

**ORIGINAL RESEARCH**

**HEAD AND NECK SARCOMAS  
OUR EXPERIENCE AT A TERTIARY CARE CENTER IN RABAT, MOROCCO**

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**ABSTRACT**

**Introduction:**

Head and neck sarcomas are rare, malignant and very heterogeneous tumors. The difficulty to manage these sarcomas requires the intervention of a multidisciplinary team to improve the prognosis. The aim of our study is to report our series (epidemiological, histological and progressive characteristics) and evaluate our treatment results.

**Patients and methods:**

This is a retrospective study on 42 cases of head and neck sarcomas, assembled at ENT and Maxillofacial Surgery department in the University Hospital of Rabat, for a period of 5 years (2010-2015). All sarcomas were confirmed by histological examination with immunohistochemical study.

**Results:**

There were 29 men and 13 women. The average age of our patients was 35.5 years (extremes of age: 13 and 70 years). All patients received a CT scan with MRI scan in 21 cases. A remote extension assessment showed lung metastases in 8 cases. The most frequently found histological type was synovial sarcoma, which was noted in 13 patients (30.9%) followed by osteosarcoma (21.2%). The osteosarcoma treatment was curative in 19 cases, based on surgery with radiotherapy. Total remission was noted in twelve patients.

**Conclusion:**

Head and neck sarcomas are rare and very aggressive cancers with poor prognosis. The treatment approach combines surgery and chemoradiation. However, in the absence of adequate and effective treatment protocols, it is necessary to establish a surgical indication in time to ensure excision as complete as possible.

**KEY WORDS:** Head and neck, ENT, sarcomas, cancers, surgery.

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**INTRODUCTION :**

Sarcomas of the head and neck are rare tumors, representing only 1% of all malignant tumors of the head and neck, and 5% of all sarcomas [1,2]. Their incidence is 3 to 4.5 on 100 000. They are a heterogeneous group of malignant tumors that share the same mesenchymal origin. They are characterized by slow growth, loco-regional aggressiveness and distant metastatic potential. The etiology of sarcoma is not yet fully elucidated.

The aim of our study is to determine, through our series of cases selected and managed within the ENT and Maxillofacial Surgery department in the University Hospital of Rabat, with a review of literature, the epidemiological, histological, clinical, and progressive characteristics, and above all treatment modalities of head and neck sarcomas and their prognosis.

## MATERIAL AND METHODS

Medical records of all patients with head and neck sarcomas diagnosed and/or managed in the ENT department of Rabat from October 2010 to April 2015 were reviewed.

All patients whose diagnosis was confirmed by a pathological report with immunohistochemistry were included in this study, including radiation-induced sarcomas.

We recorded demographic and clinicopathological characteristics, including age, sex, symptoms, tumor site, size, histology, treatment modalities, and evolution.

Histological grade was evaluated according to the classification of The French Federation of Comprehensive Cancer Centers (UNICANCER Federation) [2]. The diagnosis of radiation-induced sarcomas was based on the criteria of Arlen et al [2].

All statistical analyses were performed using SPSS software.

## RESULTS

Over a period of 5 years, 42 patients were diagnosed with sarcoma in the head and neck region. The series included 23 men and 19 women, aged from 3 to 67 years. The average age was 31.6 years.

The average consultation time was eight months, with extremes ranging from 1 month to 4 years. Concerning patients with medical history, 7.1% of our patients were smokers and 9.5% were treated for a previous cancer.

The symptoms prompting patients to seek consultation were: the presence of a mass in 30 patients (71.4%), a limitation of mouth opening in 5 cases (11.9%), dysphonia in 3 cases (7, 1%) and epistaxis in 4 cases (9.5%) (See Figure 1 and Figure 2).



**Fig. 1:** Examples of patients during the first consultations

- A- patient with mandibular osteosarcoma;
- B- patient with cervical synovial sarcoma;
- C- patient with jugal leiomyosarcoma;
- D- patient with orbital rhabdomyosarcoma.

The most frequently affected sites were bone, mandible sites (12 cases - 28.6%), maxillary sites (4 case- 9.5%), salivary glands in 7 patients (16.7% -5 parotid cases and 2 submaxillary cases), pharyngolaryngeal failure

represented 7.2% - 3 cases. The exact distribution of the different tumor sites is detailed in Table 1 and Graph 3.

Locations	Number	Percentage (%)
Larynx	1	2.4
Pharynx	2	4.8
Mandible	12	28.6
Maxilla	4	9.5
Parotid	5	11.9
Submaxilla	2	4.8
Nasopharynx	4	9.5
Nasal pits	2	4.8
Pterygoid muscle	4	9.5
Parapharyngeal space	1	2.4
Inside of the cheek	1	2.4
Ethmoido-orbital	1	2.4
Hyoid bone	1	2.4
Tongue	2	4.8
<b>Total</b>	<b>42</b>	<b>100</b>

**Table 1:** The different tumor sites in the series.

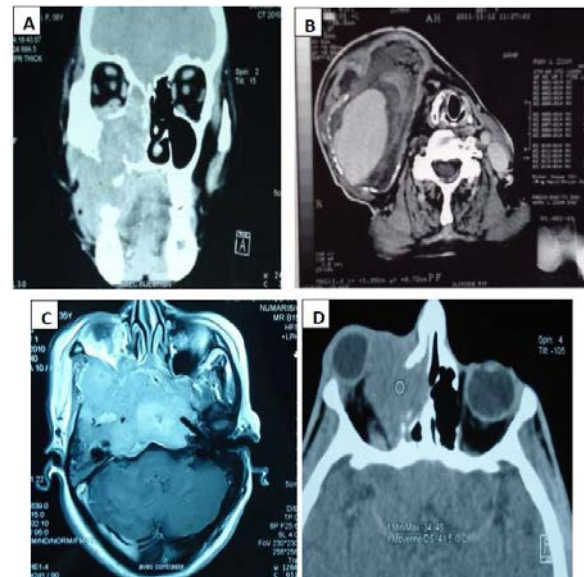
The most frequent tumor was synovial sarcoma (14 cases, 33.3%), followed by osteosarcoma (8 cases, 19%) and rhabdomyosarcoma (7 cases, 6.7%).

The rest is shown in Table 3.

Subtype	Number	Percentage (%)
Rhabdomyosarcoma	7	16.7
Osteosarcoma	8	19
Angiosarcoma	1	2.4
Leiomyosarcoma	3	7.1
Chondrosarcoma	4	9.5
Ewing's sarcoma	1	2.4
Liposarcoma	1	2.4
Synovial sarcoma	14	33.3
Fibrosarcoma	1	2.4
Sarcoma	2	4.8
<b>Total</b>	<b>42</b>	<b>100</b>

**Table 2:** The distribution of sarcoma subtypes in the series.

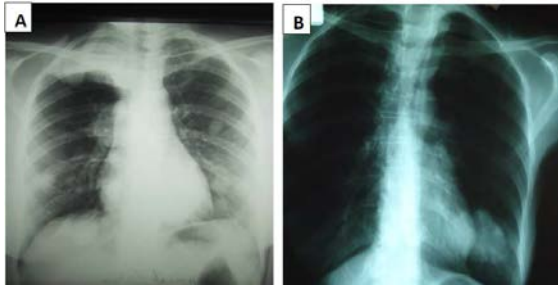
All of our patients received a CT scan, with MRI scan in 21 cases (Figure 2).



**Figure 2:** Examples of the imaging of our patients.

- A-CT image in coronal section, parenchymal window showing an orbital tumor involving the maxillary sinus;
- B- MRI axial image, showing a tumor of the mandible;
- C- MRI image in axial T2, showing a nasopharyngeal procedure;
- D- CT axial image, parenchymal window showing an orbital tumor.

A remote extension assessment showed lung metastases at diagnosis, and this was in eight patients whose evolution was marked by quick death after diagnosis (Figure 3).



**Figure 3:** Chest X-rays showing lung metastasis.

- A-** Appearance of ball release;  
**B-** Left basal pericardial metastasis.

The treatment was curative in 19 cases, based on surgery followed by radiotherapy in 4 cases, preceded by neoadjuvant chemotherapy in 6 cases (Figure 4). The remaining patients received palliative chemoradiotherapy. Most tumors were greater than 5 cm in size (75%). The most common type of management was surgery (30 cases, 71.4%), followed by radiotherapy (24 patients, 57.1%) and chemotherapy (23 patients, 54.8%). The surgical margins were clean for 17 patients, limited for 4, overgrown for 5, and not determined for 11 patients.



**Figure 4:** Postoperative appearance

- A-** patient operated for a cervical synovial sarcoma;  
**B-** patient operated for chondrosarcoma of the maxilla.

Total remission was noted in 16 patients, 10 cases of local recurrence, 5 cases of distant metastasis, 6 deaths (Figure 5). The average life span was 20 months. The 3-year survival was estimated at 50%, and 5-year survival at 9.5%.



**Figure 5:** Recurrent tumor of the mandible (synovial sarcoma).

- A-** one patient with local recurrence of synovial sarcoma;  
**B-** The scannographic appearance.

A statistical analysis was performed in single and multiple logistic regression to determine the most important factors in evolution. This analysis indicated that only surgery is the factor determining a favorable evolution (OR 1.8; CI 95% 1.1-2.5;  $P = 0.01$ ).

## DISCUSSION

Head and neck sarcomas are very rare: they account for only 1% of primary tumors of the head and neck region [3] and 4-10% of sarcomas in general [4]. In most studies, the primary sarcomas of the head and neck represent only 5% to 15% of all cases of sarcoma in [5] adults. However, in the pediatric population, 35% of all sarcomas occur in the head and neck [6].

Sarcomas occur at any age. However, head and neck sarcomas occur at a young age [7]. Biphasic presentation was noted by some authors: with 80-90% affecting adults and 10-20% affecting young people [8, 9]. However, in the pediatric population, one in three sarcomas occur at the level of the head and neck [10]. In our series, the average age was 31 years, which comes within the framework of young adult, which is, therefore, similar to the literature. The predominance of sex varies from one series to another. In our case, male dominance was clear (Table 4).

Sarcomas have various cellular origins, but they are grouped together because of their clinical, progressive, treatment similarity as well as their prognosis [7-9, 11]. They are characterized by slow growth, loco-regional aggressiveness and distant metastatic potential [10, 11]. The etiology of sarcoma is not yet well elucidated [7]. However, some factors may be responsible for sarcoma, such as: exposure to ionizing radiation, exposure to certain chemicals and association with genetic mutations [7]. Environmental and immunological factors were also responsible [11, 12]. Some point that trauma and chronic infections may play a role in the development of sarcomas [11]. Most sarcomas of the head and neck occur with nonspecific symptoms. In 65 to 95% of cases, they manifest through a palpable mass. In our series, a visible or palpable swelling was the most commonly found cause (71.4%), followed by dysphonia (7.1%), epistaxis (9.5%) and the limitation of mouth opening (11.9%).

The lung parenchyma is the preferred metastatic site of soft tissue sarcoma: an estimated 20-38% of patients will develop pulmonary metastases during their illness.

The diagnosis of primitive sarcomas is often difficult because of the rarity of these tumors, the wide variety of histogenetic types, and the existence of lesions that are benign, pseudosarcomatous, and sometimes deceitful. The majority of head and neck sarcomas are soft tissue sarcomas with only 20% of natural bone or cartilage [8-10, 13]. In descending order, osteosarcoma, rhabdomyosarcoma, malignant fibrous histiocytoma, fibrosarcoma and angiosarcoma are the most frequently encountered histologic types in the head and neck region and represent approximately 50% of all sarcomas of such region [10-11]. Over the past ten years, many genetic disorders have been described, allowing a molecular classification [14].

Treatment depends on the histologic type, stage, location, tumor size and patient age [11, 15]. It includes several means: surgery, radiotherapy and chemotherapy. The overall five-year survival is between 44% and 80% and disease-free survival varies between 45% and 62% [11]. This variability is due to the heterogeneity of these tumors and the lack of standardization of treatment modalities. For head and neck sarcomas, tumors greater than 5 cm, high histologic grade, and the limits of tumor resections correlate with increased local failure rate and decreased disease-free survival [8, 11, 12]. In our series (see Table 3), the survival rate was too low (less than 25 years to 5 years) which can be explained by the late period of consultation making the tumor too developed with greater than 5 cm in size in 68% of cases.

Types of sarcomas	2 years	5 years	10 years
Chondrosarcoma	90%	81%	70%
Chordoma	87%	74%	58%
Ewing's sarcoma	80%	68%	60%
Hemangiosarcoma	51%	34%	23%
Kaposi's Sarcoma	44%	27%	23%
Liposarcoma	92%	87%	71%
Malignant fibrous histiocytoma	85%	73%	60%
Osteosarcoma	71%	60%	52%
Rhabdomyosarcoma	71%	56%	25%
All sarcomas (literature)	70%	57%	47%
All sarcomas (our series)	68%	24%	0%

**Table 3:** Survival at 2.5 and 10 years depending on the types of sarcomas.

### CONCLUSION

Head and neck sarcomas are a rare and heterogeneous entity. Their management is mainly based on surgery with radiotherapy. Chemotherapy keeps a limited place for extended or metastatic shapes, or shapes outside any treatment resource as a palliative treatment.

### AUTHORS' CONTRIBUTIONS

The participation of each author corresponds to the criteria of authorship and contributorship emphasized in the [Recommendations for the Conduct, Reporting, Editing, and Publication of Scholarly work in Medical Journals](#) of the [International Committee of Medical Journal Editors](#). Indeed, all the authors have actively participated in the redaction, the revision of the manuscript and provided approval for this final revised version.

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Declared none.

### PATIENT CONSENT

Written informed consent was obtained from patients for publication of this study and any accompanying images.

### COMPETING INTERESTS

The authors declare no competing interests.

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