Original Article - Retrospective Study

Head and Neck Sarcomas-clinicopathological Findings, Treatment Modalities and Its Outcome - A Retrospective Study

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Abstract

Introduction: Sarcomas are malignant neoplasms constituting an important group of tumours due to their high morbidity and mortality. They can arise either from soft tissue or bone. Soft tissue sarcomas are common, but bone sarcomas are rare. The aims and objective of this study are to carry out the study of head-and-neck sarcomas between January 2006 and May 2020. **Materials and Methods:** Eighty-two patients of head-and-neck sarcoma were included. Study of demographic and clinicopathological characteristics including age, sex, gender, presenting symptoms, origin, primary anatomical location, size, tumour grade, staging, treatment modality, recurrence, metastasis, and patient status during the recent visit. **Results:** Fifty-six (68.3%) were males and 26 (31.7%) were females. The male/female ratio was 2.28:1. The most affected age group was 0–19 years. Tumour size of >5 cm in greatest dimension was seen in 67.1% cases and tumour size of <5 cm in 32.9%. Chemotherapy + radiotherapy (CT + RT) was given in 23.8% followed by surgery in 13.9%. Recurrence was seen in 19.5% and metastasis in 18.3%. **Discussion:** Rhabdomyosarcoma was the most predominant histological type. Maximum patients reported in late stage. The most commonly used treatment was the combination of CT and RT. Recurrence was seen in 19.5% and distant metastasis in 18.3% patients. Head-and-neck sarcomas are rare so epidemiological studies involving more cases must be carried out for better understanding and better outcome.

Keywords: Chemotherapy, head and neck, radiotherapy, sarcoma, surgery

Introduction

Sarcoma is a rare malignant tumour arising from mesodermal tissues, which forms the connective tissue of the body. They are an important group of tumours due to their high morbidity and mortality. Head-and-neck sarcomas constitute only 1% of all head and neck and 5% of all sarcoma cases.[1] Only 5%-15% of sarcomas affect the head-and-neck region in adults and about 35% affect paediatric patients.[2,3] Almost 80% of sarcomas arise from soft tissues, whereas 20% sarcomas arise from hard tissues.[4] Eighty to ninety percent soft tissue sarcomas are seen in adults and 10%-20% are seen in children.[4] The male/female ratio is 1.42:1.[5] The exact etiology of origin of sarcomas is still unknown. However, idiopathic, genetically predisposing factors, exposure to radiation, certain viral diseases, and chemical carcinogens are attributed to be responsible for genesis of the majority of sarcomas.[6]

Sarcomas have a large spectrum of clinical features, varying from slow growth to aggressive local and regional growth with systemic metastases. The most common symptom of soft tissue sarcoma (STS) of the head and neck is a painless swelling (80%-90%) and pain could be present occasionally, but pain is the most common presenting symptom in bone sarcomas.[7] Visual disturbance, sinusitis, epistaxis, otalgia, motor, and/or sensory disturbances may be the other symptoms.[2]

Sarcomas are classified according to types of tissue from which they arise and more than 50 histological subtypes have been

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described.[1,5] Rhabdomyosarcomas (RMS) are the most common histological type in the head-and-neck region followed by malignant histiocytomas, fibrosarcomas and neurofibrosarcomas.[8]

Imaging plays very important role in diagnosis and assessment of the head-and-neck sarcomas. Computed tomography (CT) scans and Magnetic Resonance Imaging (MRI) are two most common radiological investigations. Positron-emission tomography using fluorodeoxyglucose is a technique that assesses glucose uptake by the tumour.[6]

Treatment depends tumour size, site, histological type, stage, and age of patient. Surgery, chemotherapy (CT) and radiotherapy (RT) are commonly used treatment modalities.[9] Neck dissection is indicated when positive lymph node is identified.[10] RT for head-and-neck sarcoma is indicated in high-grade tumours, positive surgical margins, tumours >5 cm, and recurrent lesions.[11]

Head-and-neck sarcomas have a poor prognosis due to anatomic constraints and its proximity to vital structures, increasing the chances of recurrence.[12] The 5-year survival rate for sarcomas of head and neck is between 49%-55%.[11]

Materials and Methods

The study was conducted on sarcomas of the head-and-neck region between 2006 and 2020. The head and neck were defined as any site above the clavicles. The other anatomical sites were classified as follows: gnathic bone (mandible, maxilla), oral cavity (tongue, palate, lip, buccal mucosa, retromolar trigone), orbit, sinonasal tract (maxillary sinus, nasopharynx, nasal cavity), multiple sites (tumour involving more than one site), neck, face, temporal bone, masseter muscle, parotid gland, scalp, and pyriform fossa. Informed written consent was obtained from patients participating in the study. All procedures performed in the study were conducted in accordance with the ethics standards given in 1964 Declaration of Helsinki, as revised in 2013. Ethical clearance for the study was obtained from the Institutional Review Board of King George's Medical University (ECM II B/P 21) on 07/04/2020.

Settings and design

The patients were recruited from the pool of patients coming in different departments of the university. It was a retrospective, descriptive single-center study.

Confounders and variables

Following variables: age, sex, gender, presenting symptoms, origin, primary anatomical location of the neoplasm, size, tumour grade, staging, treatment modality, recurrence, and metastasis were studied.

Inclusion criteria

All patients of head and neck sarcomas were included in the study.

Exclusion criteria

Subjects with other systemic disease, pregnancy or lactation, history of drug abuse, currently on steroid therapy, and allergic to any medication were excluded from the study.

Staging

Staging was done according to Memorial Sloan– Kettering system and International Union against Cancer (UICC) staging system.

Grading

Grading was done on the basis of differentiation, mitotic count, and necrosis.

Treatment

Surgery, RT, CT, or combination of these was done according to indications. RT was given in the case of positive surgical margins not amenable to further surgical re-excision and/or high-grade tumours and tumour of >5 cm size or recurrent disease. The delivered dose was 60–62 Grays (Gy) in the case of negative margins and at least 64–66 Gy in the case of positive or close margins or gross residual disease, in fractions of 2 Gy each.

Assessment

Assessment was done after surgery, RT, CT, and combination of these. Anatomic site, tumour size, grade, treatment, recurrence and metastasis, and death were also assessed.

Statistics

The collected data were analysed with IBM. SPSS statistics software 24.0. (Chicago, Illinois, USA). Student's-*t*-test and Man–Whitney test were used for parametric and nonparametric continuous data. For categorical data, Chi-square testing was used. A $P < 0.05$ was considered statistically significant. Survival was estimated using Kaplan–Meier survival curves. Log-rank method was used to analyze the influence of various prognostic factors on survival of the patients [Figure 1].

Results

Eighty-two patients with head-and-neck sarcomas were included; 74.4% were STS and 25.6% were bone sarcomas. The mean follow-up was 32.6 ± 29.3 (mean ± standard deviation [SD]) months, median interquartile range (IQR): 24 months (12–44). Fifty-seven were males and 25 were females. The male/female ratio was 2.28:1. The most affected age group was 0–19 years, representing 64.6%, followed by 20–60 years 31.7% and least affected age group was >60 years representing 3.7%, the mean ± SD: 21.6 ± 19.4, median (IQR): 14 (6.7–34.2) [Table 1].

Gnathic bones were involved in most of the 46.3% cases, followed by oral cavity 19.5%, orbit and sinonasal tract (SNT) 7.3%, multiple sites 4.9%, neck 3.7%, face, temporal bone and masseter muscle 2.4%, parotid, scalp and pyriform fossa 1.2%. We found that 12 histopathological subtypes, the most common type was the RMS seen in 39% cases, followed by Ewing's sarcoma 14.6%, malignant fibrous histiocytoma (MFH), and osteosarcoma 10.9%. MFH had 3 variants (pleomorphic sarcoma, dermatofibrosarcoma protuberans, and myxofibrosarcoma). Synovial sarcoma and carcinosarcoma 7.3%, chondrosarcoma and fibrosarcoma 2.4%, leiomyosarcoma, epithelioid sarcoma, myeloid sarcoma

Table 1: Sample characteristic

Variable	*n*=82, *n* (%)
Age (years)	
Mean±SD	21.6±19.4
Median (IQR)	14 (6-38.0)
Age range (years)	
0-19	53 (64.6)
20-60	26 (31.7)
>60	3 (3.7)
Gender	
Male	56 (68.3)
Female	26 (31.7)
Tumour size	
<5 centimeter 55	67.1
>5 centimeter 27	33
Tumour grade	
Low grade	14 (17.1)
Intermediate grade	11 (13.4)
High grade	57 (69.5)
Follow up period (months)	
Up to 24	50 (60.9)
25-48	17 (20.7)
49-72	6 (7.3)
>72	9 (10.9)
Mean±SD	32.8±30.1)
Median (IQR)	24.0 (12-45)
Stage	
Stage I	1 (1.2)
Stage II	24 (29.3)
Stage III	40 (48.8)
Stage IV	17 (20.7)
Treatment	
CT + RT	30 (36.6)
Surgery	17 (20.7)
Surgery + RT + CT	16 (19.3)
Surgery + RT	6 (7.3)
Surgery + CT	6 (7.3)
CT	7 (8.5)
Outcome	
Died	34 (41.4)
Alive with disease	19 (23.1)
Alive without disease	29 (35.4)

SD=Standard deviation; IQR=Interquartile range; CT=Chemotherapy; RT=Radiotherapy

and malignant peripheral nerve sheath tumour (MPNST) 1.2% [Table 2].

Tumour size of >5 cm in greatest dimension was seen in 67.1% cases and <5 cm in 32.9%. High-grade tumours were observed in 69.5% patients, low grade in 17.1%, and intermediate grade in 13.4%. In Stage I, only 1.2% reported, 29.3% in Stage II, 48.8% in Stage III, and 20.7% in Stage IV, respectively.

Surgery was done in 20.7% patients. Surgery in combination with RT was given in 7.3%, surgery + CT in 7.3%, surgery + CT + RT in 19.5%, CT + RT in 36.6%. CT alone was given in 8.5% patients.

Recurrence was seen in 19.5% patients. Multiple recurrences were seen in a case of MFH of maxilla. Recurrence within a week was seen in a case of synovial sarcoma of the parotid gland. Metastasis was seen in 18.3%. Distant metastasis was seen in lung 12.2%, multiple sites 4.9%, and pelvic bone 1.2%.

Most of the patients (58.5%) were alive in the last visit at 5 years. Twenty nine (35.4%) were alive without disease (AWTD) and 23.2% were alive with disease (AWD). Thirty four (41.5%) died of disease. The mean (± SD): follow-up was 38.2 ± 30.1 months and median (IQR): follow-up 24.0 (12–45) months.

The highest number of deaths were seen due to RMS and MFH representing 9.8%, followed by synovial sarcoma and carcinosarcoma 6.1%, Ewing's sarcoma 4.9%, fibrosarcoma 2.4%, osteosarcoma and MPNST 1.2%. Anatomical site responsible for maximum number of deaths were gnathic bones 18.3%, followed by oral cavity 7.6%, multiple site 6.1%, neck 3.6%, orbit 2.4% and SNT, scalp, masseter muscle 1.2%.

Histological type responsible for highest number of AWTD patients was RMS 17.1%, followed by Ewing's sarcoma 7.3%, osteosarcoma 6.1%, chondrosarcoma 2.4%, MFH 1.2%, synovial sarcoma 1.2%. Histological type responsible for highest number of AWD patients was again RMS 12.2%, osteosarcoma 3.6%, Ewing's sarcoma 2.4%, and carcinosarcoma, myeloid sarcoma, epithelioid sarcoma and leiomyosarcoma 1.2%.

Anatomical site responsible for highest number AWTD was gnathic bone 18.3%, orbit and SNT 4.9%, multiple site 3.6%, face, parotid and masseter muscle 1.2% cases. Site responsible for highest number of AWD was gnathic bone and oral cavity 8.5%, temporal bone 2.4% and pyriform fossa, SNT and face in 1.2% cases [Table 3] and Kaplan–Meier survival curves.

Discussion

Sarcomas are important group of rare malignant tumours having high rate of morbidity, mortality, and diversity. In our study, there was a higher prevalence in men which is consistent with previous studies.[10,12-15] However, Bree *et al*. found a higher frequency among females.[1] The male/female ratio found in our study was (2.29:1), almost similar to other studies which found ratios of 2:1 and1.9:1, respectively.[14,16] However, lower male/female ratios of 1.76:1 and 1.42:1 and higher female-to-male (18:17) ratio has also been reported.[3,5,17]

STS of head and neck presents as a painless mass in 80% of the cases. Pain is the most common presenting symptom in bone sarcomas.[7] We observed majority of the STS had painless swelling but bone sarcomas (Ewing's sarcoma and osteosarcoma) had rapid growth and pain since initial visit. However, sarcomas of paranasal sinus may present symptoms of sinusitis, headache, diplopia, dystopia, dysphagia, trismus, respiratory obstruction, and dysphonia.[18-22]

Table 2: Location of tumour relation to sites

Histological type	Gnathic bone	Oral cavity	Orbit	SNT	Multiple sites	Neck	Face	Temporal bone	Masseter muscle	Parotid	Scalp	Pyriform fossa	Total
RMS	8	9	5	4	2	0	1	2	1	0	0	0	32
Ewing's sarcoma	9	0	1	0	0	0	1	0	0	1	0	0	12
MFH	5	2	0	0	1	0	0	0	0	0	1	0	9
Osteosarcoma	9	0	0	0	0	0	0	0	0	0	0	0	9
Synovial sarcoma	2	0	0	0	1	2	0	0	1	0	0	0	6
Carcinosarcoma	2	4	0	0	0	0	0	0	0	0	0	0	6
Chondrosarcoma	2	0	0	0	0	0	0	0	0	0	0	0	2
Fibrosarcoma	0	0	0	1	0	1	0	0	0	0	0	0	2
Leiomyosarcoma	0	0	0	0	0	0	0	0	0	0	0	1	1
Epithelioid sarcoma	0	1	0	0	0	0	0	0	0	0	0	0	1
Myeloid sarcoma	0	0	0	1	0	0	0	0	0	0	0	0	1
MPNST	1	0	0	0	0	0	0	0	0	0	0	0	1
Total	38	16	6	6	4	3	2	2	2	1	1	1	82

RMS=Rhabdomyosarcoma; MFH=Malignant fibrous histiocytoma; MPNST=Malignant peripheral nerve sheath tumours; SNT=Sinonasal tract; CT=Chemotherapy; RT=Radiotherapy

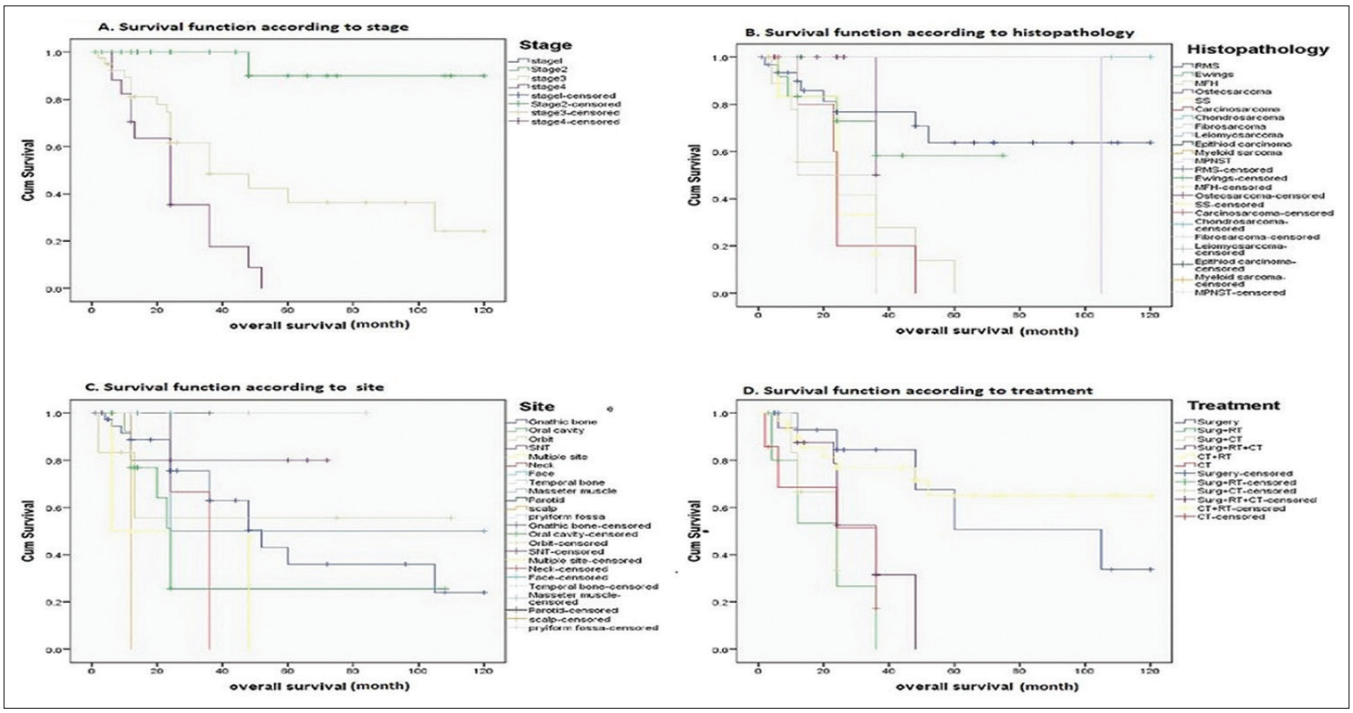

Figure 1: A log-rank test was run to determine if there was difference in survival distribution for different types of treatment, different stages of disease, primary site, and histopathological types of the tumour. The survival distribution for the different treatments (Chi-square [df = 5] =21.6, $P = 0.001$); stage of disease (Chi-square [df = 3] =25.9, $P < 0.001$); histopathological type (Chi-square [df = 11] =27.1, $P = 0.004$) was significantly different. However, the survival distribution for different primary sites of tumour was not significant (Chi-square [df = 11] =18.2, $P = 0.08$)

The World Health Organization has described more than 50–80 subtypes of sarcomas.[5,23-26] We found 12 histopathological types of sarcomas. In the head-and-neck region, the most frequent histological types were RMS, followed by MFH, fibrosarcomas, and neurofibrosarcomas.[8] In other study, most common type was MFH followed by dermatofibrosarcoma protuberans (DFSP) and fibrosarcoma.[9] MPNST was most common subtype (25%) followed by Ewing's sarcoma, liposarcoma, synovial sarcoma, and DFSP,[14] while undifferentiated pleomorphic sarcoma was most common followed by chondroblastic osteosarcoma.[17,27,28] Fifty percent of sarcomas represent osteosarcoma, RMS, MFH, fibrosarcoma, and angiosarcoma.[4] The most common subtype in our study was the RMS, (39%) which was higher than other studies where incidence was 25%.[3,13] Other studies showed lower incidence of 8%, 16.2%, and 1%–4.5%.[15,23,29]

RMS had the highest number of deaths with incidence of 9.8%.[3] RMS has a historically poor prognosis, however, progresses seen in therapy has improved substantially the clinical outcome

Table 3: Outcome status till last visit according to histopathological type, location, and outcome

	Alive with disease (%)	Alive without disease (%)	Died (%)
HPE type			
RMS	10 (12.2)	14 (17.1)	8 (9.8)
Ewing's sarcoma	2 (2.4)	6 (7.3)	4 (4.9)
MFH	0	1 (1.2)	8 (9.8)
Osteosarcoma	3 (3.7)	5 (6.1)	1 (1.2)
Synovial sarcoma	0	1 (1.2)	5 (6.1)
Carcinosarcoma	1 (1.2)	0	5 (6.1)
Chondrosarcoma	0	2 (2.4)	0
Fibrosarcoma	0	0	2 (2.4)
Leiomyosarcoma	1 (1.2)	0	0
Epithelioid sarcoma	1 (1.2)	0	0
Myeloid sarcoma	1 (1.2)	0	0
MPNST			1 (1.2)
Total	19 (23.1)	29 (35.6)	34 (41.4)
Location			
Gnathic bone	7 (8.5)	15 (18.3)	16 (19.5)
Oral cavity	7 (8.5)	2 (2.4)	7 (8.5)
Orbit	0	4 (4.9)	2 (2.4)
Sinonasal tract	1 (1.2)	4 (4.9)	1 (1.2)
Multiple sites	0	1 (1.2)	3 (3.7)
Neck	0	0	3 (3.6)
Face	1 (1.2)	1 (1.2)	0
Temporal bone	2 (2.4)	0	0
Masseter muscle	0	1 (1.2)	1 (1.2)
Parotid	0	1 (1.2)	0
Scalp	0	0	1 (1.2)
Pyriform fossa	1 (1.2)	0	0
Total	19 (23.1)	29 (35.3)	34 (41.4)
Treatment			
Surgery	3 (3.6)	9 (10.9)	5 (6.1)
Surgery + RT	1 (1.2)	0	4 (4.9)
Surgery + CT	4 (4.9)	0	3 (3.6)
Surgery + RT + CT	2 (2.4)	1 (1.2)	9 (10.9)
CT + RT	5 (6.1)	17 (20.7)	7 (8.5)
CT	4 (4.9)	2 (2.4)	6 (7.3)
Total	19 (23.1)	29 (35.3)	34 (41.4)

RMS=Rhabdomyosarcoma; MFH=Malignant fibrous histiocytoma; MPNST=Malignant peripheral nerve sheath tumours; RT=Radiotherapy; CT=Chemotherapy

of patients.[24] In our study too, RMS remains the histological type which had maximum number of deaths along with MFH constituting 9.8% of the death followed by synovial sarcoma and carcinosarcoma 6.1% each, and fibrosarcoma 2.4%.

Treatment of head-and-neck sarcoma is dependent on histological type, location, stage of disease, size, and age of patient. Combination of CT + RT is the most common treatment used.[9,16,24] CT alone is indicated in the unresectable head-and-neck sarcoma and sarcomas which extend to the unusual locations like skull base and aggressive sarcomas.[11] Surgery remains the mainstay of treatment for sarcomas of the head-and-neck region with only exceptions being most of the RMS and Ewing's sarcomas.[23] Growth of sarcoma is expansive with formation of pseudocapsule with a tendency to grow along fascial planes. Due to this reason, wider excision with adequate margins is required. Unfortunately, it is difficult to obtain wide margins in head-and-neck region due to anatomic constraints.[30,31] However, some authors have used endoscopic surgical approach wherever possible to provides better functional and cosmetic results.[32]

The incidence of lymph node metastasis is very low in head-and-neck sarcomas. Adjuvant RT is given after excision of high-grade sarcomas, large tumour or surgical margins are positive or close to positive.[33-35] Due to complexity of the anatomical sites and adjacent vital structures, it is not possible to get adequate margin after excision in all cases. Resection with postoperative adjuvant therapy is done in most of the cases.[11] Due to rarity and the diverse clinical behaviours, management of sarcoma can be challenging and should be carried out in a multidisciplinary hospital with expertise by experienced surgeons.[36-39]

Surgery was done in 17% patients by us. In most of the studies, surgery was the only treatment option. Surgery was done on 77%,[15] 52.7%,[27] and 22.2%[14] patients. In the head-and-neck region, indications for CT are unresectable sarcomas, sarcomas with extension to the unusual sites, e.g., skull base and sarcomas of aggressive nature.[11] CT and RT were given in 36.6% cases in our study. As in our Indian scenario, patients report to us in late stage of disease due to unawareness and poor socioeconomic conditions making lesions inoperable. Combination of surgery and CT was given in 7.3% cases by us whereas percentage in cases of Singh *et al*.[14] was 4.1%. A combination of surgery and RT was given in 7.3% patients by us. Lajer *et al*.[27] used in 13.8%, Penel *et al*.[13] in 21.4%, Pacheco *et al*.[3] in 27.8% cases. Singh *et al*. did surgery + RT in 36.1% of the cases.[14] 53% patients were treated with radiation therapy without surgery by Andersen *et al*.[39] In our study, combination of surgery, CT, and RT was given in 19.5% patients. Pacheco *et al*.[3] performed combination of surgery + RT + CT in 27.8% cases. Other authors find it to be used less frequently; 14.2% and 16.6%, respectively.[13,27] Surgery + RT + CT was done in 5.5% of the cases by Singh *et al*.[14] CT alone was given by us in 8.5% patients. CT along with RT, especially where wide excision is not possible improves local control.[11]

Sarcomas are known for their higher recurrence rate. Inadequate excision causes recurrence. The local recurrence rates for high-grade STS following surgery have been reported as 50%.[18] Therefore, adequate margins should be taken at the time of the surgery.

Lymph node metastasis is less common as compared to distant metastasis. Lymph node metastasis was observed in 3%–10%, while distant metastasis was seen in 28% of the cases.[11] For distant metastasis, the most frequent site was lung followed by bone, central nervous system, and

liver.[22] Distant metastases were not seen by Tejani *et al*.[23] Patel *et al*. reported nodal metastasis as 3%.[6] Singh *et al*.[14] reported metastasis in 42% of the cases. We observed 18.3% metastasis in our study.

We observed 35.4% patients alive without evidence of disease. It was seen as 41.6%, 45.1%, and 50% by others.[3,11,9] Live patients with local disease had a prevalence of 25% and 21.3%.[3,11] It found 23.2% by us. Overall, the patients who were alive at the time of the last visit in our study were 58.5%. Mean follow-up was 32.8 ± 30.1 months in our study. Overall survival reported by Singh *et al*.[14] was 48.6% in their study.

Death rate of 22.2%, 18.19%, 33.3%, and 33.5% was found, respectively.[3,9,4,11] We observed higher death rate of 41.5% compared to previous studies.

Conclusion

RMS was most common tumour. Most affected age group was 0-19 years age group. Maximum patients reported in Stage III.

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Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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