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Maxillary bone sarcoma rare disease, NCI experience

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Abstract:

Background: Head and neck sarcoma represent about less than 10% of all body sarcoma, maxillary bone sarcoma is not a common variant among cases of head and neck sarcoma usually occur in 3rd to 4th decades of life, osteosarcoma is the most common variant among maxillary bone sarcomas, surgery isthe standard treatment line of maxillary bone sarcoma.

Patients and methods: A retrospective study was conducted among 20 patients with maxillary bone sarcoma in NCI, Cairo University, during the period from 2009 to 2018 to present the experience of National cancer institute, Cairo University, in the evaluation and management of maxillary bone sarcoma.

Results: among 20 cases of maxillary bone sarcoma; 11 patients (55%) were below 30 years old, 9 patients (45%) were above 30 years old. Out of our 20 patients, there were 13 males (65%) and 7 females (35%). Osteosarcoma was diagnosed in 12 patients (60%), Chondrosarcoma was diagnosed in 6 patients (30%) while Ewing's sarcoma was diagnosed in 2 patients (10%). On uni-variant analysis tumor size and adjuvant chemotherapy intake have statistically significant effect on overall survival. On Multi variant analysis, tumor size was the only predictor that had significant effect on overall survival. On uni-variant analysis tumor size had significant effect on disease free survival. On multi variant analysis tumor size only that affected the disease free survival with statistical significance.

Conclusion: maxillary bone sarcoma is rare tumor occurs in 3rd to 4th decades of life with male predominance, osteosarcoma was the most common variant, most of them were of high grade, surgery was the standard line of treatment with good results regarding disease free survival in case of achieving negative margins (R0 resection).

Introduction:

Sarcoma is a malignant tumor arising from cells of mesenchymal lineage and accounting for 1% of all malignant tumors, head and neck sarcoma represent 4:10% among all body sarcoma.(1) Surgery with negative margins (R0 resection) is the gold standard choice for treatment of sarcoma; unfortunately this can't be achieved every time in head and neck sarcoma due to tumor invading areas of difficult surgical access.(2) In such cases; radiotherapy is usually given as an adjuvant therapy and this decreases the risk for local recurrence. (3) Head and neck sarcomas are either bone sarcoma (20% of cases) or soft tissue sarcoma (80% of cases).(4) Maxillary osteosarcoma is the most common variant among maxillary bone sarcoma.(5) Chondrosarcoma represents 1:5% among maxillary bone sarcoma cases.(6) Ewing sarcoma represents 1:4% among maxillary bone sarcoma cases. (7) Unlike extremity osteosarcomas, which occur in younger patients maxillary bone osteosarcomas usually present in the third and fourth decades. (8)

Methodology:

Medical records of 20 patients with maxillary bone sarcoma were reviewed and analyzed in the period between 2009 and 2018; patients included in this study were patients presented with maxillary bone sarcoma which was pathologically proved, patients excluded from this study were patients presented with cutaneous sarcoma (Kaposi sarcoma) and patients with maxillary sinus sarcoma. Primary endpoint of this study is to present the experience of National cancer institute, Cairo University, in the evaluation and management of maxillary bone sarcoma, to evaluate incidence, presentation, imaging characteristics, pathological features and treatment modalities of maxillary bone sarcoma. Secondary endpoint of this study is to evaluate the outcome of management in maxillary bone sarcoma patients regarding loco-regional control, overall survival and disease free survival. Tertiary endpoint of this study is to identify role of adjuvant therapy in dealing with maxillary bone sarcomas.

Results:

Among the reported 20 cases with maxillary bone sarcoma, 13 cases were males (65%) and 7 cases were females (35%), 11 patients (55%) were below 30 years old while 9 patients (45%) were above 30 years old, among our patients, no one has family history of malignancy, with only 2 patients (10%) had a history of self-malignancy (retinoblastoma). 14 cases (70%) were diagnosed via CT imaging while both CT and MRI were used in diagnosis of 6 patients (30%). Incisional biopsy was done in 6 patients (30%), punch biopsy was done in 6 patients (30%) and true cut biopsy was done in 8 patients (40%). The tumor was bilateral in 2 patients (10%), left sided in 11 patients (55%) and right sided in 7 patients (35%). Tumor was less than 8 cm in size in 16 patients (80%) and was more than 8 cm in size in 4 patients (20%). Clinically and radiologically; 3 patients (15%) were diagnosed as stage 1 while 17 patients (85%) were diagnosed as stage 2. Among all of the 20 cases no one was metastatic at initial presentation. Among all of our cases, tumor was high grade with negative cervical lymphadenopathy (N0). Regarding histopathological type, Osteosarcoma was diagnosed in 12 patients (60%), Chondrosarcoma was diagnosed in 6 patients (30%) while Ewing's sarcoma was diagnosed in 2 patients (10%). 18 patients received neo adjuvant chemotherapy, while 2 patients didn't receive neo adjuvant chemotherapy. 18 patients (90%) underwent surgical resection, while 2 patients (10%) were beyond resection and received palliative treatment (chemotherapy & radiotherapy). Out of 18 patients who underwent surgical procedure (maxillectomy); 2 patients (11.1%) underwent partial maxillectomy, 11 patients (61.1%) underwent total maxillectomy while 5 patients (27.8%) underwent extended radical maxillectomy. Reconstruction was done via free flap in 2 patients (11.1%), obturator in 11 patients (61.1%) and obturator with temporalis flap in 5 patients (27.8%). Regarding surgical margins; margins of resection were negative in 5 patients,

while were positive in 13 patients. Regarding adjuvant therapy; adjuvant CTH and RTH were given to 13 patients (whose tumor was resected with positive margins) while adjuvant CTH only was given to 5 patients. Regarding recurrence among 18 patients who underwent surgical resection tumor recurrence was reported in 12 patients; 4 local recurrence only, 6 local recurrence with lung metastases and 2 patients had lung metastases without local recurrence. Among those 12 cases who had tumor recurrence; treatment of recurrence achieved through Chemotherapy and surgery in 6 patients (50%) and palliative Chemotherapy in 6 patients (50%). 12 patients (60%) have 2 years disease free survival, 8 patients died within one year of disease **(Table 1).**

Characteristic		**Frequency**	**Percentage**
Age (years)	**Less than 30 years**	**11**	**55%**
	More than 30 years	**9**	**45%**
Gender	**Male**	**13**	**65%**
	Female	**7**	**35%**
Family history	**No**	**20**	**100%**
History of self-previous malignancy	**Yes**	**2**	**10%**
	No	**18**	**90%**
Laterality	**Bilateral**	**2**	**10%**
	Left	**11**	**55%**
	Right	**7**	**35%**
Pre-operative biopsy	**Incisional**	**6**	**30%**
	Punch	**6**	**30%**
	True-cut	**8**	**40%**
Clinical and radiological stage	**Stage 1**	**3**	**15%**
	Stage 2	**17**	**85%**
Distant Metastases at initial presentation	**No**	**20**	**100%**
Histopathological type	**Chondrosarcoma**	**6**	**30%**
	Osteosarcoma	**12**	**60%**
	Ewing's sarcoma	**2**	**10%**
Grade	**High**	**20**	**100%**
L.Ns	**No**	**20**	**100%**
Margins	**Negative**	**5**	**27.7% among resected cases**
	Positive	**13**	**72.3% among resected cases**
Neo-adjuvant CTH	**Yes**	**18**	**90%**
	No	**2**	**10%**

Surgical procedure	**Yes**	**18**	**90%**
	No	**2**	**10%**
Resection type of maxillectomy	**Partial**	**2**	**11.1% among resected cases**
	Total	**11**	**61.1% among resected cases**
	Extended radical	**5**	**27.8% among resected cases**
Reconstruction type	**Free flap**	**2**	**11.1% among resected cases**
	Obturator	**11**	**61.1% among resected cases**
	Temporalis flap and obturator	**5**	**27.8% among resected cases**
Adjuvant RTH+CTH	**Yes**	**13**	**65%**
	No	**7**	**35%**
Adjuvant CTH only	**Yes**	**5**	**25%**
	No	**15**	**75%**
Recurrence	**Yes**	**12**	**60%**
	No	**8**	**40%**
Site of the recurrence	**Local only**	**4**	**20%**
	Local and lung metastases	**6**	**30%**
	Lung metastases only	**2**	**10%**
Final status	**Alive**	**12**	**60%**
	Dead	**8**	**40%**

Table 1 : Clinic-pathological features of studied series.

Regarding overall survival analysis and within a median follow up of 36 months ranging from (11:84 months) we found that both tumor size and adjuvant chemotherapy had significant effect on overall survival on the uni-variant statistical analysis; 1 year overall survival reported in 92.3% of the patients received adjuvant CTH, compared to 66.7% of the patients who didn't receive adjuvant CTH, 3 years overall survival occurred in 83.9% of the patients received adjuvant CTH, compared to 33.3% of the patients who didn't receive adjuvant CTH, 73.4% of patients with tumor size below 8 cm had overall survival of 3 years, compared to only 50% of patients with tumor size more than 8 cm. On multi-variant analysis we found that tumor size was the only predictor that has statistically significant effect on overall survival. **(Fig., 1).**

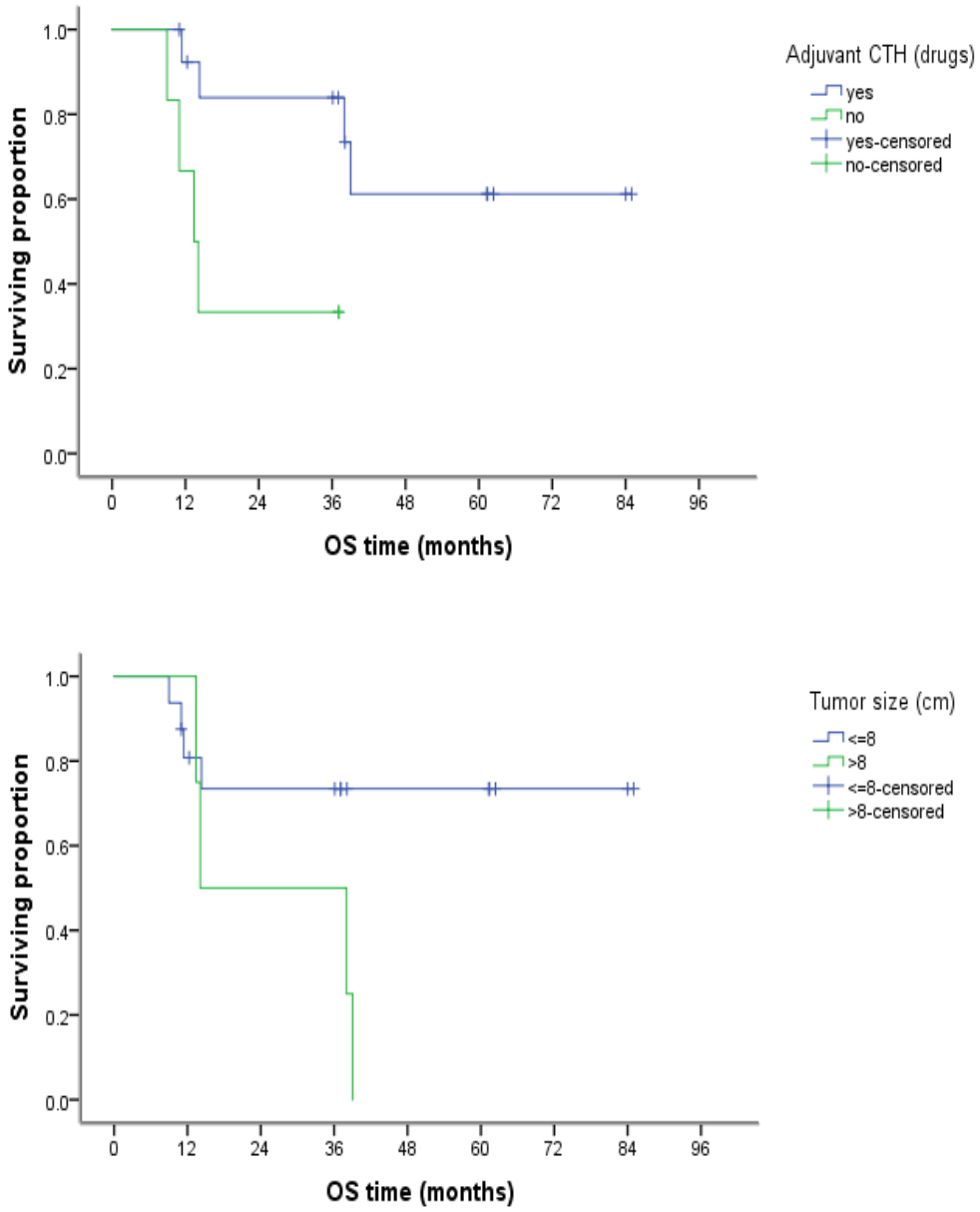

Figure2 : Effect of tumor size and adjuvant CTH on overall survival.

Regarding disease free survival analysis and within a median follow up of 36 months ranging from (11:84 months) we found that tumor size had the only statistically significant effect on disease free survival on both uni-variant analysis and multi-variant analysis; median disease free survival for patients with tumor size below 8 cm disease was 37 months compared to 4 months in patients with tumor size more than 8 cm **(Fig., 2).**

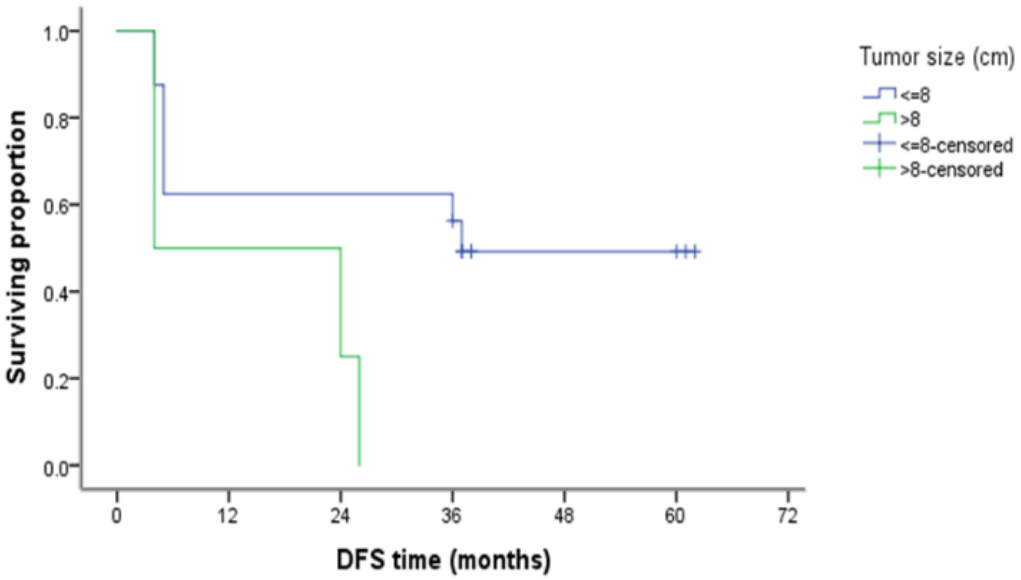

Figure2: Effect of tumor size on disease free survival.

Disscussion:

Head and neck sarcoma is a rare tumor, it accounts for less than 10% of all sarcomas. (9) Yamaguchi S et al among 32 cases of maxillary sarcoma reported that osteosarcoma was the most common variant. (10) Barosa J et al that analyzed 29 cases of head and neck sarcomas reported that osteosarcoma was detected in (34%) of patients and chondrosarcoma was detected in (6.8%) of patients. (4) Among our study we reported that most of the included bone sarcoma was osteosarcoma (60%), followed by chondrosarcoma (40%), and Ewing sarcoma (10%). Regarding age,

among 16 cases of osteogenic sarcoma of the jaw; Fernandes R et al reported that median age of the patients was 41 years. (11) Among our series we reported that median age of our patients was 30 years; 11 patients (55%) were below 30 years old, 9 patients (45%) were above 30 years old. Regarding gender distribution, Ogunlewe M et al. among 59 patients with bone sarcomas of the jaws reported male predominance. (12) Among our cases we reported male predominance as there were 13 males (65%) and 7 females (35%). Computed tomography (CT) is considered the most common diagnostic imaging used for diagnosis of maxillary bone sarcoma. (13) We used CT imaging in 14 cases (70%) while both CT and MRI were used in diagnosis of 6 patients (30%). Luna-Ortiz K et al reported in study including 21 patients with maxillary bone sarcoma that tumor was less than 8 cm in 13 cases (62%), and more than 8 cm in 8 cases (38%). (13) Among our cases tumor was less than 8 cm in size in 16 patients (80%), tumor was more than 8 cm in size in 4 patients (20%). Among our series, all of our patients were high grade and this was going with Luna-Ortiz K et al and in Fernandes R et al in which the largest proportion of patients had high grade tumors. (11&13) Among our series' most of our cases underwent surgical resection and this was reported also by Ferrari D et al. (14) Regarding disease free survival; among 12 cases of maxillary osteosarcoma Jasnau S et al. reported that 79% of patients showed 2 years disease free survival (15), we reported that 60% had 2 years disease free survival. Among a lot of literatures; tumor size is considered an important predictor that has statistically significant effect on overall survival. Among 541 patients of head and neck sarcoma Lee R et al. reported on multi-variant analysis that tumor size had significance on disease free survival. (16) Albergo et al. and Bosma et al. reported thatamong cases of jaw Ewing sarcoma tumor size less than 8cm had low risk for local recurrence. (17&18) Wang et al. among cases of jaw chondrosarcoma reported that there was low risk for local recurrence for cases with tumor size less than 10cm. (19) Patel SG et al. reported that tumor size more than 4cm and positive margins of excision were associated with local recurrence. (20) Among our cases we documented on multi-variant analysis that tumor size with cutoff point 8cm was the only significant predictor for disease free survival. Smeele L et al. and Ogunlewe M et al. reported that adjuvant chemotherapy had significant effect on overall survival among patients with maxillary bone sarcoma. (21&12) Patel SG et al. reported that adjuvant chemotherapy had no significant in both overall survival and disease free survival, also reported that negative surgical margins were the most important predictor of overall and disease-specific survival. (20) Among our study we reported that adjuvant chemotherapy had significant effect on overall survival on the uni-variant statistical analysis. We also reported that importance of achieving negative margins in surgical resection and its impact on low risk for local recurrence.

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