Pediatric Malignancies of the Facial Skeleton

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Abstract

Objectives: To analyze the demographics, survival, and treatment efficacy of pediatric tumors of the facial skeleton.

Study Design and Methods: Retrospective study of cases from the US National Cancer Institute's Surveillance, Epidemiology, and End Results database. Pediatric patients between the ages of 0 and 18 who were diagnosed with a malignant sarcoma of either the bones of skull and face and associated joints or mandible from 1973 to 2012 were identified.

Results: In total, 192 patients were included in the analysis. The average age of diagnosis was 11.21 (±5.19) with a male-to-female ratio of 1.49:1. Whites were the most commonly affected race (76.56%). Malignant mandible sarcomas account for 29.69% of the cohort (n=57). The most common pathology was Osteosarcoma, which accounted for 43.23% of the cohort (n=83). Among patients with known histologic grade (n=67), 43.3% were AJCC stage III or IV. Overall, 5-year disease specific survival was 80.62%. When stratified by treatment modality, 5-year DSS was 86.7% for surgery alone, 69.3% for radiation alone, and 74.0% for surgery with adjuvant radiotherapy (p=0.0345).

Conclusions: This study represents the largest cohort of pediatric malignant tumors of the facial skeleton and skull. The most common and most effective treatment is surgery alone, which showed significant improvement in 5-year DSS.

Introduction

- In the pediatric population, the majority of lesions of the jaw are benign.[1]
- The most common primary pediatric bone tumors are sarcomas.
- Osteosarcoma and Ewing's sarcoma accounting for the most prevalent histologies.[2,3]
- Sarcomas are rare in the head and neck.
- They comprise approximately 1% to 2% of all head and neck malignancies.[4]
- The mandible and maxilla are the most commonly involved bones.[3]
- Sarcomas are characterized by destruction of local structures and high rates of recurrence.[5]
- Head and neck sarcomas tend to present with pain, swelling, ocular symptoms, and loose teeth.[3]
- Treatment is surgical resection.
- The primary indicator of survival is the presence of negative margins.[2,3,6]
- Adjuvant radiotherapy has been shown to improve survival in osteosarcoma.[3]

Methods and Materials

- The Surveillance, Epidemiology, and End Results (SEER) 18 database was queried using the following criteria: Pediatric patients (Age at diagnosis 0-18), Sarcoma, Bones of the face, skull, mandible, and joints, 1973-2012.
- Patients were sorted into cohorts based upon: Primary site and Treatment type.
- Data was analyzed on the basis of: Age, Sex, Race, Histologic grade, Histologic type, and Treatment modality.
- Survival analysis was performed using disease-specific survival (DSS) calculations.
- Hazard analysis was performed using Cox regression.
- $P < 0.05$ was considered statistically significant for all tests.

Results

Figure 1: Population by Sarcoma Sub-Type.

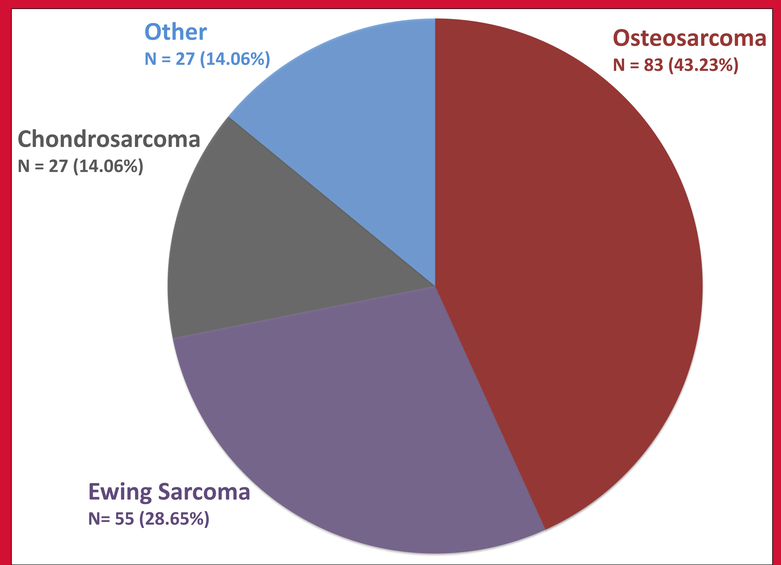

Table 1: Demographic and Clinicopathologic characteristics of Pediatric Head and Neck Sarcoma (1973-2012)

	N	%
Total	192	100.00%
Mean age at diagnosis, years	11.21±5.19	
Sex		
Male	115	59.90%
Female	77	40.10%
Race		
White	147	76.56%
Black	24	12.50%
Other	19	9.90%
Unknown	2	1.04%
Primary Site		
Mandible	57	29.69%
Bones of Skull, Face, and Joints	135	70.31%
Treatment		
Surgery	111	57.81%
Radiotherapy	23	11.98%
Multimodality	58	30.21%

Table 2: Disease-specific survival analysis of Pediatric Head and Neck Sarcoma by Site

	n	Survival	p-value (log-rank)
All Sites			**0.0345***
Overall Survival	192	80.62%	
Multimodality Therapy	58	74.01%	
Surgery Alone	111	86.66%	
Radiation Alone	23	69.29%	
Bones of Face, Skull, Joints			0.1209
Overall Survival	135	80.66%	
Multimodality Therapy	49	75.47%	
Surgery Alone	69	86.98%	
Radiation Alone	17	70.59%	
Mandible			0.2180
Overall Survival	57	80.41%	
Multimodality Therapy	9	66.67%	
Surgery Alone	42	85.95%	
Radiation Alone	6	66.67%	

Figure 2: Kaplan-Meier Survival Curves of Pedatric Head and Neck Sarcoma by Site

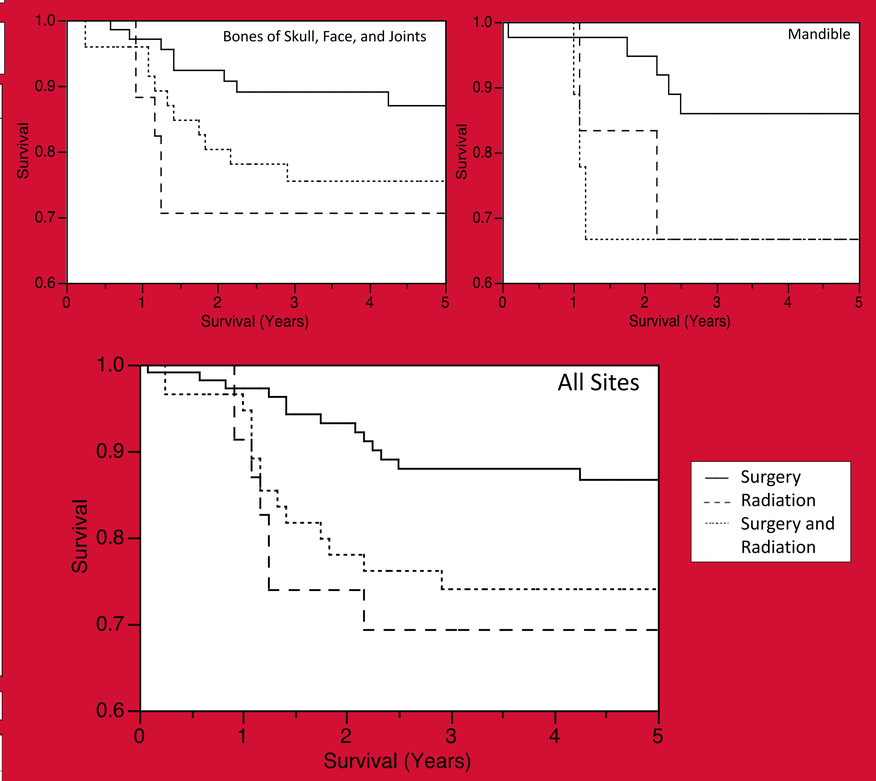

Table 3: Cox Proportional Hazard Analysis of Pediatric Head and Neck Sarcoma by Site

Treatment Modality	Hazards Ratio	95% CI	p-value
Overall			
Single Modality Therapy	ref.		
Multimodality Therapy	1.72	0.85 - 3.38	0.1289
Surgery Alone	ref.		
Radiation Alone	2.76	1.04 - 6.74	**0.0428***
Face, Skull, and Joints			
Single Modality Therapy	ref.		
Multimodality Therapy	1.58	0.70 - 3.54	0.2664
Surgery Alone	ref.		
Radiation Alone	2.78	0.84 - 8.34	0.0901
Mandible			
Single Modality Therapy	ref.		
Multimodality Therapy	2.43	0.52 - 8.76	0.2294
Surgery Alone	ref.		
Radiation Alone	2.69	0.39 - 12.50	0.2754

Discussion and Conclusions

- The most common primary pediatric bone tumors of the head and neck are sarcomas.
- A majority of these sarcomas are osteosarcomas (43.23%)
- The overall survival of these patients is 80.62% with similar survival seen in bones of the face and skull as well as the mandible.
- Surgery alone had a higher overall 5 year survival with 86.66% compared to radiation alone which had a survival of 69.29%.
- Multimodality therapy yielded an average overall 5 year survival of 74.01%
- Cox Proportional Hazard confirmed our survival analysis showing that radiation had a greater hazard of death than surgery alone.
- Chemotherapy use is an important treatment modality in this population but was unable to be analyzed due to the limitations associated with the SEER database.
- Surgery should be strongly considered in the treatment management of pediatric sarcomas of the head and neck as the primary mode of therapy when possible with radiation given adjuvantly when necessary.

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