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Maxillofacial sarcomas: a ugandan epidemiological survey

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ABSTRACT

We reviewed the case notes of 203 patients who were treated for sarcomas of the oral and maxillofacial region over a period of 5 years. There were 98 male cases (mean age 31.4 ± 12.8 years) and 105 female cases (mean age 29.3 ± 10.4). Kaposi's sarcoma accounted for 82.8% cases, and rhabdomyosarcoma 6.9% followed by osteosarcoma and chondrosarcoma at 2.5% each. Except for Kaposi's sarcoma, surgery in combination with radiotherapy and/or chemothe- rapy was the main stay of treatment. Survival data was not available for most of our patients.

Keywords: Maxillofacial Sarcomas, Kaposi's Sarcoma, Maxillofacial Tumors, Head and Neck Tumors

1. INTRODUCTION

Sarcomas of the head and neck region are rare malignancies often without a clear etiology. They present with variable symptoms usually dependent on the affected anatomical structure. Symptoms range from painless masses, hearing loss, vertigo, tinnitus, to facial paralysis. Masses that affect the oral cavity can induce dental pain and, loosening of teeth. When they affect vital structures of the neck they may cause dysphagia, hoarseness, and even dyspnea [1]. Diagnosis is based on histological evaluation highly dependent on expert and experienced pathologists who at times need to employ special immunohistochemical staining techniques to confirm the diagnosis and when in doubt outside opinion is sought. Both these avenues may not readily be avail- able in developing countries therefore reliance on the consensus of a local panel of pathologist for confirmation is the norm [2].

Generally oral maxillofacial malignancies contribute a small percentage to the overall incidence of this disease entity [3,4]. Sarcomas are even rarer contributing 1% to 15% of maxillofacial tumors [1,5-7].

Pathologic classification has been reported as critical to the ultimate treatment and prognosis of head and neck sarcomas. Different studies have reported Kaposis' sarcoma, osteosarcoma, rhabdomyosarcoma, malignant fibrous histiocytoma, and angiosarcoma as the most common types that occur in the head and neck region [8-10]. However, the HIV pandemic has changed the overall incidence and prevalence of sarcomas in sub Saharan Africa due to an increase in Kaposi's sarcoma [11]. In recent times an increase in availability of highly active retroviral therapy has been reported to reduce Kaposis' sarcoma incidence among HIV/AIDS patients [12]. Decline in Kaposi's sarcoma has been reported in Tanzania with an increase in antiretroviral therapy availability [13]. We didn't find a report on head and neck sarcomas in Uganda therefore we felt the need to carry out a retrospective survey of prevalence of this entity in the maxillofacial region.

2. MATERIALS AND METHODS

Patient data were obtained from a retrospective search of medical records at the Maxillofacial Unit, Mulago hospital, a national referral and Teaching Hospital of Makerere University College of Health Sciences seen from January 2006 to December 2010. Records of cases of histopathologically diagnosed as sarcomas were selected and patient files traced. The histological type, gender, age at initial diagnosis, presenting complaint, site of tumor and treatment were retrieved. Comparison with published data was done using simple students T test (p 0.05) with the help of Medcalc.

3. RESULTS

3.1. Prevalence and Histopathological Types

There were 2084 cases biopsied at the oral and maxillofacial unit within the study period. Out of which 203 (9.7%) were sarcomas. Eleven histological types of sarcoma were found from the retrieved data. Kaposis' sarcoma was the most common lesion followed by rhabdomyosarcoma then osteosarcoma and chondrosarcoma (**Table 1**).

Table 1. Gender and histopathological distribution of sarcoma.

Neoplasm	Sex		No. (%)
	Female	Male	
Kaposi's sarcoma	87	81	168 (82.8)
Rhabdomyosarcoma	5	4	9 (4.4)
Osteosarcoma	3	3	6 (2.9)
Embryonal rhabdomyosarcoma	3	2	5 (2.5)
Chondrosarcoma	2	3	5 (2.5)
Hemagio pericytoma	1	3	4 (1.9)
Malignant fibrous hystiocytoma	0	2	2 (1)
Myxoid liposarcoma	1	0	1 (0.5)
Fibro sarcoma	1	0	1 (0.5)
Dermato fibrosarcoma	1	0	1 (0.5)
Reticular cell sarcoma	1	0	1 (0.5)
Total	105	98	203 (100)

3.2. Sex and Age Distribution

There was no gender preponderance, with a male to female ratio of 1:1.1 (**Table 1**). The age range was 1 to 76 years (mean 30.3 ± 11.6 years, median and mode were both at 30 years) in **Table 2**. The third and fourth decade equally dominated at 32% each.

3.3. Tumor Site Distribution

There were more soft tissue sarcomas in the maxillofacial region than hard tissue (5.4%). Vascular tumors as a group (Kaposi's sarcoma and hemagiopericytoma) were the commonest followed by the muscular group (rhabdomyosarcoma and embryonal rhabdomyosarcoma). Kaposi's sarcoma mainly was seen on the palate but the oral cavity as a whole accounted for the second highest reported site. The mandible was the most commonly affected jaw bone by both rhabdomyosarcoma, osteosarcoma and chondrosarcoma (**Table 3**).

3.4. Clinical Features and Duration of Symptoms

As per **Table 4**, a combination of pain and swelling were the commonest chief presenting complaints of the patients (43.6%). This was followed by swelling alone. Seven (6.4%) cases all of Kaposi's sarcoma came because they were referred. Patients presented within 1 - 205 weeks of onset of symptoms. However, the 205 weeks case was an outlier diagnosed as a fibrosarcoma. The others that presented after long durations were Kaposi's sarcoma cases. Two cases of chondrosarcoma were initially diagnosed as pleomorphic adenoma and chondroma hence they were treated conservatively with surgery only to fail loco-regionally. We don't routinely stage but we always have chest x-rays and abdominal ultrasound scans done before treatment except for Kaposi's sarcoma. In all cases they were no significant findings.

3.5. Treatment and Follow-Up

The treatment modalities differed as per the diagnosis. Kaposis sarcoma was mainly managed by chemotherapy and highly active antiretroviral therapy (HAART). There is an ongoing blinded study treatment Kaposi's sarcoma so we couldn't establish who received what. However, we were unable to establish HIV/AIDS management situation for 41.2% of the patients with Kaposi's sarcoma. Out of those whose HIV treatment history was retrievable, 17.7% were on HAART at time of diagnosis, while 11.3% were on cotrimazole (septrin) prophylaxis and an equal percentage reported not being on any treat- ment. 1.0% had stopped taking HAART due to perceived increase in sickness. Otherwise non Kaposi's sarcoma cases were managed by a combination of surgery followed by chemotherapy and/or radiotherapy (**Table 5**). Specific drugs used, dosages and number of courses were determined by a consultant oncologist whereas those who underwent radiotherapy were handled by consultant radiotherapy oncologists. One of the patients with chondrosarcoma of the mandible had total mandibulectomy followed by radiotherapy but died after

Table 2. Distribution of sarcomas according to age groups.

Neoplasm	Age (years)							Mean
	0 - 9	10 - 19	20 - 29	30 - 39	40 - 49	50 - 59	60+	
Kaposi's sarcoma	7	9	58	58	28	5	1	30.9 ± 10.0
Rhabdomyosarcoma	2	2	2	1	1	-	1	26.6 ± 21.9
Osteosarcoma	1	-	1	2	2	-	-	32.3 ± 13.7
Embryonal rhabdomyosarcoma	-	3	2	-	-	-	-	8.0 ± 6.4
Chondrosarcoma	-	-	2	2	1	-	-	30.8 ± 7.7
Hemagio pericytoma	1	-	-	1	1	1	-	32.5 ± 19.1
Malignant fibrous hystiocytoma	-	-	-	1	1	-	-	38.5 ± 2.1
Myxoid liposarcoma	-	1	-	-	-	-	-	27*
Fibro sarcoma	-	-	-	-	-	-	6	62*
Dermato fibrosarcoma	-	1	-	-	-	-	-	14*
Reticular cell sarcoma	-	-	1	-	-	-	-	23*
Total	11	16	66	65	34	6	8	

*Denotes age of the lone patient in the group; #2 Patients did not have their age specified.

Table 3. Distribution of sarcomas as per affected anatomical site.

Neoplasm	Site (%)									Total %
	Palate	Whole body	Tongue	Mandible	Maxilla	Oral cavity	Parotid	lip	Retro molar area	Row total
Vascular										
Kaposi's sarcoma	22.6	14.3	13.1	2.3	1.2	19.1	0.6	1.8	0.6	75.6
Hemagio pericytoma	-	-	-	-	-	-	25.0	50.0	-	70.0
Muscular										
Rhabdomyosarcoma	-	-	-	44.4	11.1	33.3	-	-	-	100
Embryonal rhabdomyosarcoma	-	-	-	60.0	40.0	-	-	-	-	100
Bony										
Osteosarcoma	-	-	-	83.3	16.4	-	-	-	-	100
Cartilagenous										
Chondrosarcoma	-	-	-	60.0	40.0	-	-	-	-	100
Fibrous										
Malignant fibrous hystiocytoma	100	-	-	-	-	-	-	-	-	100
Fibro sarcoma	-	-	-	-	-	100	-	100	-	100
Dermato fibrosarcoma	-	-	-	-	-	-	-	-	-	100
Fatty										
Myxoid liposarcoma	-	-	-	-	-	100	-	-	-	100
Unclear Histiogenesis										
Reticular cell sarcoma	-	-	-	-	-	-	100	-	-	100

The missing row percentages are for those cases whose site was not established.

Table 4. Distribution of sarcomas as per chief complaint/reason for coming to hospital.

Neoplasm	Chief complaint of patient N)									Duration (weeks)
	Swelling	Referral	Pain+ swelling	Bleeding	Pain	Difficult eating	Swelling+ bleeding	Pain+ Numbness	Wound	
Kaposi's sarcoma	30	7	37	5	3	5	2	1	2	1 - 156
Rhabdomyosarcoma	1	-	2	-	-	-	-	-	-	1 - 24
Osteosarcoma	1	-	3	-	-	-	-	1	-	6 - 22
Embryonal rhabdomyosarcoma	1	-	-	-	-	-	-	-	-	4*
Chondrosarcoma	1	-	3	-	-	-	-	-	-	3 - 60
Hemagio pericytoma	-	-	1	-	-	-	-	-	-	3*
Malignant fibrous hystiocytoma	-	-	1	-	-	-	-	-	-	7*
Myxoid liposarcoma	1	-	-	-	-	-	-	-	-	8*
Fibro sarcoma	-	-	1	-	-	-	-	-	-	250
Dermato fibrosarcoma	1	-	-	-	-	-	-	-	-	4
Reticular cell sarcoma	-	-	-	-	-	-	-	-	-	1*
Total	36	7	48	5	3	5	2	2	2	1 - 205

*Data available for one case or only one case was found.

Table 5. Distribution of sarcomas as per treatment modality.

Neoplasm	Treatment (% of cases)						
	Chemo and or HAART	Surg	Radio	Surg Radio	Surg+Chemo	Surg Chemo Radio	Not established
Kaposi's sarcoma	73.3	-	-	-	0.6	-	26.2
Rhabdomyosarcoma	-	-	-	55.6	-	-	44.4
Osteosarcoma	16.7	-	-	16.7	50.1	16.7	-
Embryonal rhabdomyosarcoma	20.0	-	-	-	-	-	80.0
Chondrosarcoma	-	-	-	-	20.0	80.0	-
Hemagio pericytoma	50.0	50.0	-	-	-	-	-
Malignant fibrous hystiocytoma	50.0	-	-	-	-	-	50.0
Myxoid liposarcoma	100	-	-	-	-	-	-
Fibro sarcoma	100	-	-	-	-	-	-
Dermato fibrosarcoma	100	-	-	-	-	-	-
Reticular cell sarcoma	-	-	-	-	-	-	100

Those under not established are cases whose files couldn't be traced.

the second dose of chemotherapy. We were only able to follow a few patients due to distances involved.

4. DISCUSSION

Little is known of the epidemiology of oral maxillofacial sarcomas in a Ugandan population. The data analyzed here differed a bit with the general epidemiology of the disease probably due to the high number of Kaposi's sarcoma cases (**Table 1**) seen at our centre [1,3,7,8]. Although a study from Kenya [8] reported Kaposi's sarcoma as the commonest sarcoma, their percentage was not as high as that seen in this study (P 0.001 chi 72.19). On the other hand, a Nigerian [7] study reported only one case therefore either other centers have most of the biopsies for Kaposi's sarcoma diagnosis taken by other specialties or the oral facial component of the disease is the main manifestation among our population. This has been alluded to by Ziegler *et al.* [14]. All Kaposi's sarcoma patients were HIV positive with up to 17.7% being on HAART at the time of diagnosis. This was not a surprise given the fact that increased incidence of HIV/AIDS associated Kaposis sarcoma has been reported by other authors [15]. We could not ascertain the exact treatment that our Kaposi's sarcoma patients got due to an ongoing research randomizing them in HAART group and other arms. However, the main stay of treatment in the country is HAART except in a few cases that are given chemotherapy as well. It has become standard practice in this era of HAART to expect a resolution of the lesions with improving immunity and increase in CD4 cell count [16,17].

Rhabdomyosarcoma was the second commonest sarcoma in our series followed by osteosarcoma and then chondrosarcoma. The data in this study is slightly different in that respect when compared to other reports that rank rhabdomyosarcoma second to fourth in prevalence [1,7,8,18,19]. Chondrosarcoma is reported to be rare in the head and neck region [1,7,18] therefore the prevalence in this study was rather high but not statistically significantly different from that reported by a Kenyan study that picked just 3 cases over a ten year period (P = 0.12 chi 2.38). A Nigerian [7] report had only 2 cases of chondrosarcoma that were loco-regional failures from other centre in a twenty year study. It is hard to tell if the prevalence in Uganda is higher or it's because our unit is the only functional maxillofacial unit in the country at present. Unfortunately the risk factors for sarcomas have not been well addressed and it was not any different in this study therefore we could not establish if there are any particular factors to explain this high occurrence.

The majority of our patients presented with swelling and pain as the chief complaints. This differed with reports from developed country [18,20]. Sarcomas are known to present as painless swelling therefore pain on set is either because of advancement or supra infection. In this study painful swellings were the commonest chief complaints. This probably was due to delayed reporting and supra infection. In fact one of the chondrosarcoma patients decided to try traditional healers until pain set in before she came back for surgery.

In this study surgical intervention for non Kaposi's sarcoma disease was the primary treatment modality just as reported by other authors [1,7,8,18]. Surgery was accompanied by some form of adjuvant therapy with either radiation and/or chemotherapy. However, the few whose outcome was established, the results were discouraging as many died within a short time.

Future research should focus on survival of patients and the factors that affect treatment outcomes. A mechanism for long term follow up has to be implemented. With increasing mobile telephone penetration, a window of opportunity is available to aid us in follow up.

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