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ORIGINAL ARTICLE

Sarcomas of the Oral and Maxillofacial Region: Analysis of 26 Cases with Emphasis on Diagnostic Challenges

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Abstract

Sarcomas of the Oral and Maxillofacial Region (SOMR) are rare lesions which pose diagnostic and management challenges. We analyzed 26 cases of SOMR with respect to clinical presentation, histopathological subtype, treatment modalities, recurrence, and treatment outcome. In our series, Osteosarcoma (OS) was the most common type of sarcoma (7 cases), followed by 5 cases of Ewing's Sarcoma (ES), 3 cases each of Chondrosarcoma (CS) and Leiomyosarcoma (LMS), 2 cases each of Malignant Peripheral Nerve Sheath Tumor (MPNST), Pleomorphic Undifferentiated Sarcoma (PUS), Myeloid Sarcoma (MS)and Rhabdomyosarcoma (RMS). Surgery was the primary treatment modality in most cases and was combined with adjuvant chemo/ radiotherapy in few cases. 24 of the 26 cases were followed up for an average period of 40.67 months. Adverse disease outcomes like recurrence were seen in 2 cases whereas death due to the disease was reported in 7 cases. In view of the diagnostic challenges faced in SOMRs, it appears practical to stress on the underlying genetic aspects of the disease process rather than histological subtyping to improve disease outcome.

Keywords Sarcoma · Oral and maxillofacial region · Molecular diagnostics

Introduction

Sarcomas are a heterogeneous group of rare neoplasms that constitute about only 1% of all malignancies. They are even rarer in the oral and maxillofacial region and constitute less than 1% of neoplasms occurring in this region [1].The true incidence of these tumors is relatively difficult to ascertain due to rarity of their occurrence and difficulty in diagnosis and classification. Sarcomas originate from diverse tissues and more than 50 histopathological subtypes have been identified [2].

Sarcomas of the oral and maxillofacial region **(SOMR)** are known to arise more often from soft tissue (80%) than from bone/cartilage (20%) [1].They confer mortality by virtue of local recurrence rather than distant metastasis as is observed in sarcomas arising in sites other than oral and maxillofacial region [3]. Acceptable surgical margins are difficult to obtain in case of SOMR due to delicate anatomy of this region and proximity to vital structures [4].

The diagnosis and classification of sarcomas has been aided by use of immunohistochemistry (IHC) and molecular markers; however as many as 20% of these tumors still remain

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unclassified and are thus difficult to manage [5]. There is a paucity of evidence based data in the literature which is a direct result of the controversies in sarcoma grading and classification, rarity of these lesions and lack of clear guidelines for successful management [6].

Although SOMR are rare tumors, they cause high mortality and morbidity and hence should be familiar to all healthcare professionals enabling early diagnosis and treatment. The aim of this study was to analyze the clinical, histopathological features; treatment outcomes and survival in patients of SOMR seen at our institute over a 10 year period for better understanding of these rare lesions. Commonly encountered diagnostic pitfalls have also been discussed.

Material and Methods

The present study was carried out retrospectively, using archival records of patients diagnosed with SOMR at our institution from 2007 to 2017. As per the institutional guidelines, this study was exempted from ethical clearance.

After careful examination of all records and reassessment of histopathological diagnosis, a total of 26 patients diagnosed with primary SOMR were included in the study.

The clinical records of the included patients were reviewed for demographic details, histopathological diagnosis, treatment modality, recurrence and outcome. For overall survival, period from the time of diagnosis till last follow-up/death was considered. None of these patients presented with a prior history of irradiation/surgery.

Statistical analysis was done using IBM SPSS version 20.0 software. Kaplan-Meier method was used for evaluating survival curves with area under the curve calculated at 95% confidence interval.

Results

A total of 26 patients with SOMR were diagnosed at our institution during the study period. Of these, 19 were male (73.07%) and 7 were female (26.9%). The age of initial presentation tumour varied greatly, ranging from 4 years of age to 75 years. 9 patients (34.61%) were less than 18 at the time of presentation.

In the present case series, the tumors showed an almost equal distribution between maxilla (12 cases) and mandible (11 cases). One case each of maxillary buccal vestibule, mandibular gingiva and buccal mucosa was seen.

Out of the 26 cases of SOMR, only 3 cases involved the soft tissues with remaining 23 presenting within the jaws. Painless swelling was seen in majority of patients ranging in duration from 15 days to 11 months. History of trauma was associated with two cases, one each of osteosarcoma and Ewing's sarcoma. Histopathologically the most common hard tissue sarcoma was osteosarcoma and the most common soft tissue sarcoma was Ewing's sarcoma. The patient details have been summarized in Table 1.

The treatment protocol has been summarized in Table 2. The primary treatment modality in our cases was surgery with adjunctive chemotherapy and radiotherapy in some cases. In cases of myeloid sarcoma only chemotherapy was administered. The average follow-up time was 40.67 months with 2 cases lost to follow-up. Overall survival of patients with SOMR is shown in Fig. 1.The age-specific survival of the pediatric cohort was comparatively better than the adult cohort. (Fig. 2).

Discussion

SOMR pose a great challenge for survey studies due to the rarity of their occurrence and diversity of presentation. Literature shows presence of some survey studies which add to the limited knowledge available on these lesions. Most of these studies investigate patient's profile; treatment and survival [2].We have attempted to address the challenges faced while diagnosing sarcomas in addition to the above.

Due to the prevalence of tobacco habit in the Indian subcontinent, oral squamous cell carcinoma is the most frequently encountered malignancy whereas sarcomas account for less than 1% of malignant head and neck tumors [7]. Out of the total 1222 case of reported malignancies in our department during the study period, only26 (2.1%)were SOMRs.

The average age of occurrence of SOMR is 50–60 years as evidenced by previously published reports. In the present series, the average age was 52 years. This value may be skewed due to inclusion of 2 young children aged 4 and 5 years suffering from Ewing's sarcoma and myeloid sarcoma respectively. A marked male predilection ($n = 19$) similar to published literature was also observed [8].

Clinically SOMRs present with vague features with majority of the patients reporting with a painless swelling of relatively short duration and are otherwise asymptomatic [9].Pain is usually seen in association with sarcomas of bone [10].In the current series only 2 patients presented with painful swelling (one each of Ewing's sarcoma and rhabdmyosarcoma).Other common presenting signs were tooth mobility, tenderness, perforation of cortical plates and ulceration of overlying mucosa.

Radiographic investigations are crucial not only to the diagnosis of SOMRs but also help in tumor staging and treatment planning. However, conventional radiographs are inadequate for this purpose and use of special investigations such as computerized tomography (CT) and magnetic resonance imaging (MRI) is of paramount importance [11].

CT and MRI help in assessing tumor size for staging, determining extent of tumor including intramedullary and

Table 1 Summary of thePatients with Sarcomas of the Oral and Maxillofacial Region (SOMR)

Case No.	Histological Diagnosis	Sex	Age (Yrs)	Site	Treatment S- Surgery C- Chemotherapy R- Radiotherapy	Adjuvant treatment	Recur	Survival (In Months)	Status
1.	Osteosarcoma	M	18	Post. Mandible	S	C	–	90	Alive
2.	Osteosarcoma	M	65	Post. Mandible	S	C	12 months	19	Dead
3.	Osteosarcoma	M	21	Post. Maxilla	S	–	–	38	Alive
4.	Osteosarcoma	M	22	Ant. Maxilla	S	C + R	–	61	Alive
5.	Osteosarcoma	M	60	Post. Maxilla	S	C	–	26	Dead
6.	Osteosarcoma	M	18	Post. Maxilla	S	–	–	12	Dead
7.	Osteosarcoma	F	14	Ant. Maxilla	S	–		11	Alive
8.	Ewing's Sarcoma	M	17	Ant. Maxilla	R	C	–	119	Alive
9.	Ewing's Sarcoma	M	30	Ant. Mandible	S	C	after 60 months	120	Alive
10.	Ewing's Sarcoma	M	18	Post. Mandible	S	C	–	54	Alive
11.	Ewing's Sarcoma	M	4	Post. Mandible	S	C + R	–	33	Alive
12	Ewing's Sarcoma	F	15	Post. Mandible	S	C	–	50	Alive
13.	Chondrosarcoma	M	75	Post. Maxilla	S	–	–	25	Dead
14.	Chondrosarcoma	M	40	Post. Mand.	S	–	–	–	Lost To Follow-Up
15.	Chondrosarcoma, Extraskeletal	M	21	Max Buccal Vestibule	S	R	Within 1 Month	78	Alive
16.	Leiomyosarcoma	F	21	Ant. Maxilla	S	R	3 months	24	Dead
17.	Leiomyosarcoma	F	20	Post. Mandible	S	–	–	12	Alive
	Leiomyosarcoma	M	17	Mand. Gingiva	S	–	–	–	Lost To Follow-Up
18.	Pleomorphic Undifferentiated Sarcoma	M	55	Post. Maxillary Alveolus	C	–	+	8	Dead
19.	Pleomorphic Undifferentiated Sarcoma	M	20	Maxilla (Mid palate)	C	R	–	23	Dead
20.	Rhabdomyosarcoma	M	59	Maxillary Tuberosity	S	C + R	–	59	Alive
21.	Rhabdomyosarcoma	F	23	Buccal Mucosa	C	–	–	13	Alive
22.	Malignant Peripheral Nerve Sheath Tumor	F	38	Ant. Mandible	S	R	–	18	Alive
23.	Malignant Peripheral Nerve Sheath Tumor	F	60	Post. Maxilla	S	C + R		25	Alive
24.	Myeloid Sarcoma	M	28	Post. Mandible	C	–	–	30	Alive
25.	Myeloid Sarcoma	M	5	Post. Mandible	C	–	–	28	Alive

Case # 8,10,11,12(Ref. 26), 13(Ref. 50), 15(Ref. 51), 25 and 26 (Ref. 44) have been published previously as case reports/ series

Table 2 Patient characteristics and treatment

S.no	Characteristics	No.	Percentage
1	Age, Y		
	Mean	30	–
	Range	4–75	–
2	Sex, no. (%)		
	Male	19	73%
	Female	07	27%
3	Location, no. (%)		
	Maxilla	12	46%
	Mandible	12	46%
	Others	02	7.6%
4	Pathologic Types, no. (%)		
	Osteosarcoma	07	26.9%
	Ewing's's Sarcoma	05	19.2%
	Chondrosarcoma	03	11.5%
	Leiomyosarcoma	03	11.5%
	Pleomorphic Undifferentiated Sarcoma	02	7.7%
	Rhabdomyosarcoma	02	7.7%
	MPNST	02	7.7%
	Myeloid Sarcoma	02	7.7%
5	Treatment Modality, no. (%)		
	Only Surgery	07	26.9%
	Surgery + Chemotherapy	06	23%
	Surgery + Chemotherapy + Radiotherapy	04	15.3%
	Surgery + Radiotherapy	03	11.5%
	Chemotherapy + Radiotherapy	02	7.7%
	Only Chemotherapy	04	15.3%

extramedullary involvement;invasion into contiguous structures and tumor calcification [12].

Metastasis to other body parts from the primary tumor may be assessed by the use of PET scans. PET scans are superior to CT or MRI for detecting primary tumors, staging and assessing tumor volume [13].

Detection of subcentimeteric tumors has been made possible by the advent of combined PET–computer tomography (CT) scanners, which provide both functional and structural information.This enables early detection of the disease and reduces false-positive lesions [14].

Most intraosseous sarcomas in the current series presented as ill-defined radiolucent defects. Periosteal bony reaction was seen in few tumors of bony origin. However radiographic findings are not pathognomic and should not be solely relied upon for diagnosis. Radiographic presentation of sarcomas may mimic certain benign lesions such as hemangiomas [15].

The role of cytology in the diagnosis of SOMRs is limited with the standard approach being to categorize tumor as per the predominant cell type i.e. into round cell, spindle cell,

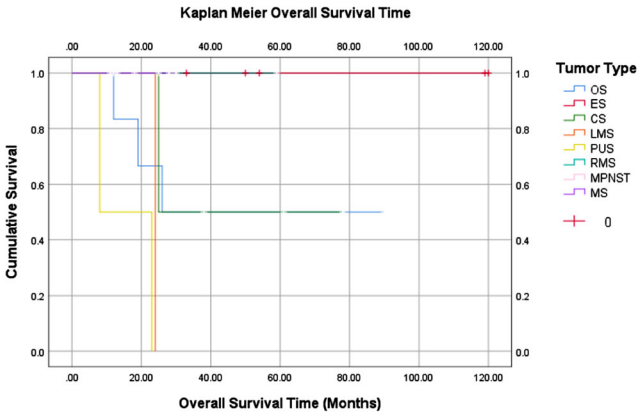

Fig. 1 Kaplan-Meier overall survival curve

pleomorphic and myxoid sarcoma [16]. It is advisable to use FNAC for metastatic and recurrent lesions and as an adjunct to diagnosis.

Histopathological Challenges in Sarcoma Diagnosis

Histopathology is the main stay in sarcoma diagnosis but is frequently problematic with an abundance of diagnostic pitfalls.

Oral lesions that may be misdiagnosed as sarcomas are benign bony lesions like fibrous dysplasia and ossifying fibroma for a low-grade osteosarcoma, lymphomas for round cell tumors and fibromatosis for low-grade fibrosarcomas. Epithelial malignancies like sarcomatoid carcinoma may be misinterpreted as an epitheloid sarcoma in the absence of surface epithelium in a small biopsy sample. Thus conventional histology needs to be supplemented with ancillary methods like immunohistochemistry and molecular diagnostic techniques for accurate diagnosis.

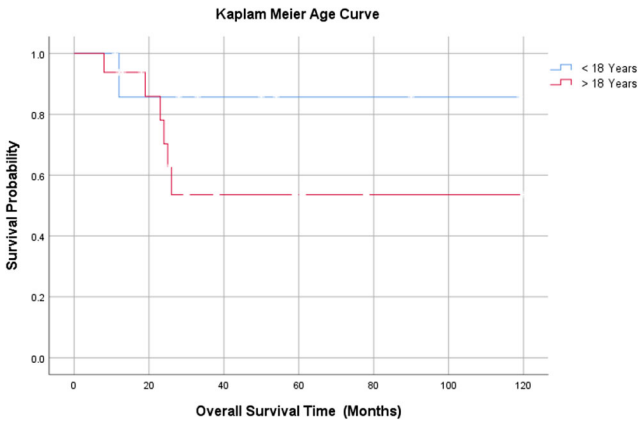

Fig. 2 Kaplan-Meier Age curve

Osteosarcoma (OS)

The most commonly observed sarcoma in our series was osteosarcoma accounting for (23%) of cases. Jaw osteosarcomas are particularly challenging in nature as anatomic complexity of this region render them amenable to local recurrence after surgical resection.

Histologically, OS is characterized by unequivocal production of osteoid by malignant mesenchymal cells. Osteosarcoma of jaws show low mitotic activity and metastasize rarely making differentiation from fibro-osseous lesionsand osteoblastomas difficult. IHC is of limited use; however Yoshida et al. suggested that MDM2 and CDK4 are reliable markers to differentiate low grade osteosarcomas from benign lesions, although discordant results were found in other studies [17, 18].

Chondroblastic and osteoblastic variants are the most common subtypes, with the osteoblastic variant predominating in this series as also in the studies by Argon A et al. and Yildiz FR et al. [19, 20]. Other series have shown an increased prevalence of chondroblastic variant [21],and therefore no absolute consensus can be reached on the more prevalent type. It appears prudent to consider these lesions as histologically heterogeneous with divergent differentiation and need clinical and radiographic corroboration for correct diagnosis.

A diagnostic dilemma also arises when differentiating a chondrosarcoma from chondroblastic variant of osteosarcoma especially considering that management of each tumor is different. IHC marker galectin-1 (positive in osteosarcoma) has been shown to effectively differentiate between the two tumors [22].It has also been suggested to consider malignant chondroid tumors in an adolescent patient as chondroblastic osterosarcoma unless proven otherwise [23].

Chondroblastic variant of osteosarcoma is known to have a better prognosis than the osteoblastic variant. Thus it is postulated that prognosis of jaw osteosarcoma is better in comparison to that of long bones due to prevalence of chondroblastic subtype in jaws [24].

Fibroblastic variant of osteosarcoma can be difficult to diagnose if tumor osteoid is absent as may sometimes be seen in a small biopsy sample. Adding to the diagnostic dilemma in our case (# 7)was diffuse positivity for pan-CK and vimentin. On incisional biopsy, the tumor was signed out as monophasic synovial sarcoma and not spindle cell carcinoma due to the presence of an intact non-dysplastic surface epithelium separated from the tumor by a band of connective tissue. On excisional biopsy, the tumor showed malignant osteoid formation and was negative for EMA.SYT gene rearrangement assay involving 18q11.2 was done using Fluorescence in-situ Hybridization (FISH) and was found negative. A final diagnosis of fibroblastic variant of osteosarcoma was thus given. In a study of 131 osteosarcomas by Okada et al., only 4.5% showed CK positivity, more often than not in epitheloid areas [25]. Thus, though extremely rare, CK positive osteosarcoma is an established entity and the role of ancillary molecular techniques becomes decisive to an accurate diagnosis.

Ewing's's Sarcoma (ES)

ES is a rare malignant round cell tumor believed to be of neuroectodermal origin. Head and neck region accounts for only about 1–4% of all cases of ES. In the 5 cases of ES seen in current series a marked predilection for mandible and male gender was observed. The immunohistochemical diagnostic criteria is already well established, commonly used markers are CD99, FLI1, S-100 and NSE. However CD99 and FLI 1 are also frequently positive in lymphoblastic lymphomas [26]. LCA positivity seen in lymphoblastic lymphomas can help distinguish them from ES [27].

Chondrosarcoma (CS)

1–2%of all Chondrosarcomas occur in head and neck region. Of the three cases reported here one occurred in extraskeletal location and all three were of mesenchymal type histologically. Extraskeletal chondrosarcomas are rare aggressive pathological variants with only one previously reported case of the same arising in the buccal space [28].Diagnostic pitfalls encountered in differentiating chondrosacomas from Chondroblastic variant of osteosarcomas have been addressed in the previous section. The absence of cartilaginous foci makes differentiation of mesenchymal CS from other small round blue cell tumor difficult. Sox 9, a transcription factor [29] believed to be the 'master regulator' of chondrogenesis has been proven to be consistently positive in the cartilaginous as well as primitive mesenchymal cells of CS and may thus enable differentiation from other round cell tumors. However it has also been identified in other tumors of neuroectodermal phenotypes [30]. Thus, routine histology remains the gold standard for CS diagnosis. Mesenchymal CS of the jaw bones has a more indolent course in comparison to those at other anatomic sites. However, extraskeletal mesenchymal CS has a rapid clinical course with frequent metastasis unrelated to patient age or cellular differentiation and a 5 year survival rate of 54.6% [31, 32].

Leiomyosarcoma (LMS)

Although LMS accounts for 7% of all soft tissue sarcomas, it is extremely rare in oral cavity due to the paucity of smooth muscles in this region. The tumor generally does not show any specific signs and symptoms and usually presents as a non-ulcerated painless mass [33].All our cases were seen in young individuals with two arising within jaw bones. Vilos et al. [34] and Yan B et al. have reported that the jaw bones are the most frequent sites for primary LMS of the oral and maxillofacial region with 70% of the lesions arising here. LMS can be differentiated from other spindle cell sarcomas like fibrosarcomas and MPNSTs on the basis of histopathology, special stains and immunohistochemistry. Problems in diagnosis may arise when the tumorshows low mitotic index making differentiation from a benign neoplasm like leiomyoma difficult. Case # 17 in the current series showed low mitotic activity but was still classified as LMS as mitotic activity greater than 1 per10 high power fields in a tumor of smooth muscle origin should be considered malignant [35].Desmin negative LMS needs to be differentiated from myofibrosarcomas. The latter tumor shows diffuse positivity for calponin but only focal expression of h-caldesmon in occasional cases. LMS on the other hand usually expresses both calponinand h-caldesmon [36].

Pleomorphic Undifferentiated Sarcoma (Malignant Fibrous Histyocytoma) (PUS)

It is agreed that the term MFH and PUS may be used synonymously when a combination of sampling and immunohistochemistry show no definable line of difference. Since there are no reproducible criteria for recognition, PUS is essentially a diagnosis of exclusion [37].The degree to which PUS may be sub-classified as a sarcoma of an alternative subtype largely depends on the ancillary techniques that are available to the pathologist. These tumors are notoriously aggressive; however data regarding the prognosis of oral PUS is not well documented. Both cases in the present case series had a poor outcome culminating in death within 2 years of diagnosis.

Rhabdomyosarcoma (RMS)

RMS is the most common soft tissue sarcomas in children and adolescents with about 30% of pediatric RMS presenting in the head and neck region [38]. Adult RMS is a relatively rare tumor that manifests frequently in the extremities and is extremely rare in the head and neck region with a decidedly poorer outcome when compared to pediatric RMS of equivalent site [39].The clinical presentation of adult RMS can be deceptive as was seen in our cases. Case # 20 presented as a soft tissue growth in an extraction socket and was excised on the assumption that it was a peripheral reactive lesion. Case # 21 presented as an extremely tender soft tissue swelling of left side of face giving the impression of a space infection. Due to the absence of any obvious odontogenic infection, an incisional biopsy was performed that revealed RMS. Both cases of adult RMS presented here were treated primarily with surgery and have had an uneventful clinical course for 5 and 1 year respectively. Differentiation from tumors having rhabdoid features such as carcinosarcoma, dedifferentiated chondrosarcoma, MPNST (malignant triton tumor) requires a battery of immunostains in addition to careful evaluation of clinical data [40].

Malignant Peripheral Nerve Sheath Tumor (MPNST)

MPNSTs lack standardized diagnostic criteria and thus pose difficulty in diagnosis. They represent malignant tumors arising from peripheral nerves or displaying differentiation along the lines of elements of nerve sheath. Sporadic cases as opposed to those associated with Neurofibromatosis-1 account for approximately half of reported tumors with slight male predominance [41].Both cases presented here were seen in females and presented within the jaws. The tumors display immunohistochemical variability with only about half showing S-100 positivity and the remaining have fibroblastic, perineural or mixed features to a varying degree. Other markers like LEU-7, Nestin, H3K27me3, PGP9.5, CD56and myelin basic protein are inadequately reliable for diagnosis [42, 43].

Myeloid Sarcoma

MS can be a diagnostic challenge when presenting in the absence of AML, myeloproliferative neoplasm or mixed myelodysplastic/proliferative disorders. Important markers that help differentiate MS from other round cell tumors include CD43, lysozyme, MPO, CD68 and CD34. The differential diagnosis and the cases included here have been discussed in detail in our previous publication [44].

Molecular Diagnostics in Sarcomas

Although historically immunohistochemistry has been an invaluable tool in sarcoma diagnosis, it requires a panel of antibodies to distinguish between the differential diagnoses and even then lacks specificity. Rapid advances have been made in the field of sarcomagenesis due to unmasking of genetic and genomic alterations associated with this group of tumors. This has further led to refinement of morphologic sarcoma classification by providing diagnostic information and the identification and development of novel immunohistochemical markers [45, 46].

Currently, based on distinct and recurrent abnormalities sarcomas have been classified:

A) *Chimeric fusion gene associated tumors*- accounting for approximately 20% of sarcomas with the prototype being Ewing's sarcoma/PNET (EWSR1-FLI1 translocation) and synovial sarcoma (SS18-SSX translocation). Other tumors with distinct chromosomal amplification such as low grade osteosarcomas (12q14–15 amplification) have also been classified along with translocation associated sarcomas. These tumors show simple karyotype with low mutation rates and a relatively monomophic morphology with a wide range of clinical behavior. CIC-rearranged sarcoma (CRS) were until recently described as a subset of the Ewing's sarcoma family. Emerging data suggests that CIC-DUX4 tumors are aggressive soft tissue sarcomas with a distinct pathogenesis. They differ from ES by virtue of their location i.e. somatic soft tissues in contrast to skeletal location of ES. Also, CRS tends to occur at an older age in comparison to pediatric presentation of ES [47].
B) *Oncogenic mutation associated sarcoma*- Gastrointestinal stromal tumor (GIST) represent the prototypic tumor of this type. Also included in this category is GNAS1 mutation seen in fibrous dysplasia that may be useful in distinguishing these tumors from low grade osteosarcomas.
C) *Complex karyotype sarcomas*- This group represents genomically unstable and high grade sarcomas seen predominantly in adults where molecular testing provides limited scope in diagnosis. These tumors show high mutational load with frequent loss of tumor suppressor genes and have no effective targeted therapies. Pleomorphic undifferentiated sarcomas, leiomyosarcomas, pleomorphic rhabdomyosarcomas are some of the tumor which fall under this category [45, 46].

Although molecular and immunohistochemical studies contribute immensely to accurate sarcoma classification, accumulating evidence suggests that from a therapeutic perspective, it is the underlying genomic, genetic and transcription anomalies that matter more than the histological phenotypes [48].

Treatment and Follow Up

Sarcoma therapy is multimodal with surgery being the mainstay of treatment in non-metastatic and chemotherapy the primary treatment modality in metastatic cases. Adjunctive radiotherapy is being increasingly used and immunotherapy is also under trials. For soft tissue sarcomas it is recommended to use all three treatment modalities for achieving best results [49]. For osteosarcomas, the objective of surgery is removal of tumor with at least 3 cms clear margins with preservation of anatomy.

It has been suggested that head and neck sarcomas have a poorer prognosis in terms of recurrence and survival when compared to other sites [6].The effect of disease sub site, however remains unsubstantiated due to the small sample size in most studies. Seven of the twenty six patients in our series died due to the disease; six out of the seven were located in the maxilla which may indicate a poorer prognosis for this subsite. Pleomorphic Undifferentiated Sarcoma being an aggressive tumor resulted in death within 2 years of diagnosis. Recurrence was seen in 4 cases post-treatment with wide variation in time of recurrence, one case of chondrosarcoma (case# 15) showed recurrence within one month after surgery while a case of Ewing's sarcoma (case# 9)in a 30 year old male recurred 60 months after treatment.

Conclusion

SOMRs are a diverse group of tumors requiring multimodal treatment with distinct prognostic implications. The ever evolving field of sarcoma diagnosis has been strengthened by the addition of molecular methods. However, from a therapeutic stand point, it seems more prudent to stress on the genetic alterations underlying the disease process rather than accurate histological classification to improve prognosis. SOMRs are rare lesions and single center studies usually provide small sample size with heterogeneous data. The need for multicenter studies and a cumulative database with cross specialty collaboration is the need of the hour to improve disease outcome. Although our series is small and composed of retrospectively analysed data, we hope that it adds to the existing and established data on this rare group of lesions.

Compliance with Ethical Standards

As per the institutional guidelines, this study was exempted from ethical clearance.

Conflict of Interest The authors declare no conflict of interest.

The Precis After analysis of 26 cases of SOMRs, it is concluded that SOMRs are a diverse group of tumors requiring multimodal treatment and have distinct prognostic implications. From a therapeutic stand point, it is more prudent to stress on the genetic alterations underlying the disease process rather than accurate histological classification to improve prognosis.

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