Osteosarcoma of the Maxillofacial bones in Kenyans

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SUMMARY. Osteosarcoma (OS) is a highly malignant tumour and is the most common primary neoplasm of bone, although rare, especially in the maxillofacial skeleton. This article presents 14 Kenyan cases of OS of the maxillofacial bones seen between January 1991 and July 1997: 11 in the mandible, two in the maxilla and one in the right zygomatic arch. Patients ranged in age from one week to 50 years (Mean = 29.7), with an equal gender distribution. While pain and rapid swelling were the commonest clinical features, the radiographic and histopathological characteristics were as varied as has been described elsewhere. Generally, effective management of most of the cases was poor due to late presentation for treatment.

INTRODUCTION

Osteosarcoma (OS) is a highly malignant tumour and is the most common primary neoplasm of bone, although overall it is rare, especially in the maxillofacial bones (Caron et al., 1971; Russ and Jesse, 1980). In the jaws, OS characteristically affects patients considerably later than in the long bones, namely at a mean age of 30 to 39 years (Archer and Langdon, 1992). Approximately 6.5 and 10% of all OS arise in the cranial bones (Caron et al., 1971; Batsakis, 1987). In one series of 21 cases of OS in a West African population, Ajagbe et al. (1986) found a frequency of 1.5% and an age range of 10 to 47 years, with a mean of 27 years.

In Eastern Africa, there is a dearth of information regarding the pattern of occurrence and clinical characteristics of maxillofacial OS. In this paper, we present the clinico-radiological and histopathological characteristics of 14 Kenyan cases of OS seen over a 6½-year period.

PATIENTS AND METHODS

Between January 1991 and July 1997, 14 patients with OS of the maxillofacial region were diagnosed in the Oral and Maxillofacial clinic of the University of Nairobi Dental Hospital (UNDH). Patients were documented as they were referred for specialist evaluation and management. In addition to the comprehensive clinical examination, relevant radiological investigation was performed on each patient. Confirmatory histopathological examination of each case was then independently performed by pathologists at the UNDH and the Southern African Centre for Orofacial Diseases (SACOD).

In each case, Haematoxylin and Eosin (H & E) staining was employed in the usual way for histopathological evaluation. In some of the cases, additional special stains, including those for cytokeratin and vimentin, were utilized. In all the cases documented, the histopathological reports prepared by both centres (UNDH and SACOD) were unanimous.

RESULTS

Of the 14 cases of maxillofacial OS, seven were male and seven female, with an age range of one week to 50 years (mean = 29.7 years). While two lesions occurred in the maxilla, 11 were in the mandible: five in the right and six in the left mandible. One case had a lesion in the right zygomatic arch. Table 1 shows the general clinical characteristics of all the patients. Pain and rapid swelling were the most salient features at presentation.

<table>
<thead>
<tr>
<th>Case</th>
<th>Age (Years)</th>
<th>Sex</th>
<th>Site of lesion</th>
<th>Clinical features</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.</td>
<td>1 week</td>
<td>F</td>
<td>Left mandible</td>
<td>Swelling, ulceration</td>
</tr>
<tr>
<td>2.</td>
<td>17</td>
<td>M</td>
<td>Left mandible</td>
<td>Swelling, pain, ulceration</td>
</tr>
<tr>
<td>3.</td>
<td>11</td>
<td>F</td>
<td>Right mandible</td>
<td>Swelling, pain, ulceration</td>
</tr>
<tr>
<td>4.</td>
<td>26</td>
<td>F</td>
<td>Right mandible</td>
<td>Swelling, pain, ulceration</td>
</tr>
<tr>
<td>5.</td>
<td>45</td>
<td>F</td>
<td>Left mandible</td>
<td>Swelling, pain, ulceration</td>
</tr>
<tr>
<td>6.</td>
<td>13</td>
<td>M</td>
<td>Left mandible</td>
<td>Swelling, pain, ulceration</td>
</tr>
<tr>
<td>7.</td>
<td>23</td>
<td>F</td>
<td>Right maxilla</td>
<td>Swelling, pain, ulceration</td>
</tr>
<tr>
<td>8.</td>
<td>48</td>
<td>F</td>
<td>Right mandible</td>
<td>Swelling, pain, ulceration</td>
</tr>
<tr>
<td>9.</td>
<td>30</td>
<td>M</td>
<td>Left mandible</td>
<td>Swelling, toothache</td>
</tr>
<tr>
<td>10.</td>
<td>27</td>
<td>M</td>
<td>Left maxilla</td>
<td>Swelling, pain</td>
</tr>
<tr>
<td>11.</td>
<td>50</td>
<td>F</td>
<td>Right mandible</td>
<td>Swelling, pain</td>
</tr>
<tr>
<td>12.</td>
<td>26</td>
<td>M</td>
<td>Right zygomatic arch</td>
<td>Swelling, pain</td>
</tr>
<tr>
<td>13.</td>
<td>50</td>
<td>M</td>
<td>Right mandible</td>
<td>Swelling, pain</td>
</tr>
<tr>
<td>14.</td>
<td>50</td>
<td>M</td>
<td>Left mandible</td>
<td>Swelling, pain</td>
</tr>
</tbody>
</table>

n = 14; Average age = 29.7 years.
Radiographic evaluation revealed a variable pattern of osteoblastic and osteolytic activities in almost all the lesions. The lone right zygomatic arch lesion exhibited an exuberant osteoblastic activity radiographically, as depicted in Figure 1.

Histologically, there was also a varied pattern of osteoblastic, chondroblastic and myxoid differentiation activity among the cases. Figure 2 shows a highly chondroblastic type of OS (specimen from the youngest patient in the present series).

The youngest patient in this series (a 1-week-old female neonate) was most intriguing at presentation (Fig. 3 taken at 3 weeks of life) as OS was least expected at this age. Wide extirpation of the tumour was accomplished under general anaesthesia (Fig. 4) and the postoperative recovery was uneventful (Fig. 5).

The modalities of tumour management for all the patients are summarized in Table 2. Generally, the response to all the modalities of management was poor due to the fact that most of the patients presented late for specialist evaluation. Poor compliance with postoperative follow-up is our main obstacle in attempting to evaluate the overall prognosis of the management of OS. Most of the patients (at least six) who were consistently followed-up for periods ranging from 2 to 6 months, had extensive recurrences from
which they subsequently died. The neonate was lost to follow-up within 2 months of operation. One case still remains free of the tumour at least 5 years after surgery and neoadjuvant chemotherapy (case 9, JM).

DISCUSSION

While OS has also been called osteogenic sarcoma, the term osteosarcoma should be preferred as it emphasizes the fact that this malignant neoplasm produces bone and is a primary neoplasm of bone (Pindborg and Eversole, 1992). However, approximately a quarter of osteosarcomas show relatively little ossification and are commonly referred to as osteolytic types (Hu vos et al., 1982). In the maxillo-mandibular region, osteosarcomas are frequently parosteal and osteoblastic in nature and their biological behaviour differs from that of tumours involving other skeletal bones in that the average age of onset is 20 years later than for skeletal lesions, the histopathological variables are more favourable, distant metastases occur less frequently and survival rates are higher (Russ and Jesse, 1980).

In the present series, the documented clinical features, radiographic appearances and the histopathological variables are consistent with those published elsewhere. Radiologically, the findings may be the same as those of OS in other sites – osteolytic, osteoblastic or a mixture of the two with the poorly defined irregular margins that are characteristic of malignant lesions (Doval et al., 1997). Furthermore, because of the complexity of the clinical features and radiography, the final diagnosis can be made only after histopathological analysis is carried out, whereupon evidence of sarcomatous stroma must be seen to elaborate osteoid or primitive bone.

Remarkably, our youngest patient at diagnosis of OS was only one week old. The patient’s mother had apparently noticed the growth within about 5 days of birth. Therefore, it may be pertinent to infer that the lesion could have manifested itself during the infant’s intrauterine life. Such an occurrence would be intriguingly rare. The histopathological appearance of the biopsy specimen was highly chondroblastic to the extent that a diagnosis of chondrosarcoma could have been entertained until foci of osteoblastic activity were identified. As a general consensus among pathologists worldwide, the hallmark of the diagnosis of OS is the ability of the malignant cells to produce osteoid even in a highly chondroblastic lesion. Occurrence of neonatal malignant lesions of the mesenchyme may tend to produce generally intriguing cellular patterns that can be difficult to characterize. Rapid diagnosis of such lesions may often be rendered difficult.

Although only a few biopsy specimens were subjected to a variety of the emerging histocytological techniques, it appears that there may be no specific pattern that may emanate from their utilization to facilitate the rapid diagnosis and characterization of the various OS entities of the skeleton in general.

The essential feature of treatment of OS of the maxillofacial bones is aggressive local therapy involving adequate surgical extirpation (Russ and Jesse, 1980). In combination with neoadjuvant chemotherapy as currently practised, encouraging results with regard to disease control may be obtained. In our experience, aggressive treatment may only be useful if patients report early and the neoplasm is identified promptly. Notably, adequate chemotherapy with radical surgery has probably played an important part in locoregional control and survival in patients with OS of the jaw bones (Doval et al., 1997).

Unfortunately, most patients in resource-poor conditions are seen late and the definitive diagnosis may take weeks to be available. Under these circumstances the surgical treatment of an extensive highly malignant tumour in the maxillofacial bones can only yield highly unreliable expectations for the patient and clinician. Furthermore, with the advent of cost-sharing for health provision, the ability for most, if not all patients to afford the current neoadjuvant chemotherapy regimes has diminished tremendously. Therefore, our experience with the management of
osteosarcoma has sadly remained generally dismal as we have been able to offer only salvage pain treatment for nearly half of the patients who presented with extensive tumours.

ACKNOWLEDGEMENTS

We are greatly indebted to Prof. Eric J. Raubenheimer of the Southern Africa Centre for Orofacial Diseases for the verification of the histopathological analysis of most of the cases included in this report. We are also grateful to the administration of the Kenyatta National Hospital, Dental Unit and University of Nairobi Dental Hospital for permission to present these patients. Finally, sincere thanks to Keziah Mbuguah for preparing the manuscript.

REFERENCES


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Paper received 9 December 1997
Accepted 22 January 1998