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Management of Sarcomas of Head and Neck from Oral and Maxillofacial Surgeons Prespective.

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Abstract:
Objectives- *Sarcomas of head and neck (HN) are rare malignant tumors of mesenchymal origin which account 1% of all HN malignancies. The objective was to review the literature to emphasize on classification, aetiology, diagnosis and staging, various treatment modalities and recent updates on HN sarcomas.*
Method- *An electronic data search was done in April 2020 without any time-bound factor on "PubMed," National Library of Medicine, the National Institute of Health, "Science Direct," and "Web of Science" databases. Keywords searched were "head and neck sarcomas" AND "osteosarcoma" AND "Ewing's sarcoma" AND "chondrosarcoma" AND "malignant fibrous histiocytoma" AND "fibrosarcoma" AND "Liposarcoma" AND "rhabdomyosarcoma" AND "angiosarcoma" AND "hemangiopericytoma" AND "synovial sarcoma" AND "malignant schwannoma" AND "leiomyosarcoma" AND "alveolar soft part sarcoma" AND "Kaposi sarcoma" AND "radiation-induced sarcoma". Papers written in only English were included.*
Results- *Abstracts of 6018 articles were reviewed and 80 articles including randomized clinical trials, non-randomized clinical trials, high evidenced observational studies and Systematic reviews were selected.*
Conclusion- *Diagnosis of these tumors depends on physical examination, imaging and biopsy. Computerized tomography showed better evaluation for small tumors and hard tissue sarcomas. Positron Emission Tomography has high sensitivity and specificity for detection of metastasis and recurrent sarcomas. Aggressive surgical resection with adequate margins remains the mainstay treatment except in Ewing's sarcoma, skull base tumors and unresectable cases. Adjuvant chemotherapy/radiotherapy are reserved for high grade tumors and resected tumors with inadequate margins. This literature review highlights the challenges encountered during the diagnosis of sarcomas and the intricacies in the treatment planning.*
Keywords- *'sarcoma', 'head and neck sarcoma', 'soft tissue sarcoma','radiation induced sarcoma', 'osteosarcoma', 'chondrosarcoma'*

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I. Introduction

Sarcoma (derived from the Greek word "*sarx*" meaning fleshy), is an unusual variety of cancer. National Institute of Cancers, have defined sarcomas as "*A type of cancer that begins in bone or the soft tissues of the body, including cartilage, fat, muscle, blood vessels, fibrous tissue, or other connective or supportive tissue*"[1]. Due to the rare incidence of these tumours which is 1% of head and neck (HN) malignancies, there is an inadequate number of published data[2]. The neck region is the commonest site for soft tissue sarcomas (STS). The anatomical heterogenicity of HN region makes surgical therapy a challenging task. Most of the sarcomas of the HN regions are diagnosed late because they essentially are asymptomatic until they involve other neural or vascular structures. Due to the paucity of these malignancies, no systematic protocol or randomized clinical trial focusing on their management.

So, we carried out a literature review to emphasize on classification, aetiology, diagnosis, various treatment modalities, and recent updates on the management of head and neck sarcomas (HNS).

II. Historical Background

Alexis Boyer (1757-1833) was the first to equate exostosis, gumma of bone, spina ventosa, and osteosarcoma. John Abernethy (1764-1831) was the first to classify tumours based on their appearance where he enlisted eight types of sarcomas. Rudolf Virchow, a German physicist, was the one who stated that sarcomas evolve from nonepithelial and non-hematogenous tissues and separated them from other cancers. He distinguished six major types of sarcomas: 1) fibrosarcoma, 2) myxosarcoma, 3) gliosarcoma, 4) melanosarcoma, 5) chondrosarcoma, and 6) osteosarcoma[3].

III. Material methodology

An electronic data search was done in April 2020 without any time-bound factor on the following database "PubMed," National Library of Medicine, the National Institute of Health, "Science Direct", and "Web of Science". Keyword searched were "head and neck sarcomas" AND "osteosarcoma" AND "Ewing's sarcoma" AND "chondrosarcoma" AND "malignant fibrous histiocytoma" AND "fibrosarcoma" AND "Liposarcoma" AND "rhabdomyosarcoma" AND " angiosarcoma" AND "hemangiopericytoma" AND "synovial sarcoma" AND "malignant schwannoma" AND "leiomyosarcoma" AND "alveolar soft part sarcoma" AND "Kaposi sarcoma" AND "radiation-induced sarcoma". Studies reported other than English language were excluded. Abstracts of 6018 articles were reviewed and finally 80 articles including randomized clinical trials, non-randomized clinical trials, high evidenced observational studies and Systematic reviews were selected for the reviews. Studies reported other than English language were excluded.

IV. Aetiology

The precise aetiology of HNS is unknown. Nevertheless, it can be briefly attributed as idiopathic, genetically predisposing, radiation-induced, viral, and chemical carcinogens induced. Quite a few hereditary disorders like Li-Fraumeni syndrome are associated with an increased risk of soft tissue sarcomas of head and neck (STSHN). The pathogenesis of the development of sarcoma remains an enigma.

V. Classification

Sarcomas were classified according to a multitude of criteria, including the previously mentioned tissue of origin, the histologic grade of the neoplasm, and the anatomic subsite within the HN from which the tumour arises. Histopathology plays a major role in the classification of sarcoma[4]. Based on their origin, sarcomas are classified into STS and hard tissue sarcomas (Table 1). Radiation-induced sarcoma (RIS) is considered a separate category, as it can arise in soft tissues or bone. The current 2013 World Health Organization (WHO) Classification of sarcomas of soft and hard tissue was published 11 years after the 2002 volume[5]. Clinical, histological, and genetic data were integrated with the WHO classification.

Table 01- Classification of sarcomas of Head and Neck[4]

Hard tissue sarcomas	1. Osteosarcoma 2. Chondrosarcoma 3. Ewing sarcoma
Soft tissue sarcomas	4. Angiosarcoma 5. Hemangiopericytoma 6. Synovial sarcoma 7. Rhabdomyosarcoma 8. Malignant schwannoma 9. Liposarcoma 10. Leiomyosarcoma 11. Fibrosarcoma 12. Malignant Fibrous Histiocytoma 13. Alveolar soft part sarcoma 14. Kaposi sarcoma 15. Undifferentiated/other category sarcomas
Radiation-induced sarcomas (RIS)	1. Soft tissue sarcoma 2. Hard tissue sarcoma

VI. Diagnosis, Staging & Grading-

The clinical presentation of HNS can be highly variable depending on the specific area of involvement and the proximity to surrounding anatomic structures. Painless mass is the most common symptom in STSHN with which patient's report to the surgeon. Detailed physical examination, although always a requirement and can be augmented with imaging studies. Computerized tomography (CT) has a better evaluation for small tumours in hard tissues and near the air-filled paranasal sinuses and skull base. MRI plays a crucial role in the evaluation of STSHN as it shows the presence of oedema, haemorrhage, necrosis, cystic degeneration, and fibrosis[6]. A chest X-ray or CT thorax should be taken to rule out any metastasis and it also helps in staging. Positron Emission Tomography (PET) has high sensitivity (91%) and specificity (85%) for the detection of sarcomas & has a valuable role in the detection of metastasis and recurrence cases[6-8]. Biopsy from multiple sites has a gold standard value in confirming the diagnosis. Molecular biology has a significant role in the differentiation of a heterogeneous group of sarcomas; however, it is not completely understood. Immunohistochemistry (IHC) has a significant role in the diagnosis of oral carcinomas, oral melanoma, tumours of the salivary gland, benign mesenchymal tumours of the oral cavity, and oral sarcomas. IHC markers

commonly used for diagnosis osteosarcomas, Ewing's sarcoma, rhabdomyosarcoma, leiomyosarcoma, angiosarcomas, and peripheral nerve tumours[9].

The staging system most often used for STS is the American Joint Committee on Cancer (AJCC) TNM system[10] (Table-02). In 2002, WHO changed the histologic grading from the four-grading system to a three-grading system for STS4. Another grading system was given by French or Fédération Nationale des Centres de Lutte Contre Le Cancer (FNCLCC) in which histologic grading was classified into three grades (grade 1, 2 & 3) based on differentiation, mitotic count & tumour necrosis[11]. Both systems are valuable in assessing the prognosis of HNS, the one given by WHO is commonly followed as it is easy to interpret.

Table 02- TNM Classification of Sarcomas of HN[10]

Category		**Criteria for STS**	**Criteria for Hard tissue sarcomas**
Primary tumor (pT)	pTX	Primary tumor cannot be assessed	Primary tumor cannot be assessed
	pT1	Tumor ≤ 2 cm in greatest dimension	Tumor ≤ 8 cm in greatest dimension
	pT2	Tumor > 2 cm and ≤ 4 cm	Tumor > 8 cm in greatest dimension
	pT3	Tumor > 4 cm	Discontinuous tumors in the primary bone site
	pT4	Tumor with invasion of adjoining structures ○ **pT4a:** Orbital, skull base / Dural, central compartment viscera, fascial skeleton or pterygoid muscle invasion ○ **pT4b:** Brain parenchymal invasion, carotid artery encasement, prevertebral muscle invasion or central nervous system involvement via perineural spread	-
Regional lymph nodes (pN)	pN0	No regional lymph node metastasis	No regional lymph node metastasis
	pN1	Regional lymph node metastasis	Regional lymph node metastasis
Distant metastasis (pM)	pM0	No distant metastasis	No distant metastasis
	pM1	Distant metastasis	Distant metastasis M1a- Lung M1b- Bone or other distant site

VII. Discussion

The general concepts of sarcoma management are not unanimously applied in the HN region. The classical treatment modalities employed in HNS are surgery, radiotherapy (RT), and/or chemotherapy. Various factors help in deciding the treatment plan like tumour size, patients age and co-morbidities, tumour location, histologic type, sub-type, and differentiation. Due to the complex anatomy of the HN area, the ability to obtain wide surgical margins becomes limited, leading to a higher local recurrence rate. Distant metastases are rare in the absence of regional metastases if it is there commonly in the lung, and the sight of nodal metastases should incite a quest for distant metastases. The sporadic nature of these tumours accounts for the difficulties encountered in making definitive treatment decisions based on the present literature.

Hard Tissue Sarcomas

Osteosarcoma (OS) is a principal malignant bone tumour that is defined by the presence of cells of mesenchymal origin i.e. the spindle cells, which deposits immature osteoid matrix. An incidence of 3.4 per million people per year has been reported worldwide[12]. OS the most common primary malignancy in children[13]. OS shows a bimodal distribution across age; occurring in the prepubertal phase and after the age of 60 years[13]. OS patients complain of symptoms of nonspecific pain in the affected area. Also, it can frequently present as a pathological fracture. Surgery with negative margins remains the mainstay therapy for all hard tissue sarcomas except Ewing's sarcoma (EWS)[14-17].

Chondrosarcoma of the HN region representing approximately 0.1% of all HN neoplasms and give rise to abnormal bone or cartilage growth. It commonly occurs in the age range of 8–80 years with a peak between 30 and 60 years[18]. It involves almost every site of HN. Tumours of maxilla and antrum more challenging to excise with adequate surgical margins and consequently are less acquiescent to cure. Better survival rate had shown by chondrosarcomas of the mandible and highly differentiated histologic grade. Surgical resection with postoperative radiation is reported as the most widely employed primary treatment[19]. According to WHO, the survival rate of patients with grade 1 chondrosarcoma is 89%. 53% of patients with grade 2 and 3 showed a 5-year survival rate.

EWS is the second most common primary bone malignancy in children and adolescences after OS[20]. They are locally aggressive, commonly occur in males than females, and the first three decades of life[21]. EWS

has site predilection for the skull, mandible, and maxilla[13, 22]. The most common symptom is pain or swelling. EWS is unique among the more common tumours of the HN in that there is a myriad of treatment combinations used with a multidisciplinary approach[23]. No difference has been reported in patients of EWS and OS treated with surgery, RT, or combined surgery and RT[24]. RT had been the standard treatment for EWS, but until the 1960s RT or surgery were the mainstay treatments for EWS[25]. Postoperative RT is indicated/advocated if the margins achieved by surgery are inadequate for local control. By using a combination of local therapy and aggressive multi-drug chemotherapy, there is a decrease both in the incidence of local disease recurrence and the development of metastases, thereby exacting the cure rate from 10% to 75%[26, 27]. The treatment modalities of hard tissue sarcomas are summarized in table 3

Table 03- Comment on management modalities of Hard tissue SHN

Sr. no	Name of the tumor	Management modalities		
		Surgical therapy	**Chemotherapy**	**Radiotherapy**
1	Osteosarcoma	En-bloc resection with three dimensional free margins of at least 1cm[14-17, 21, 28.]	-Adjuvant chemotherapy[29, 30]. -Decreases recurrence & increases survival rate	- Adjuvant RT in those with close or positive margins
2	Chondrosarcoma	Most effective treatment[29, 31].	-limited role -adjuvant in aggressive/high grade tumor	-Controversial - Proton therapy for skull base[32, 33].
3	Ewing's sarcoma	-second line treatment after neoadjuvant chemotherapy[23].	- 3-6 cycles at 2-3 weeks interval before surgery - 6 to 10 cycles post-surgery[34].	-most preferred treatment 31-59 Gy) -adjuvant in positive margins -exclusively for non-operable[23].

Soft Tissue Sarcomas

STSHN exhibits borderline pathological appearance concerning benign or malignant behaviour. Even though they have identical histological patterns, the clinical result is often contrasting and challenging to predict. In adults, Malignant fibrous histiocytoma (MFH) is the most common contributing for 20%–30% of all STS & approximately 1-3% of occurring in the HN[35, 36]. MFH is pleomorphic sarcoma comprising of partly fibroblast-like, partly histiocyte-like cells. Overall, males are more commonly affected than females with a ratio of 3:2[28]. Pain, swelling, or a pathologic fracture are the most common features seen in patients. Radical excision with wide safety margins along with dissection of loco-regional lymph nodes is the primary treatment for MFH[37]. Many studies have shown overall median 5-year survival rates and increases with clear surgical margins[38]. Fibrosarcoma is a malignant tumour of fibroblast accounting 5% in the HN region among all the extraoral fibrosarcoma's[39, 40]. High local recurrence rate and low incidence of local-regional lymph node metastasis are it's distinguishing features[41]. Definite treatment regimen still appears to be ambiguous[42]. The most important part to be considered in treatment and survival rate is adequate resection[43, 44]. Also, prophylactic neck dissection is controversial. The 5-year survival rate is 52% for periosteal fibrosarcoma and 27% for medullary origin fibrosarcoma which suggests a better prognosis for periosteal fibrosarcoma[43]. Rhabdomyosarcoma (RMS) is a malignant tumour beginning in striated muscle. Of all the malignancies, RMS constitutes 3-5% and is the commonest paediatric STS[45]. Most of the cases shown to be male predominance with a ratio of 1.3:1[46]. For the precise diagnosis of RMS detailed physical examination, past medical history, radiological imaging, histology, and molecular tests are essential. Chemotherapeutics agent like Irinotecan and carboplatin with concurrent RT in RMS gives favourable tolerability, efficacy, and local control[46]. An increased survival rate was seen with surgical resection and adjuvant chemo or RT[42, 47].

Angiosarcoma (AS) is a malignant tumour of the inner lining of blood vessels. Less than 5% of cases of STS involve HN, 10% of which are AS[48]. The site and size of the tumour, marginal status, and age are important determinants for poor prognosis. Radical surgery and postoperative RT remains the mainstay treatment with these tumours[49, 50]. Because of the extensive microscopic spread of AS of the scalp, obtaining negative surgical margins become difficult. RT reported to be an inadequate treatment for a potentially curable disease, and RT is also avoided for radiation-induced AS. Liposarcoma is one of the most common malignant mesenchymal neoplasms, accounting to 15% of all STS[51, 52]. Liposarcomas are typically diagnosed in the fourth through seventh decades and are more common in men[6]. It is agreed that surgery with negative margins is the gold-standard treatment for all histologic subtypes of liposarcomas[49]. The difficulty with these tumours is local recurrence. Radiation improves local recurrence; however, it may not have any impact on overall survival[53]. Hemangiopericytoma (HPC) is a mesenchymal vascular sarcoma originating from the pericytes of Zimmerman. HPC was first described as a solitary fibrous tumour by Wagner in 187054. Stout and Murray[55] gave the term ''hemangiopericytoma,'' in 1942. The lesion is not red, which is the main characteristic of other vascular tumours, suggesting a vascular origin. Surgery is considered the mainstay of treatment, though RT and chemotherapy have an integral role[10, 42, 54]. Some authors have suggested presurgical embolization to reduce the risk of intraoperative bleeding when treating this lesion.

Synovial sarcomas (SS) are the malignant tumours which originate from pluripotent mesenchymal cells which constitutes for almost 8% of all STS and often involve the extremities[56]. The most common age group of patients is between 25 and 35 years with male preponderance[57]. The most common site of presentation reported in the literature is the parapharyngeal space, followed by the hypopharynx, however, orbital involvement is reported rarely[58, 59]. Presently, the mainstay of treatment is complete resection with wide margins to constraint local recurrence. Malignant schwannoma (MS) is a rare tumour yet one of the most aggressive malignant lesions in the HN area. In most instances, MS originates from the trigeminal nerve and its branches. The most common clinical sign MS is a painless, rapidly growing mass. Complete surgical removal is the main treatment and most important prognostic factor; however, the resecting margin of the tumour is always difficult and controversial[60]. Adjuvant RT is advised by the oncology consensus group, as part of a uniform treatment policy for MS[61]. Aggressive surgical intervention and RT can result in good survival. The role of chemotherapy in MS is similar to other STSHN but has shown no significant effect on survival rate. Doxorubicin–ifosfamide regimen has been found to be a more superior which warrants further investigation[62-64]. Leiomyosarcoma (LMS) is a malignant tumour of smooth muscle which accounts for only 4% of HN sarcomas. Elderly individuals are among the commonly affected population[65]. Oral cavity, scalp, paranasal sinuses, and jaws are the frequently affecting areas of HN region in LMS[65]. The primary treatment of LMSs is surgical excision with 1 cm free margins. Radical neck dissection is only reserved for late lymph node metastasis. Adjuvant RT or systemic chemotherapy has also been used to treat LMSs[66]. No clear survival benefit has been demonstrated with the use of adjuvant chemotherapy or RT.

Alveolar soft part sarcoma (ASPS) is rare, having incidence of less than 1% of STS with only about 25% of those occurring in the HN. Surgery followed by chemotherapy has been commonly employed as a treatment strategy in most of the studies. Lingual ASPS in young children's are having good prognosis compared to other parts of the body. Kaposi Sarcoma (KS) is a neoplastic antiproliferative disorder characterized by multiple violaceous nodules on the skin of the upper and lower extremities, but rarely in the mucosa of the HN. Palate is the most common site within the oral cavity[67]. Skin lesions are painless bluish-red macule which slowly grows to form indurated plaques and nodules. The most common sites of cutaneous involvement reported are the postauricular region, scalp, and neck. Guidelines for the treatment of patients with AIDS-KS are not well established and are based on limited clinical experience. In the literature, treatment modalities for cutaneous KS include surgery, conventional and megavoltage RT, chemotherapy, immunotherapy, antiviral drugs, and cessation of immunosuppressive therapy in iatrogenically immunosuppressed patients. Doxorubicin, bleomycin, vinblastine, and dacarbazine were the drugs initially used because of success with lymphoreticular lesions. Interferon and thalidomide have shown to have a significant role[68]. The treatment modalities of STSHN and RIS are summarized in table 4

Table 04- Management modalities of STSHN

Sr. no	Name of the tumor	Management modalities		
		Surgical therapy	**Chemotherapy**	**Radiotherapy**
1	Malignant Fibrous Histiocytoma	-primary treatment	-neoadjuvant role is contravercial[38].	-as adjuvant (60 Gy)[69].
2	Fibrosarcoma	- Adequate resection[43] -prophylactic neck dissection is controversial	-adjunctive role is unclear	-primary role in unresectable cases
3	Rhabdomyosarcoma	- primary treatment[70].	- combination of vincristine and dactinomycin commonly used[70] -reduces RT dose if given as induction therapy	- adjuvant to surgery -brachytherapy reduce the dose of external beam RT[70] -50.4 Gy recommended for gross disease
4	Angiosarcoma	-radical surgery[49].	-combined neoadjuvant approach with RT[49] -RT plus rIL-2 immunotherapy[71].	-adjunctive measure to surgery and chemotherapy -improves survival rate by 33%[49]
5	Liposarcoma	-surgery with negative margins is the gold-standard[72].		- adjuvant RT for high-grade tumors, large tumors and positive margins -20% improved survival rate using surgery+RT[73].
6	Hemangiopericytoma	-main treatment with lowest recurrence[74].	-only for infantile HPC[75].	
7	Synovial sarcomas	-complete surgical resection	-long term survival with chemotherapy	-in resection cases with questionable margins[76].
8	Malignant peripheral nerve sheath tumor	-primary treatment modality[60].	-adjuvant role has questiontionable survival benifit[62].	- Adjuvant modality - Dose of 50-60 Gy preferred for intermediate- to high-grade lesions[61].

9	Leiomyosarcoma	-surgical excision with 1cm free margins		
10	Alveolar soft part sarcoma	- complete resection with tumor-free zone of 1 to 1.5 cm[77].	-adjuvant to surgery -monotherapy with IFN alpha-2b showed significant role[78].	
11	Kaposi sarcoma	- recommended modality with limited indications.	- first line of therapy - Doxorubicin, bleomycin, vinblastine, and dacarbazine used in combination therapy	-RT dose of 15 – 30 Gy has curative role
12	Radiation Induced Sarcoma	-shown to have difficult local control[79].		-used as adjuvant

Radiation Induced Sarcomas (RIS)

The incidence of RIS ranges from 0.03 to 0.3% with a predilection for the maxillary region, including the maxillary sinus, alveolar process, palate, and adjacent nasal cavity[79, 80]. The occurrence of RIS is within fibrotic and hardened tissues thus making the early clinical diagnosis challenging. The radiological findings are not pathognomonic and hence differentiation from other neoplastic entities is obscure. The threshold dose for RIS is unknown; however, it has been detected between dose ranges of 1340 cGy to 16,440 cGy with a mean of 6000 cGy[79]. Management of RIS is complicated and truly challenging. The same principles of de novo sarcoma patients should be used to treat these tumours, but, there are limitations given of the therapy. Surgical resection, chemotherapy, further irradiation (external beam or brachytherapy), or a combination of the above are the different treatment options for RIS. Bjerkehagen et al[79] reported a high percentage of positive margins and local recurrence in those patients with RIS who underwent surgery, concluding that local control is difficult to achieve in this type of tumours. Chemotherapy is often used as an adjuvant to patients after surgical resection.

VIII. Conclusion

Sarcomas of the HN region are very rare in nature. as most of the sarcomas are asymptomatic or shows painless mass diagnosis becomes difficult and commonly diagnosed in late the stage the of disease. No standard protocols were given for ST and hard tissue sarcomas of HN. Surgical therapy with adequate margins remains the gold standard treatment for most of the sarcomas. Due complexity of the HN region obtaining negative margins becomes quite difficult so adjuvant CT or RT should be used. Multicentric randomized clinical trials are necessary for definitive treatment.

Abbreviation's-

HN- head and neck, STS- soft tissue sarcoma, STSHN- soft tissue sarcoma of head and neck, RIS – radiation induced sarcoma, CT- computerized tomography, PET- positron emission tomography, IHC- immunohistochemistry, HNS- head and neck sarcoma, RT- radiotherapy, OS- osteosarcoma, ES- ewing's sarcoma, MFH- malignant fibrous histiocytoma, AS- angiosarcoma, HPC- hemangiopericytoma, MS- malignant shwannoma, LMS- leimyosarcoma, ASPS- alveolar soft part sarcoma, KS- Kaposi sarcoma

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