Management of Maxillofacial Osteosarcomas in Kenya: A Case Series
Symon Guthua, Martin Kamau, Nicholas Abinya
University of Nairobi

Correspondence to: Dr. Martin Kamau, University of Nairobi P.O Box 19676-00202, Nairobi, Kenya. Email: mrtnkamau@yahoo.com

Abstract
Background: Though uncommon, Maxillofacial osteosarcomas represent a unique challenge in management due to a multitude of factors such as difficulty in diagnosis, local complex anatomy making surgical excision difficult as well as debate on usefulness of adjunct treatment modalities such as chemotherapy and radiotherapy. Nonetheless they represent a significant health burden due to their high morbidity and mortality. Methods: Retrospective cross-sectional study of records archived in the School of Dental Sciences. Results: 25 cases of Maxillofacial sarcomas were seen over a span of 26 years. Mean age of occurrence was 35.68 years with a preponderance for females (17 cases). The mandible was the most affected site accounting for 18 cases.

Discussion: The management of maxillofacial sarcomas presents significant challenges arising from multiple factors such as lack of standardised treatment protocol as well as late presentation of patients, diagnostic challenges and loss to follow-up in our setting.

Key words: Maxillofacial osteosarcomas, Management Protocol, Kenya
Ann Afr Surg. 2019; 17(1):***
DOI: http://dx.doi.org/10.4314/aas.v16i1.*

Conflicts of Interest: None

Funding: None © 2019 Author. This work is licensed under the Creative Commons Attribution 4.0 International License.

Introduction
Osteosarcoma is the most common primary neoplasm of bone, with an incidence of 1: 100,000 (1,2). Most head and neck sarcomas are of the soft-tissue type with only 20% being of bony or cartilaginous origin (3). In the USA, it is estimated that less than 4% of all recorded osteogenic sarcomas occur in the jaw (4). The mandible and maxilla are the predominant sites of head and neck osteosarcoma (HNOS), although extragnathic bone as well as soft tissues sites may be affected [6]. The 5-year disease-specific survival rate for patients with HNOS has been poor, with most studies reporting survival rates of 23-37% with the maxillary tumours showing worse prognosis (5,6,7,8). Maxillofacial sarcomas have been noted to have a biological behaviour that differs from that of the long bones in that the average age of onset is 10-20 years later than their skeletal variants, distant metastasis are rarer and survival rates are more favourable (9). The mainstay of treatment for osteosarcoma of the jaws is complete surgical excision with free margins as well as reconstruction to improve function and quality of life post treatment. The use of multidrug chemotherapy (both neoadjuvant and adjuvant) in the management of skeletal OS is well established with notable improvements in survival rates but the same cannot be said for OS in the maxillofacial region. Two meta-analyses reviewing the outcomes related to the addition of chemotherapy regimes in Head and neck OS showed conflicting results (10). Nonetheless, several individual centre reports have shown a benefit with the addition of neoadjuvant and/or adjuvant chemotherapy (11). In the maxillofacial region extensive lesions may provide a challenge where the complex local anatomy and proximity to vital structures may preclude the wide excision with adequate tumour free margins.

In this paper, we review 25 cases of OS seen over a period of 26 years in one specialized department in a teaching institution.

Materials and Methods
The study was conducted at the Department of Oral and Maxillofacial Surgery, School of Dental Sciences, University of Nairobi utilising patient clinical and histopathologic records archived at the Department. This was a retrospective cross-sectional study analysing all the records of patients who had a histologic diagnosis of maxillofacial OS. Patient clinical and histopathologic records were retrieved and data recorded in a data collection form. Incomplete patient
records were excluded from the study. Ethical approval was obtained from the Ethics, Research and Standards committee of the Kenyatta National Hospital and the University of Nairobi (Approval number: P170/6/2009).

**Results**

A total of 25 cases were analysed. With regard to gender predilection, women were more affected (17 cases) than males (8 cases). The mandible was the most common site afflicted (18 cases) while 7 cases were in the Maxilla. The age range was from 1 week old to 70 years. 15 patients were below the age of 39 years. The mean age of occurrence was 35.68 years. Table 1 shows the distribution of the cases by age.

<table>
<thead>
<tr>
<th>Age group (years)</th>
<th>Osteosarcoma Cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>0-9</td>
<td>1</td>
</tr>
<tr>
<td>10-19</td>
<td>4</td>
</tr>
<tr>
<td>20-29</td>
<td>6</td>
</tr>
<tr>
<td>30-39</td>
<td>4</td>
</tr>
<tr>
<td>40-49</td>
<td>3</td>
</tr>
<tr>
<td>50-59</td>
<td>3</td>
</tr>
<tr>
<td>&gt;60</td>
<td>4</td>
</tr>
</tbody>
</table>

The histopathologic subtypes varied between the osteoblastic, fibroblastic and chondroblastic high grade variants. Of note, one patient was diagnosed with an aggressive telangiectactic/vascular type of osteosarcoma while another had a discordant diagnosis: initial histological diagnosis was osteoblastoma later revised to osteosarcoma after review by panel of pathologists. Musculo-skeletal Tumour Society Cancer Centre staging was only noted for 20 patients who were in the surgery and chemotherapy group as well as the palliative chemoradiotherapy group (3). The patients were all in stage II. No staging was noted for 5 patients who had been treated earlier in this series with either surgery alone or surgery and radiotherapy. No details were noted from the clinical records of why these patients were offered these treatment modalities.

Over the past 26 years the modalities of management have mainly been multimodal therapy involving neoadjuvant chemotherapy then cytoreduction therapy followed by postoperative adjuvant chemotherapy a noted in the table 2 below.

<table>
<thead>
<tr>
<th>Treatment modality</th>
<th>Number of Patients</th>
<th>Outcomes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Surgery alone</td>
<td>4</td>
<td>Unknown-All lost to follow-up post-treatment</td>
</tr>
<tr>
<td>Surgery + chemotherapy</td>
<td>13</td>
<td>5 patients still on follow-up 10 years. Stable post-treatment</td>
</tr>
<tr>
<td>Surgery + radiotherapy</td>
<td>1</td>
<td>Unknown- Lost to follow-up post-treatment</td>
</tr>
<tr>
<td>Palliative chemoradiotherapy</td>
<td>7</td>
<td>All deceased</td>
</tr>
</tbody>
</table>

**Case – Mandibular osteosarcoma**

Patient MM, a 54-year-old female, was referred with a swelling of the right side of the mandible of one-month duration (figure 1). She had impaired sensation of the right inferior alveolar nerve. There was no significant medical history apart from hyperacidity related to non-steroidal anti-inflammatory drugs.

![Fig. 1A: Extraoral appearance at presentation](image1)

![Fig. 1B: Intraoral appearance at presentation](image2)

A CT scan revealed a lytic soft tissue mass arising from the right mandibular alveolus and extending to the surrounding soft tissues (Figure 2).

![Fig. 2A: CT scan – axial view.](image3)

The mass is marked by green arrows

![Fig. 2B: Coronal view](image4)

Histopathology sections (Figure 3) showed features of Osteosarcoma – osteoblastic type (high grade).
There was no evidence of metastasis of the tumor to the chest as confirmed by a high-resolution chest CT scan (Fig. 4).

Management protocol for the patient included multimodal treatment as follows:
1. Neoadjuvant chemotherapy - 3 cycles
2. Surgery (post-chemotherapy) and initial primary reconstruction - Tumor specimen was resected en bloc and reconstructed with a 2.5mm preformed titanium plate and soft tissue (cheek and floor of mouth) augmentation with a pectoralis major pedicled flap (Figure 5).
3. Post op chemotherapy – 4 cycles after 6 weeks post-operatively (Figure 6).

Discussion
Generally, maxillofacial mesenchymal malignancies are rare. In the present study, 25 maxillofacial osteosarcomas were noted over a 26-year period. The demographic distribution of maxillofacial sarcomas noted some trends. In our study, the most frequent site of occurrence was the mandible which concurred with most of the reports in literature (1,3). Analysis of age distribution of maxillofacial osteosarcomas in this study noted that the mean ages at first presentation of maxillofacial osteosarcoma were consistent with data from other African studies (12). The lower mean age of occurrence of osteosarcoma is probably influenced by the lower age expectancy in the Kenyan population (52 years in 2006) (12). As regards gender distribution, there was a female preponderance which was in contrast to literature which did not note any real preference to any gender (1,2,12). The management of Osteosarcoma remains a challenge. Firstly, these tumours remain relatively rare such that they may be considered as “orphan diseases” (13). In our case series we encountered 25 cases over a span of 26 years. Secondly, they remain a challenge to diagnose. Our series did have a case that was initially diagnosed as an osteoblastoma. The clinical behaviour of the lesion did not correspond with the histological examination. As such re-evaluation of the tissue
specimen was requested and a diagnosis of osteosarcoma was made. Finally, there are challenges in the management of the tumours due to not only their aggressive nature but also due to a lack of consensus as to the mode of management. There is a dearth of literature regarding management of maxillofacial sarcomas and the few that are available do not provide consensus as to the treatment protocol. While it is clear that aggressive surgical extirpation is a necessity, the use of chemotherapy in Maxillofacial osteosarcomas remains controversial (5,10). While the utility of chemotherapy in the management of appendicular (long bone) osteosarcoma is well proven, there has been conflicting evidence concerning its use in the head and neck (11). Nevertheless, it does appear that its use does suggest some advantage over the surgery alone (4). The use of radiotherapy is not as effective as chemotherapy as Osteosarcomas have been shown to be relatively radio-resistant. The five patients in this study who remained disease free over a period of 10 years had gone through the management approach involving neoadjuvant chemotherapy (3 cycles) followed by complete surgical extirpation and then adjuvant chemotherapy (4 cycles). From our case series, the overall outcomes were relatively poor with all patients presenting in stage III and this may have been attributed to late presentation of patients with these aggressive lesions. There still remains a challenge of follow-up of patients in our setting. Of the 25 cases in our case series, 12 patients were lost to follow-up possibly due to socio-economic factors, 7 patients were deceased and 5 patients are still on follow-up (10 years median time).

In general, the management of sarcomas remains a challenge essentially due to the rarity of these cancers, their biological diversity and resultant difficulty in their diagnosis, the lack of a large multicentre evidence base for treatment, the complexity and intensity of the treatment regimens and the high mortality from these cancer types. The National Cancer Control Network (NCCN, US) and the National Institute for Health and Clinical Excellence (NICE, UK) have developed detailed recommendations limiting their management to MDT sarcoma teams with Specialist Sarcoma - Surgeons, Pathologists, Radiologists, Oncologists as well as support staff (14).

Conclusion
The management of Jaw sarcomas in Kenya remains a challenge. Possibly, embracing the sarcoma centre model would improve outcomes by concentrating patient load and expertise in these centres with attendant advantages of long-term follow-up and development of evidence-based treatment protocols.

Acknowledgements
The authors would like to thank the staff of the Department of Oral and Maxillofacial Surgery, School of Dental Sciences, University of Nairobi, for their support.

References