January 1987 – December 2002. Tissue blocks were re-examined independently by two pathologists. Where necessary, reticulin special stains were carried out to determine the origin of the tumour. Information collected from the records (case notes and referral notes) included age, sex, site of lesion and HIV status. Clinically suspected cases of classical Kaposi’s sarcoma lesions were excluded as biopsy was not done.

**Results**

A total of 4757 histologically confirmed malignant tumours were recorded over the period of 16 years. Of this, 65 (1.37%) were Kaposi’s sarcoma. Out of the 65 cases of Kaposi’s sarcoma, 45 were males and 20 females (M: F = 1:0.25).

Table 1 shows the anatomic distribution of the lesions. The commonest site in both sexes was the legs and foot which accounted for 58.5%. A total of 16 cases were HIV positive out of 45 patients who were screened. Patients with confirmed diagnosis of Kaposi’s sarcoma and who were also HIV positive were only available between 1995-2002. The age range was 12 - 60 years, and peak age 26-45 years.

Table 2 shows the phases of Kaposi’s sarcoma. The commonest phase was the mixed type which consists of a mixture of spindle cells, vascular clefts filled with red blood cells and haemosiderin pigments.

**Table 1: Anatomical distribution of Kaposi’s sarcoma in 65 patients**

<table>
<thead>
<tr>
<th>Anatomical site</th>
<th>Male</th>
<th>Female</th>
<th>No. of HIV positive cases</th>
<th>Total</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Leg</td>
<td>14</td>
<td>6</td>
<td>2</td>
<td>20</td>
<td>30.8</td>
</tr>
<tr>
<td>Foot</td>
<td>13</td>
<td>5</td>
<td>8</td>
<td>18</td>
<td>27.7</td>
</tr>
<tr>
<td>Lymph node</td>
<td>5</td>
<td>-</td>
<td>-</td>
<td>5</td>
<td>7.7</td>
</tr>
<tr>
<td>Gastrointestinal tract</td>
<td>5</td>
<td>2</td>
<td>2</td>
<td>7</td>
<td>10.8</td>
</tr>
<tr>
<td>Face</td>
<td>8</td>
<td>7</td>
<td>3</td>
<td>15</td>
<td>23</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>45</strong></td>
<td><strong>20</strong></td>
<td></td>
<td><strong>65</strong></td>
<td><strong>100</strong></td>
</tr>
</tbody>
</table>

**Table 2: Histological phases of Kaposi’s sarcoma in 65 patients**

<table>
<thead>
<tr>
<th>Type</th>
<th>No. (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mixed</td>
<td>40 (61.5)</td>
</tr>
<tr>
<td>Monomorphic</td>
<td>13 (20)</td>
</tr>
<tr>
<td>Anaplastic</td>
<td>12 (19.5)</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td>65 (100)</td>
</tr>
</tbody>
</table>

**Discussion**

Sixty five histologically confirmed cases of Kaposi’s sarcoma were seen during the 15-year period. This represents 1.37% of all cancers diagnosed during the period. This figure is small compared to reports from central, east and southern Africa which was put at 1.5-12% of all cancers. \textsuperscript{1,2-12} In the present study the M: F ratio of 1:0.25 confirms that the disease is predominant in males. The lesions were predominantly on the legs and feet as noted in other reports from Nigeria and other part Africa. \textsuperscript{2, 4, 6 - 8} Why Kaposi’s sarcoma is common on the lower limbs is not clearly understood. In the past the disease was believed to be multicentric in origin. \textsuperscript{5, 6} However, recent findings show that it is monoclonal in origin and that the transformed cells home to various tissues, including blood vessels. \textsuperscript{10, 12-15} It has been reported that KSHV is present in both sporadic and endemic of Kaposi’s sarcoma. The current hypothesis is that the monoclonal cells arising from Kaposi’s sarcoma may have receptors for the virus, which may act as a promoter in an already initiated cell. \textsuperscript{5, 6} The hypothesis is yet to be confirmed.

The recently discovered HHV8 in the lesions of Kaposi’s sarcoma has given it a strong role in the pathogenesis of this disease. KSHV is reported to be present in more than 90% of Kaposi’s sarcoma lesion. This virus releases cytokines as well as HIV tat protein which is believed to contribute to the pathogenesis of Kaposi’s sarcoma. However the presence of KSHV appears to be a necessary factor in the development the disease. \textsuperscript{12-17}

The few cases of Kaposi’s sarcoma documented histologically do not represent all the cases of Kaposi’s sarcoma seen within the period. Clinically suspected cases that had no biopsies were excluded from the study. No case of transplant recipient or patient on immunosuppressive therapy was biopsied. Studies in Toronto and France showed that Kaposi’s
sarcoma occurs in 1-5.3% of patients who either have transplantation or are on immunosuppressive therapy. The commonest histological pattern was mixed type in this study as described by Kyalwazi in 1976.

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Reference