Head and neck soft tissue sarcomas: 40 Year casuistry from ENT Department of IPO-LFG

Original Article

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Abstract

Objective: To identify prognostic factors for Head and Neck Soft Tissue Sarcoma. Assess survival according to sublocation and treatment.
Study design: Observational retrospective study
Material/methods: Analysis of patients data with Head and Neck Soft Tissue Sarcoma followed at the ENT department at IPO-LFG between 1980 and 2022.
Results: 59 patients (64% men) with a mean age of 47,2 years were selected. Chondrosarcoma and rhabdomyosarcoma were the most diagnosed and the larynx was the most affected local. 83% were operated; 28%% relapsed locally; and 10% had metastasis. 17% of patients died from the disease. The patients who received RT exclusively died. The disease-specific survival rate at the 5th year was 83%, with the spindle cell sarcoma having the worst results.
Conclusion: Head and Neck Sarcomas have worse outcomes than other locations. Spindle cell sarcoma and primary treatment with RT were associated with higher mortality. Early surgical treatment is essential.
Keywords: sarcomas, head and neck, tumors

Introduction

Head and neck sarcomas are neoplasms originating from the embryonic mesenchyme, and can be categorized based on their origin into soft tissue sarcomas and bone sarcomas.[1] These are exceedingly rare tumors (with an estimated annual incidence of 5 per 100,000 individuals), with 5–15% of sarcomas occurring in the head and neck region, and are associated with significant morbidity and mortality.[1-3] Factors influencing the prognosis include the type and grade of the tumor, surgical margins, and anatomical location.[4] However, the various histological subtypes of head and

neck sarcomas present challenges for the rigorous study of this class of neoplasms.

The preferred treatment approach is en bloc surgical resection with negative margins, combined with neoadjuvant or adjuvant therapies.[1] Due to the challenge of achieving complete tumor resection in this anatomical location, head and neck sarcomas are known to have higher local recurrence rates and lower overall survival rates compared to sarcomas in other locations.[4] This study examined patients with head and neck soft tissue sarcomas who were referred to the Otorhinolaryngology Service (ORL) at the Portuguese Oncology Institute of Lisbon Francisco Gentil (IPO-LFG), a reference center, with the aim of identifying the prognostic factors associated with these tumors, as well as assessing patient survival in relation to histological subtype and treatment.

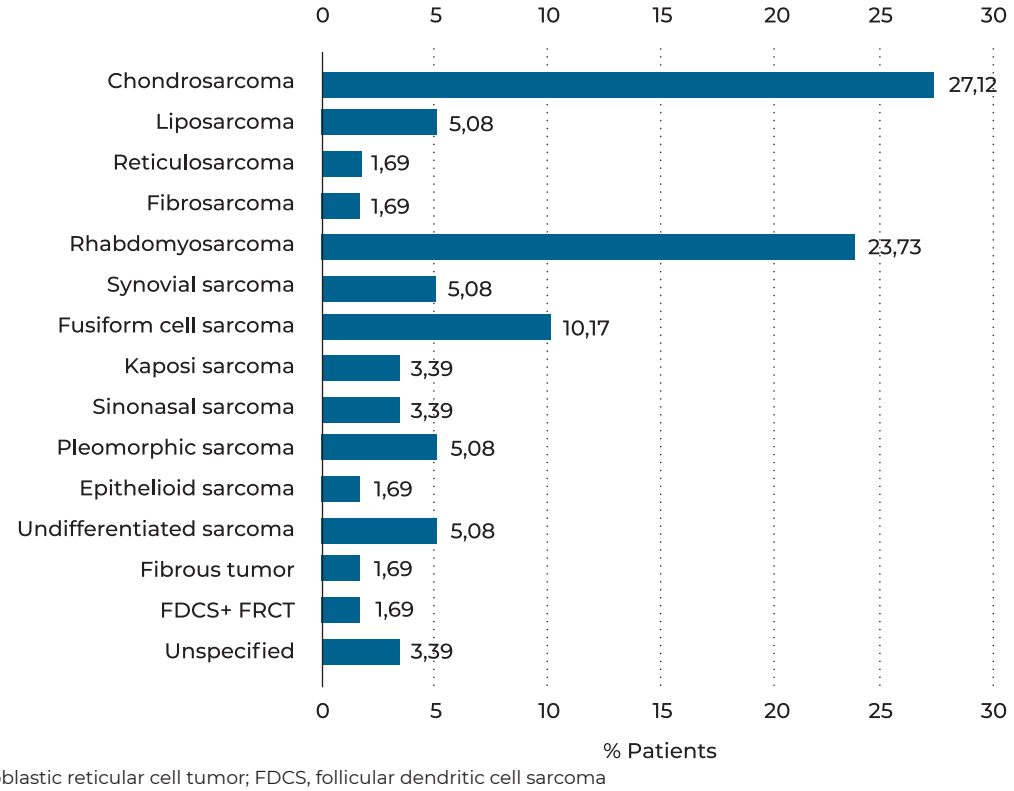
Materials and Methods

To conduct this study, we reviewed the data from the clinical records (both physical and electronic) of patients with head and neck soft tissue sarcoma followed at the OR L department of the IPO-LFG from 1980 to 2022. Subsequently, a statistical analysis was conducted using Microsoft Excel® and SPSS®.

Results

A total of 59 patients were eligible, comprising 64% males and 36% females, with an average age of 47.2 years (range 1–94 years), of which 18% (n=11) were pediatric cases. Additiona lly, 20% and 17% of the cases had tobacco and alcohol habits, respectively. The average follow-up period was 61 months (ranging from 0 to 406 months). The histological diagnosis was varied. As illustrated in Graph 1, the most frequently diagnosed sarcomas were chondrosarcoma (n=16), rhabdomyosarcoma (n=14), and spindle cell sarcoma (n=6). These were followed by liposarcoma (n=3), synovial sarcoma (n=3), pleomorphic sarcoma (n=3), undifferentiated sarcoma (n=3), Kaposi's sarcoma (n=2), sinonasal sarcoma (n=2), reticulosarcoma (n=1), sclerosing epithelioid fibrosarcoma (n=1), epithelioid sarcoma (n=1), solitary fibrous tumor (n=1), and both fibroblastic reticular cell tumor (FRTC) and

Graph 1
Identified histological subtypes of sarcomas



FRTC, fibroblastic reticular cell tumor; FDCS, follicular dendritic cell sarcoma

follicular dendritic cell sarcoma (FDCS) (n=1). In two cases, the histological subtype was not identified. Regarding pediatric patients, 82% (n=9) had rhabdomyosarcoma, and 18% (n=2) had synovial sarcoma. The most commonly reported symptoms were dysphonia, dyspnea, painless mass, and epistaxis, with an average time to presentation of 5 months. The most indolent tumors were chondrosarcoma, Kaposi's sarcoma, and epithelioid sarcoma, with average times to presentation of 16, 8, and 6 months, respectively. The sarcomas with the shortest time to presentation were sinonasal and pleomorphic sarcoma, each with a duration of 1 month, and rhabdomyosarcoma, with an average duration of 3.7 months.

These lesions most frequently originated in the larynx, followed by the nasal cavities and paranasal sinuses. In Table 1, the various histological subtypes are grouped by the anatomical region. A tumor biopsy yielded the correct histopathological outcome in 72% of the cases, with chondrosarcoma having the lowest percentage of accurate diagnoses through biopsy at 67%. Fine needle aspiration cytology (FNAC) was performed in 33% of the cases, and it yielded the correct histological subtype in only 37.5% of the cases. In 12.5% cases, a sarcoma was correctly detected, but the histological subtype of the sarcoma was incorrectly identified; in another 12.5%, malignant cells were detected, but a diagnosis of sarcoma was not made; in 12.5% of cases, the result was inconclusive, and in 25% of the cases, the test was negative for malignancy. A core-cut biopsy was performed in only two c ases, failing to provide the correct diagnosis of neoplasia in one instance; in the other case, while it did diagnose sarcoma, the identified histological subtype was incorrect.

Regarding the staging of chondrosarcomas, 56% were grade I, 25% grade II, and in 19% of the cases, the grade was not identified. No cases were identified as either Grade III or IV.

Regarding the other tumors, 20% were T1, 40% T2, 30% T3, and 10% T4; 28.6% of the cases exhibited regional metastases, and 10% had distant metastases.

Table 1
Localization of the identified histological subtypes of sarcomas

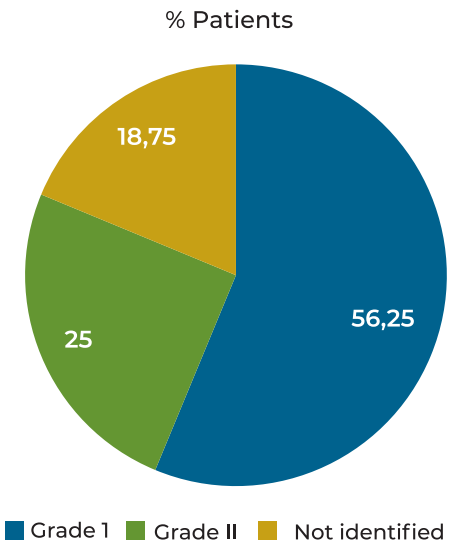
Histology	Base of Skull	Nose & SP	Parotid and Neck	Face	Oral	Trachea/ Larynx/ Pharynx	Heard
Chondrosarcoma						16	
Rhabdomyosarcoma	2	7		2	1	1	1
Fusiform cell sarcoma	1	4	1				
Liposarcoma			1			2	
Synovial sarcoma			1			2	
Pleomorphic sarcoma	1		2				
Undifferentiated sarcoma		1		1	1		
Kaposi sarcoma				1	1		
Sinonasal sarcoma		2					
Unspecified		2					
Reticulosarcoma						1	
Fibrosarcoma				1			
Epithelioid sarcoma			1				
Malignant solitary fibrous tumor		1					
FDCS+ FRCT	1						

Only 17% patients did not undergo surgery as the primary treatment. This was because one patient refused surgery, 5% underwent chemotherapy and radiotherapy, and 12% underwent radiotherapy alone, of which 29% received this treatment with palliative intent. All undifferentiated sarcomas and 66% of spindle cell sarcomas were primarily treated with radiotherapy. All patients who solely underwent radiotherapy (RT) with curative intent died. In addition, 8% of the patients received neoadjuvant chemotherapy (66% of synovial sarcoma cases and 21% of rhabdomyosarcoma cases). The most commonly performed surgical procedures were: excisional biopsy (22%), total parotidectomy, laryngeal microsurgery with debulking of the lesion (a procedure performed exclusively for chondrosarcomas) (16%), partial laryngectomy (14%), maxillectomy (6%), lateral petrosectomy (2%), and partial pharyngectomy (2%). In 8% of the cases, unilateral cervical lymph node dissection was performed – 50% of these cases were synovial sarcoma, 25% were spindle cell sarcoma, and 25% were of an unspecified subtype. The post-surgical histopathological examination revealed surgical margins with tumor presence in 12% of the cases and close-lesional excision in 4% of the cases; 17% underwent RT, 2% received both adjuvant chemotherapy and RT, and 2% underwent adjuvant chemotherapy. The rhabdomyosarcoma was the sole histopathological subtype that received adjuvant chemotherapy. Overall, 28% of the patients experienced loco-regional recurrence/persistence within an average period of 5 months; 66% of pleomorphic sarcomas, 50% of spindle cell sarcomas, 50% of sinonasal tumors, and 38% of chondrosarcomas exhibited recurrence/persistence. In cases of chondrosarcoma, surgery was the chosen treatment for recurrences. In the remaining cases, 40% underwent chemotherapy, 40% received RT, and 20% (n=1-sinonasal) were treated with photon therapy outside of the service. Distant metastases occurred in 10% of the cases, with all of these cases showing pulmonary metastases. One case also exhibited hepatic and retroperitoneal metastases. The tumor most commonly associated with recurrence was spindle cell tumor (66% of cases), followed by rhabdomyosarcoma (7%). The disease-free survival rate was 91.3% at the 1-year mark and 83.05% at the 5-year mark. However, when calculated specifically for each type of tumor, fusiform sarcoma, rhabdomyosarcoma, and pleomorphic sarcoma had the worst outcomes. The disease-free survival rates at 1 and 5 years were 67% and 33% for spindle cell sarcoma, 86% and 71% for rhabdomyosarcoma, and 67% and 67% for pleomorphic sarcoma, respectively. Chondrosarcoma demonstrated the most favorable survival rates, with 100% disease-free survival at both the 1st and 5th year marks. Specifically in the pediatric population, the disease-free survival rate was 100% at 1 year and 81% at 5 years. The overall survival rates at 1 and 5 years were 91.3% and 79.66%, respectively.

Table 2
Staging of other sarcomas

T	N	M
1 (20%)	N0 (71,4%)	M0 (90%)
2 (40%)	N+ (28,6%)	M+ (10%)
3 (30%)		
4 (10%)		

Graph 2
Staging of chondrosarcomas



Discussion

This study is an endeavor to delineate the characteristics and outcomes of head and neck soft tissue sarcomas, and to identify the significant prognostic factors for this highly heterogeneous pathology in a contemporary sample from Portugal. The IPO-LFG is a reference center for oncological pathology, and as such, the ENT (Ear, Nose, and Throat) service ends up treating a significant number of patients with head and neck sarcoma, despite it being a rare condition.

Regarding demographic characteristics, our findings align with those from other studies, such as the one conducted by *Tejani MA et al.* in the United States, which reported that the average age at diagnosis was 50 years. Additionally, pediatric cases account for 10–20% of the total cases, with a male-to-female ratio of 2:1.[2,4,5] Theoretically, rhabdomyosarcoma, neuroblastoma, and fibrosarcoma are the histological subtypes most commonly found in the pediatric age-group.[2] Indeed, in our series, 82% of the pediatric cases were rhabdomyosarcomas.

Despite the two most commonly reported symptoms being dysphonia and dyspnea, followed by a painless mass and epistaxis in our study, the latter two are the symptoms most frequently documented in the literature.[2,6] However, in our study, we had a higher proportion of chondrosarcoma compared to the previous studies, which may be the reason why the two laryngeal symptoms are more common in our sample.

Histopathological analysis of a biopsy sample is currently the only reliable technique available for a definitive diagnosis. However, in our study, it only provided the correct diagnosis in 72% of the cases. This percentage was even lower for chondrosarcomas, where biopsies failed to provide the correct diagnosis in 33% of the cases.[4,7] Although FNAB is an easy, quick, and inexpensive procedure, it does not yield an accurate diagnosis in a considerable percentage of cases and does not allow tumor classification. In our study, FNAB did not test positive for malignant cells in 37.5% of the cases.[4] Despite core needle biopsy being performed in only two patients in our series, it is a good alternative as it allows for the acquisition of sufficient tissue for an accurate diagnosis, including tumour classification.[8] Excisional biopsies should be reserved for lesions smaller than 3 cm in diameter.[7]

Surgery remains the treatment of choice, as in this and previous studies, it demonstrated superior outcomes compared to RT.[2,4] Indeed, in our case series, all patients who underwent exclusive RT had a fatal outcome.

The tumor size and surgical margins were found to affect survival, consistent with the findings in the literature. Therefore, every effort must be made to achieve complete resection of the tumor. However, due to the anatomical complexity and presence of vital anatomical structures in the head and neck region, wide resection with adequate margins is not feasible in all cases. Consequently, the chosen treatment often involves surgery followed by adjuvant RT.[2,9] Therefore, it is also crucial that these patients are immediately treated at a center experienced in reconstructive surgery to reduce morbidity.

Adjuvant RT is recommended for high-grade tumors, positive surgical margins, lesions larger than 5 cm, and recurrences.[2,4] Recurrences are the sole indication for neoadjuvant RT.[9]

Exclusive chemotherapy is not recommended for the treatment of soft tissue sarcomas of the head and neck, and indeed in our retrospective study, no patient underwent chemotherapy as a sole treatment.[4,11] Nevertheless, head and neck sarcomas respond well to chemotherapy when combined with RT, particularly when tumor resection is not feasible or in cases with a higher risk of distant metastases.[2,11] It may also be indicated alongside RT and surgery for high-grade soft tissue sarcomas. Chemotherapy is sometimes administered prior to surgical procedures in order to shrink the size of large sarcomas, especially those located near vital structures.[2,9,11]

Head and neck sarcomas present with cervical lymph node metastases in approximately 3% of the cases, which is why cervical lymph node

dissection is not routinely performed. It is only recommended in patients with palpable cervical lymphadenopathy or high risk of nodal metastases.[2] Synovial sarcoma and spindle cell sarcoma were the only subtypes in which cervical lymph node dissections were performed in our study population. However, according to the international literature, nodal metastases are more commonly associated with embryonal rhabdomyosarcoma, epithelioid sarcoma, clear cell sarcoma, synovial cell, and vascular sarcomas.[2] As observed in our series, the lungs are the most frequent site of metastases.[2]

The overall survival rates at 1 and 5 years were 91% and 79%, respectively, higher than the 5-year survival rates reported in the literature, ranging from 46 to 55%.[2,4,9,10,11] Local control and distant metastases are the most significant factors related to the survival of patients with soft tissue sarcomas. In contrast to other case series where rhabdomyosarcoma was the most commonly encountered tumor, in our study, chondrosarcoma was the most frequently diagnosed histological type. This type of tumor rarely presents with metastatic disease and is more amenable to local control, which may partly explain the higher overall survival observed in our cohort.[2,4] This is further supported by the fact that the 5-year disease-specific survival rate was lower in the pediatric population, a group in which no cases of chondrosarcoma were diagnosed. However, head and neck sarcomas exhibit higher rates of local recurrence and lower overall survival rates compared to sarcomas occurring in other anatomical regions, primarily due to the anatomical constraints in the head and neck region that limits complete functional resection.[4,11]

Conclusion

The treatment of head and neck sarcomas is complex, and the outcomes are generally less favorable compared to sarcomas originating in other locations. Despite the fact that favorable outcomes can be achieved with early surgery, the surrounding anatomical complexity presents a challenge. Early and effective surgical treatment has a significant effect on the recurrence rates and prognosis; thus, it is essential that these cases are managed in specialized tertiary centers with multidisciplinary teams.

Conflicts of Interest

The authors declare that there is no conflict of interests regarding the publication of this paper.

Data Confidentiality

The authors declare having followed the protocols in use at their working center regarding patients' data publication.

Protection of humans and animals

The authors declare that the procedures were followed according to the regulations established by the Clinical Research and Ethics Committee and to the 2013 Helsinki Declaration of the World Medical Association.

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Availability of scientific data

There are no datasets available, publicly related to this work.

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