A Study on 13 Cases of Bone and Soft Tissue Sarcomas in the Head and Neck

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Since bone and soft tissue sarcomas arising from the head and neck region are very rare, the standard treatment protocol has yet to be established. In the present study, 13 cases with bone and soft tissue sarcomas treated from 2007 to 2014 were analyzed. The mean period from initial visit to final pathological diagnosis was approximately 3 weeks, and 38.5% of the cases needed several biopsies for an accurate diagnosis. Death occurred early for those cases in which more than 40 days was needed to obtain the histological diagnosis. The 3-year overall survival in 13 cases was 35.2%. As the cause of death was uncontrolled local lesion, the locoregional control was more important than distant metastasis control for the prognosis. Tumor location, TNM stage, and histological grade affected the prognosis of the sarcomas. Radiation therapy including heavy ion and proton beam radiotherapy was not so effective as the primary treatment, as was the case with salvage treatment of sarcomas in the head and neck. Although the primary treatment of sarcomas was surgical treatment with an adequate margin, an efficient treatment protocol should be established to obtain a better prognosis.

Keywords : bone and soft tissue sarcomas, head and neck, radiation therapy, prognostic factor, FNCLCC classification

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Primary site and pathology of the 13 cases
Sinonasal sarcoma accounted for 46% of the cases.

Pathology		Site					
		Face	Neck	Oral/oropharynx	Sinonasal cavity	Larynx	Middle ear
Rhabdomyosarcoma	3		1 (Submandibular region)	1 (Tongue)	1 (Sinus)		
Undifferentiated pleomorphic sarcoma	2			1 (Soft palate)		1	
Osteosarcoma	2				2 (Sinus)		
Leiomyosarcoma	2				2 (Sinus, Nasal cavity)		
Synovial sarcoma	1				1 (Nasal cavity)		
Angiosarcoma	1						1
Unclassified pleomorphic sarcoma	2	1 (Buccal region)		1 (Oral floor)			
Total	13	1	1	3	6 (46%)	1	1

Summary of 13 cases in relation to the interval from the first visit to the diseases diagnosis, number of biopsies needed to obtain the pathological diagnosis, tumor size, presence/absence of lymph node metastasis, AJCC stage classification, and FNCLCC classification

More than 40 days were required before a pathological diagnosis could be obtained in the three cases. Tumor size of less than 5 cm are two cases. Three cases are disease-free survival.

Case	Sex	Age	Site	Pathology	Pathological diagnosis period	Count biopsy	Tumor diameter Major axis × minor axis	Lymph node metastasis	AJCC Stage				FNCLCC Classification			
									T	N	M	Stage	Tumor differentiation	Mitosis count	Tumor necrosis	Grade
1	F	55	Sinus	Rhabdomyosarcoma	20	1	5.3×2.2	+	T2b	1	0	IV	3	3	0	3
2	M	9	Submandibular region	Rhabdomyosarcoma	21	1	5.7×4.5	–	T2b	0	0	III	3	3	0	3
3	M	9	Tongue	Rhabdomyosarcoma	22	1	5.1×3.2	–	T2b	0	0	III	3	3	2	3
4	F	49	Soft palate	Undifferentiated pleomorphic sarcoma	165	2	5.1×4.9	–	T2b	0	0	III	3	1	0	2
5	M	77	Larynx	Undifferentiated pleomorphic sarcoma	23	1	1.0×1.0	–	T1a	0	0	I	2	1	0	1
6	M	22	Maxillary sinus	Osteosarcoma	27	1	7.6×5.5	–	T2b	0	0	III	3	1	0	2
7	M	34	Maxillary sinus	Osteosarcoma	22	2	6.0×3.1	–	T2b	0	0	III	2	1	2	2
8	M	56	Maxillary sinus	Leiomyosarcoma	22	1	6.4×5.0	–	T2b	0	0	I	2	1	0	1
9	F	59	Nasal cavity	Leiomyosarcoma	22	1	3.6×1.9	–	T1b	0	0	I	2	1	0	1
10	M	57	Nasal cavity	Synovial sarcoma	90	2	7.6×3.9	–	T2b	0	0	III	3	1	1	2
11	M	72	Middle ear	Angiosarcoma	41	2	5.2×4.0	–	T2b	0	0	I	2	1	0	1
12	M	38	Buccal region	Unclassified pleomorphic sarcoma	33	2	5.3×4.0	–	T2b	0	0	III	3	1	0	2
13	M	45	Oral floor	Unclassified pleomorphic sarcoma	24	1	8.5×5.0	–	T2b	0	0	III	3	1	1	2

Treatment and prognosis of the 13 cases

Only one case received palliative treatment, while the remaining 12 cases received curative treatment. Three of the four cases in whom surgery was performed first remain alive. All of the cases who received chemotherapy prior to surgery or radiotherapy died. In 10 of the cases in which surgery was performed, the surgical margins were negative, however, 6 of these cases (60%) died of local recurrence.

Case	Site	Pathology	Curative treatment	Surgical margin	Course of treatment	Prognosis (Month)
1	Sinus	Rhabdomyosarcoma	Chemo > Proton beam 72.6Gy	—	Local recurrence > Chemo > Distant metastasis > BSC	Died of the primary cancer (22)
2	Submandibular region	Rhabdomyosarcoma	Chemo+Bone marrow transplantation > Ope (ND)	Negative	Local recurrence > RT 54Gy > Chemo > BSC	Died of the primary cancer (28)
3	Tongue	Rhabdomyosarcoma	Chemo+Bone marrow transplantation > Ope (Partial exision of tongue)	Negative	Local recurrence > Chemo+Proton beam 54Gy > Brainstem recurrence > Palliative RT > BSC	Died of the primary cancer (36)
4	Soft palate	Undifferentiated pleomorphic sarcoma	Ope (Resection+ND+Rec)	Negative	Local recurrence > BSC	Died of the primary cancer (7)
5	Larynx	Undifferentiated pleomorphic sarcoma	Ope (Subtotal resection of Larynx)	Negative	Observation (No adjuvant therapy)	Alive without disease (48)
6	Maxillary sinus	Osteosarcoma	Chemo > Ope (Subtotal resection of Maxillary sinus+*ND+Rec) > Chemo	Negative	Local recurrence > Chemo > BSC	Died of the primary cancer (24)
7	Maxillary sinus	Osteosarcoma	Chemo+Proton beam (Primary: 77Gy, Lymph node: 64Gy)	—	Tumor remaining+Distant metastasis > BSC	Died of the primary cancer (12)
8	Maxillary sinus	Reiomyosarcoma	Chemo > Ope (Total resection of Maxillary sinus+ND+ Rec) > Chemo (VAC)	Negative	Local recurrence > Ope+RT > Distant metastasis > BSC	Died of the primary cancer (18)
9	Nasal	Reiomyosarcoma	Ope (Subtotal resection of Maxillary sinus) > Chemo (VAC)+RT (66.6Gy)	Negative	Chemo (VAC)	Alive without disease (21)
10	Nasal	Synovial sarcoma	Chemo > Ope (Resection)	Negative	Local recurrence > Heavy ion 64GyE > Local recurrence+Distant metastasis > Chemo > BSC	Died of the primary cancer (14)
11	Middle ear	Angiosarcoma	Palliative RT 40Gy	—	BSC	Died of the primary cancer (12)
12	Buccal region	Unclassified pleomorphic sarcoma	Chemo > Ope (Resection)	Negative	Local recurrence > Chemo+Ope+RT > Local recurrence > BSC	Died of the primary cancer (43)
13	Oral Floor	Unclassified pleomorphic sarcoma	Ope (Resection+Rec) > Chemo (VAC)	Negative	Local recurrence > Ope (Mandibular partial resection+Rec)	Alive without disease (50)

BSC: Best supportive care; Chemo: Chemotherapy, Gy: Gray; ND: Neck Dissection; Ope: Operation; RT: Radiation, VAC: Vincristine/ActinomycinD/Cyclophosphamide; Rec: Reconstruction