



Sarcoma in Head and Neck: A Study of 25 Cases in Regional Cancer Centre

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Abstract

Soft tissue sarcomas are rare tumours that account for less than 1% of all malignancies. Histopathologically diagnosed cases at Regional Cancer Centre (RCC) BCCI Guwahati between July 2010 and December 2016 were analyzed retrospectively. A total of 25 cases were seen during the study period. The mandible, tongue, orbit, scalp and maxilla were the commonest sites affected. None of the patients had neck nodes or distant metastasis at presentation. 18 patients were treated with surgical resection primarily and this was followed by adjuvant treatment. The majority of the patients failed within first and second year. Grade of tumour and T stage of the tumor were found to have a significant effect on prognosis.

Keywords: *Head and neck, sarcoma, lung metastasis, radiotherapy, chemotherapy, VAC.*

Introduction

Soft tissue sarcomas of the head and neck are mesenchymal malignant neoplasms¹. They account for less than 10% of all soft tissue sarcomas and comprises 1% of all head and neck neoplasms². Of soft tissues sarcomas, 10–20% are seen in children and 80–90% affect adults³. Soft tissue sarcomas (STS) have tri-modal age distribution with peaks in less than 10 years of age, between 11 and 40 years and the last is over 40 years. Most head and neck sarcomas occur with localized disease. Overall regional metastases is found to occur in 10-15% of head

and neck sarcomas; in most of these cases they arise from high-grade primary lesions. Distant metastases are rare in the absence of regional metastases at presentation, and if the nodal metastases is present one should search for distant metastases. Lung is the most common site of distant metastases, followed by the liver and bone. They are associated with significant morbidity and mortality. There are several histological subtypes and they present with a variety of clinical characteristics and many often require treatment with combination of surgery, radiotherapy and chemotherapy. Considering the

rarity of the disease, there is currently a dearth of studies in the literature, so we report our experience of the management of head and neck soft tissue sarcomas.

Material and Methods

Type of study: Retrospective

Number of cases: 25 patients

Study period: July 2010 to December 2016

Inclusion criteria: Patients in whom diagnosis was confirmed by a pathology report.

Patients who has not taken any treatment outside our institute

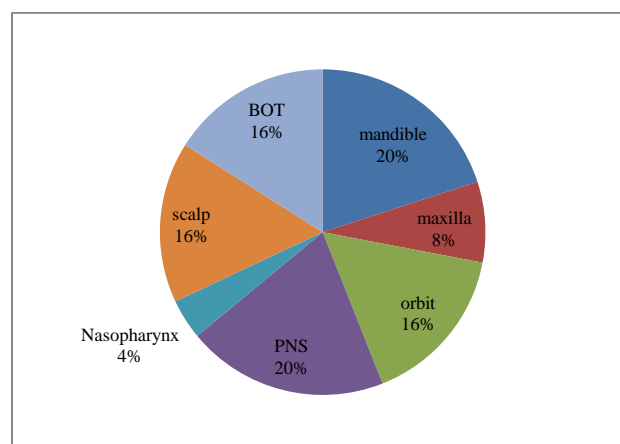
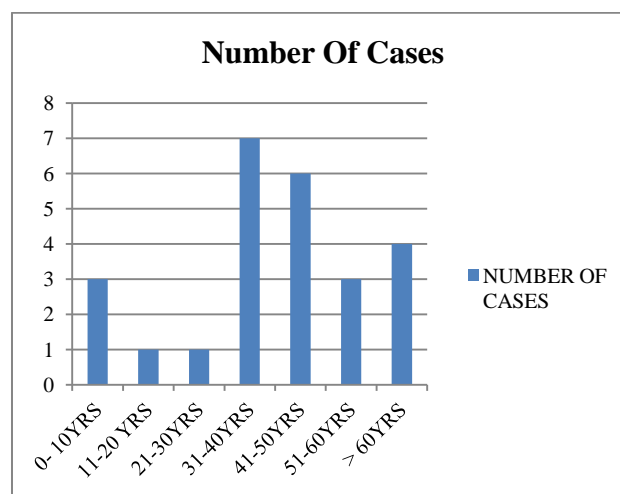
Results and Observations

Out of 25 patients, 16 were males & 9 females (male:female = 1.7:1) & 4 were of paediatric age (16%). 16 cases underwent surgery , elective neck dissection was not performed in view of the low rate of regional nodal metastases in sarcomas. 14 patients received post-op adjuvant radiotherapy they were either high grade tumor (11 cases) or had positive surgical margin (3 cases) . Patients with mandibular, maxillary and paranasal sinus sarcoma were treated with composite resection and necessary reconstruction as a definitive surgical procedure. 2 patients who did not receive radiation had tumors well below 5 cm in size with clear resection margins. 2 carcinosarcoma and 2 spindle cell sarcoma in BOT(Base of tongue) ,4 orbital sarcoma and 1 nasopharyngeal sarcoma were treated with combined modality chemotherapy and radiation. Chemotherapy was administered on individualized basis Vincristine, Adriamycin, Cyclophosphamide (VAC) regime . Ifosfamide and itopside based therapy was used for other high-risk tumors. 10 patients developed metastasis at follow up. All of them developed lung metastasis ; 7 of them have died . Out of 10 patients who developed metastasis 5 were of high grade, 3 of intermediate grade and 2 were of low grade tumor tumors . Size of the primary tumor also has an important prognostic role, as patients developing metastasis 6 were having size of primary tumor > 5 cm. This shows that grade of

tumor and size of the primary tumor are important prognostic factors.

AGE	NUMBER OF CASES
0-10 Yrs	3
11-20yrs	1
21-30yrs	1
31-40yrs	7
41-50yrs	6
51-60yrs	3
60yrs above	4

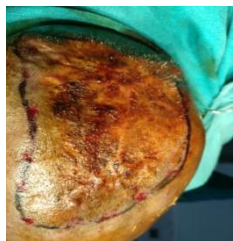
Histology	Mandible	Maxilla	Orbit	PNS	nasopharynx	Scalp	BOT
Rhabdomyosarcoma	3	1		2			
Angiosarcoma				1			
Ewing's sarcoma	2	1	2	2	1		
Carcinosarcoma							2
Spindle cell sarcoma			2			1	2
Fibrosarcoma						3	



T stage	Metastasis
T1	4
T2	6

Size	Low grade	Intermediate grade	High grade
<5cm (T1)	7	3	7
>5cm (T2)	1	3	4

Grade of Tumor	Metastasis
High	5
Intermediate	3
Low	2



Dermato fibrosarcoma scalp



Ewing's sarcoma mandible



Ewing's sarcoma mandible intraoperative pic

Discussion

Head and neck sarcomas are rare and there is lack of studies about their management and outcome. Early detection and diagnosis is clearly essential. There is high chance of local recurrence in high grade sarcomas as high as 50% in the literature^{4,5,6}. Barker et al in their study reported the median time of local recurrence after treatment with surgery and/or radiotherapy to be 4 months.⁷ The risk of local recurrence was higher with positive surgical margins as shown by other authors. Regional metastasis of head and neck sarcomas is almost exclusively in high-grade sarcomas. Lung metastasis is very common and also it is the commonest cause of death, this is

similar to our study. It has been suggested by Mendenhall et al¹ that patients should undergo a chest CT before treatment and also suggested that in the absence of pulmonary metastases, other distant metastases are highly unlikely. Most patients had lesions of the mandible, which along with the PNS, is the most common site of primary tumors^{7,8}. A large retrospective series from MD Anderson, however, indicated that high grade tumors, radiation induced tumors, and involved margins were associated with a poorer disease free survival and overall survival at five years⁸. High local failure rates in the head and neck have historically been associated with poorer treatment outcomes in this group. Since this is a rare disease, it is difficult to establish a standard treatment for head and neck sarcomas⁹. The optimal treatment is complete resection¹⁰. Because of the small space of the head and neck region and its proximity to vital structures, surgical removal of head and neck sarcomas many a times cannot achieve the ideal "wide" resection margins which are preferred in other anatomic sites. The various other important considerations include the morbidity related to breathing, swallowing, speaking, and cosmetic deformity that may follow facial and neck resection. Treatment usually includes multiple treatment modes, especially when it is not possible to totally resect the tumor¹¹; nonetheless, the best treatment option is total tumor resection. In our study, most of the patients were treated with surgery + radiotherapy with or without chemotherapy.

Conclusion

Head and neck soft tissue sarcoma is an aggressive tumor, having a very heterogeneous group. Prognosis is poor in overall survival. The recommended treatment is mainly surgical. The treatment plan should aim at resection with negative margins however aesthetic and functional concern should be taken care of too, which are due to close anatomy of the head and neck region.

Radiation therapy has an important role in case of negative factors such as higher grade or positive resection margins.

The role of chemotherapy needs further evaluation. There is a great need of further studies both prospective and retrospective to assess the treatment protocol, role of chemotherapy and need of genetic and molecular study to improve the survival of the patients.

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