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Soft tissue sarcomas of the head and neck. Clinical and pathological evaluation of 108 cases in Mexico☆

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ABSTRACT

Background: Sarcomas constitute 1% of all malignancies, but 10% occur in the head and neck (HN), and they are poorly understood. We present a large series of Soft Tissue HN sarcomas in Mexican patients with survival analysis.
Study design: This is a retrospective study of cases in a cancer hospital.
Methods: Review of files and pathology material. Literature review.
Results: We analysed 108 patients (55 men / 53 women). The age at presentation was 37 years. The original diagnosis changed in nine cases (8.3%). The most common subtype was rhabdomyosarcoma. Ninety percent of tumours were deep, 91% were high grade, 44% had metastasis, 63% measured >5 cm, overall 5-year survival (5y-OS) was 48%, and histological high grade was associated with poor survival (p=0.026).
Conclusion: Sarcomas of the HN are rare. The most affected sites were paranasal sinuses. The majority of tumours were deep, > 5 cm and high grade, 50% had metastasis, the 5y-OS was 48% and the only independent factor associated with 5y-OS was histologic grade.

1. Introduction

Sarcomas are mesenchymal neoplasms that constitute 1% of human malignancies, however, 4–10% of them present in the head and neck area (Huber et al., 2006). Soft tissue sarcomas of the head and neck (STSHN) comprise 50 different histological subtypes with a wide spectrum of biological behaviour. Most sarcomas are slow growing and locally aggressive neoplasms, but nodal metastases occur in 3–10% and distant metastases in 28% (more common in lung, bone, liver and central nervous system), more frequently found in high-grade sarcomas (De Bree et al., 2010). The histopathological classification has evolved in recent decades due to advances on immunohistochemistry (IHC), electron microscopy and cytogenetics, becoming rare the diagnoses of unclassifiable sarcoma and undifferentiated pleomorphic sarcoma (UPS, so called malignant fibrous histiocytoma) (García and Folpe, 2010).

Surgical treatment is the cornerstone of patient care. The extension of resection and surgical margins are important predictors for persistence, recurrence and overall survival. However, STSHN often arise in hidden anatomical areas of the head and neck, making surgical access and complete resection very difficult in comparison to counterparts of the extremities (Hoffman et al., 2004 and Colville et al., 2005). At this location, the most common site of involvement reported in previous series is the scalp, followed by sinonasal tract. The most common histological types reported are unclassifiable sarcoma, angiosarcoma and UPS (Guillou et al., 1997).

Our current knowledge about STSHN is based on several series published few decades ago, where there is no staging in as many as 39% of cases (Eeles et al., 1993) and also in isolated case reports (Lukšić et al., 2011 and da Silva et al., 2012) and small series of tumours located in bone. In this report, we present a series of STSHN in a Mexican population with the objective of evaluate the

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demographics, clinicopathological data and survival analysis. Also, we compare our findings with similar published studies and a literature review.

2. Methods

From a database of 1652 adult patients (>14 years-old) with primary sarcomas collected at our institutions in the period between 1985 and 2010, and excluding cases corresponding to bone sarcomas, odontogenic sarcomas, cutaneous sarcomas, Kaposi sarcoma, iatrogenic sarcomas (radiation induced) and metastasis. Of total, 108 (6.5%) patients met the criteria for inclusion. Documented data were age, sex, tumour location, histological type, histological grade, tumour size, depth of tumour, nodal status, presence or absence of metastases, surgical resection and clinical stage.

We used the American Joint Committee on Cancer AJCC staging, seventh revision (2010) to reclassify all cases, in addition to those established by the College of American Pathologists (CAP). We classify the sarcomas according to current World Health Organization (WHO) classification (Fletcher et al., 2002). Surgical resection R2 is considered when macroscopically visible tumour is on the section margin, R1 is considered when microscopic tumour is present in the section margin, and R0 is a surgical limit without neoplastic cells. The used grading system was the French system FNCLCC (Fédération Nationale de Centres de Lutte Contre le Cancer), which is based on three categories, based on the criteria described by Trojani which consist of necrosis, atypia and mitosis (Coindre et al., 1986). All histologic material was reviewed to corroborate the histological diagnosis, even with the corresponding molecular and immunohistochemical markers. To compare differences between qualitative variables (nominal and ordinal) we used the nonparametric Pearson Chi square with p value for statistical significance <0.05. For survival analysis, we used the Cox regression method with a Kaplan–Meier test. Finally, we review the literature in MEDLINE, EBSCO and OVID databases under keywords *Head and Neck sarcomas* and *soft tissue sarcoma* by limiting the search for works written in English, Spanish, French or Italian, studies in adults and STSHN. Thirteen series met inclusion criteria. We compared our results with the data of literature series.

3. Results

3.1. Demographic and clinical characteristics

Of the 108 patients, the mean age was 37.2 years (range 14–79 years), for men the average age was 34.5 years, and for women 38.5 years. The other demographic and clinical characteristics of the cases registered in this series are shown in Table 1. The most frequent sites of origin as shown in Table 2 were the paranasal sinuses with 23 (21%) cases. The most common histological type was rhabdomyosarcoma in 18 patients (17%) (Table 2). There were nineteen sarcomas initially diagnosed as unclassifiable sarcoma, all of them had paraffin-embedded tissue, which allowed new IHC studies to reclassify four of them as malignant peripheral nerve sheath tumour (MPNST), two as synovial sarcoma, one as leiomyosarcoma and two as UPS.

Most sarcomas occurred in the 20–39 years old group followed by the 40–50 years old group. Sarcomas not showed to be more prevalent in a subsite in the HN area ($p = 0.07$) but Table 3 shows that 66% of scalp located sarcomas were primitive neuroectodermal tumours; in orbit, nasal cavity and jaw the most prevalent subtype was the rhabdomyosarcoma; and the frequency locations of the rest of the various sarcomas was similar.

Table 1
Clinic and pathologic characteristics of 108 head and neck sarcomas.

Feature	Number (%)	p
Sex		
Male	55 (51)	0.847
Female	53 (49)	
Age		
<20 years	19 (17)	0.001
20–39 years	44 (41)	
40–59 years	29 (27)	
≥60 years	16 (15)	
Size and deep		
T1a	6 (5.5)	<0.001
T1b	31 (29)	
T2a	4 (3.7)	
T2b	60 (55.5)	
Without data	7 (6.3)	
Lymph node metastasis		
N0	89 (82.4)	<0.001
N1	10 (9.2)	
Without data	9 (8.4)	
Distant metastasis		
Si	89 (82.4)	<0.001
No	10 (9.2)	
Without data	9 (8.4)	
Histologic grade		
G1	10 (9)	<0.001
G2	35 (32)	
G3	63 (59)	
Clinical stage (AJCC)		
IA	3 (2.7)	<0.001
IB	7 (6.4)	
IIA	28 (26)	
IIB	17 (16)	
III	35 (32.5)	
IV	10 (9)	
Without data	8 (7.4)	
Resection $n = 72$		
R0	52 (72)	<0.001
R1	20 (28)	

AJCC = American Joint committee on cancer.

10% of cases occurred in a localized stage with little metastatic potential while 45% of cases with high-grade tumours have high metastatic potential, and 44% of the cases had metastasis.

Seventy-two patients underwent surgery which of complete resection R0 in 52 (72%) cases and R1 resection in 20 (28%) cases,

Table 2
Location and subtype of 108 head and neck sarcomas.

Site	Number (%)	Subtype	Number (%)
Paranasal sinuses	23 (21)	Rhabdomyosarcoma	18 (17)
Soft tissues of neck	17 (16)	Malignant nerve sheath tumour	14 (13)
Orbit	11 (11)	Undifferentiated pleomorphic sarcoma	13 (12)
Soft tissues of head	10 (9)	Synovial sarcoma	11 (10)
Perimandibular soft tissues	9 (8)	Unclassifiable sarcoma	11 (10)
Salivary glands	9 (8)	Angiosarcoma	8 (7)
Oral cavity	6 (5)	Fibrosarcoma	7 (6)
Nasal cavity	4 (4)	Leiomyosarcoma	5 (5)
Parapharyngeal space	4 (4)	Liposarcoma	5 (5)
Meninge	4 (4)	Neuroectodermal tumour/Ewing sarcoma	5 (5)
Scalp	3 (3)	Hemangioendotelioma	2 (2)
Conjunctive	2 (2)	Dermatofibrosarcoma	4 (4)
Thyroid	2 (2)	Myofibrosarcoma	2 (2)
Larynx	2 (2)	Other	3 (2)
Pharynx	2 (2)		