World Journal of Advanced Research and Reviews
eISSN: 2581-9615 CODEN (USA): WJARAI
Cross Ref DOI: 10.30574/wjarr
Journal homepage: https://wjarr.com/

(RESEARCH ARTICLE)

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Head and neck sarcomas: A series of 20 cases

Hajar Ouazzani Chahdi *, Oumaima Oueriagli Nabih, Amal Akammar, Nizar El Bouardi, Meriam Haloua, Badreedine Alami, Moulay Youssef Alaoui Lamrani, Mustapha Maaroufi and Meryem Boubbou

Hassan II university hospital of Fez, Department of Mother and Child Radiology, Faculty of medicine, pharmacy and dental care, Sidi Mohammed Ben Abdellah University, Fez, Morocco.

World Journal of Advanced Research and Reviews, 2024, 21(03), 192–202

Publication history: Received on 06 January 2024; revised on 27 February 2024; accepted on 29 February 2024

Article DOI: https://doi.org/10.30574/wjarr.2024.21.3.0692

Abstract

Head and neck sarcomas are rare and histologically diverse malignancies, which represents a diagnostic challenge for radiologists because they have many overlapping imaging features. They account for less than 1% of all malignant tumors in the body. People of all ages can be affected, regardless of gender. Rhabdomyosarcoma is the most common histological form, especially in children. Due to the risk of metastasis, extension is absolutely necessary.

Their treatment requires the intervention of a multidisciplinary team in specialized units to improve prognosis.

The aim of our work is to review, through our series, the contribution of imaging resources in the diagnostic approach of tumor nature, to establish the "baseline" workup and post-therapeutic follow-up, as well as to analyze the epidemiology, clinical and histological characteristics of head and neck sarcomas.

Keywords: Head and neck; Soft tissue; Sarcoma; CT and MRI findings; Surgery; Cancer.

1. Introduction

Sarcomas are rare malignant tumors, comprising less than 1% of all malignant tumors in the body. They arise from connective and supporting tissues, distinguishing themselves from the more common carcinomas of epithelial origin. Sarcomas exhibit a broad range of pathological diversity, with over 50 histological subtypes classified according to the specific tissue and cell type from which the tumor originates. There are generally two main groups of sarcomas: bone sarcomas and soft tissue sarcomas.

Radiological examinations, such as ultrasound, CT, and MRI, play a crucial role in the detection and characterization tumor, thus facilitating better management. The diagnosis of sarcoma is based on anatomopathological findings.

The therapeutic management of sarcomas mainly involves a combined approach of surgery and radiotherapy.

2. Methods

This is a retrospective descriptive study involving 20 cases of head and neck sarcomas collected at the Radiology and Otolaryngology Department of Hassan II University Hospital in Fez over a 6-year period, spanning from January 2017 to January 2023.

* Corresponding author: Hajar Ouazzani Chahdi

All sarcomas underwent at least one imaging examination (CT and/or MRI) and were confirmed by anatomopathological examination of the biopsy or excision material.

3. Results

The study included 12 female and 8 male participants, resulting in a sex ratio of 0.66. The median age of the patients was 33 years, with ages ranging from 6 years to 77 years. The average time to consultation was 7 months, with a range of 25 days to 12 months.

Table 1 Various locations of head and neck sarcomas

locations	Number of patients (%)
Maxillary sinus	8 (40)
Orbit	4 (20)
Nasal fossa	3 (15)
Mandible	1 (5)
Infratemporal fossa	1 (5)
Mastoid	1 (5)
Parotid	1 (5)
Base of skull	1 (5)

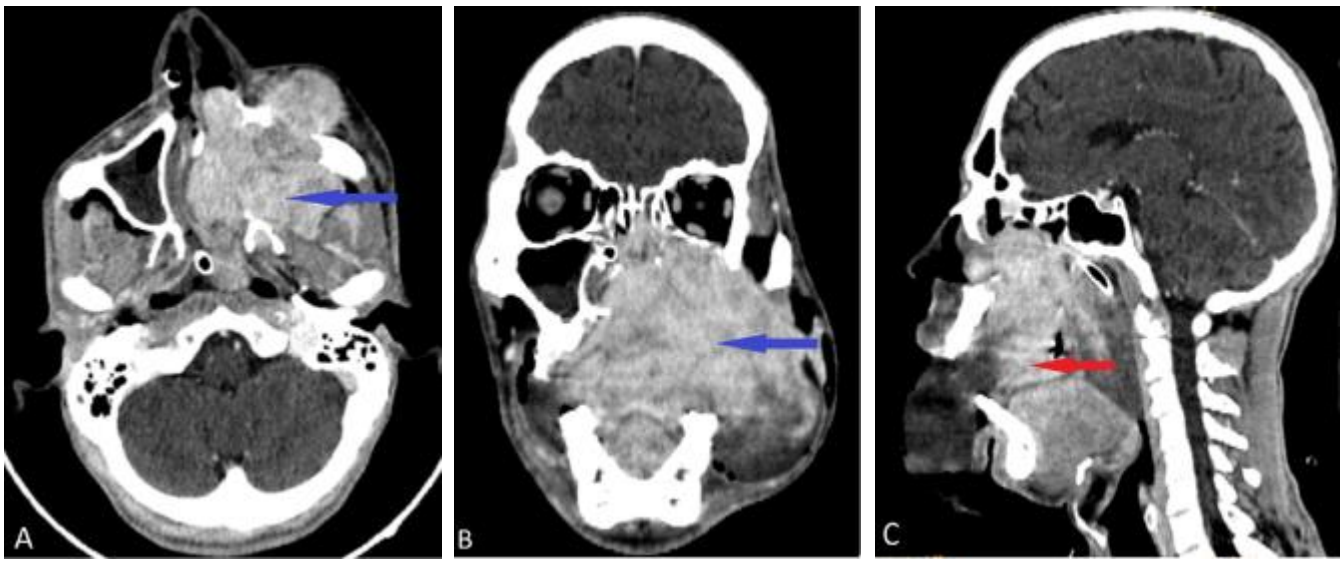

Figure 1 Head and neck CT scan after contrast injection, in axial sections (A), with coronal (B)and sagittal (C) reconstrucions:

Locally advanced tumor centred on the left maxillary sinus (blue arrow), heterogeneously enhanced after contrast, extends towards the infra-temporal fossa laterally, and protrudes and almost completely fills the rhinopharynx, oropharynx and oral cavity poteriorly (red arrow)

[Reference: Mother-child radiology department/ Hassan II university hospital of Fez]

The most common mode of discovery was a mass or swelling, which was found in 8 patients (40%). Exophthalmos was present in 4 cases (20%), while nasal obstruction was present in 3 cases and facial pain in 2 cases, associated with rhinorrhea and epistaxis in one patient.The most frequent sites of head and neck sarcomas in our series were the

maxillary sinus with 8 cases, accounting for 40%, followed by the orbital cavity in 4 cases, representing 20%. Various locations have been illustrated in **Table 1**.

Cervico-facial CT scans were performed on all patients to detect and characterize the mass (size, structure, vascularity) and assess tumor extension at the time of diagnosis. Complementary MRI was requested for only 7 out of all the patients, which is 35%. This was done to improve the pre-therapeutic study of the soft tissues.

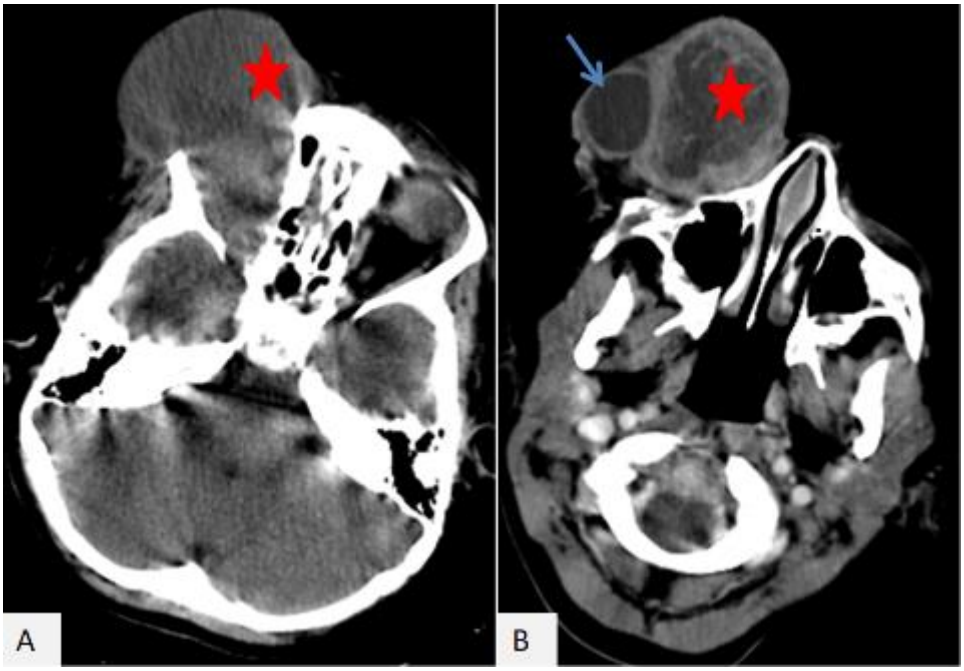

Figure 2 Contrast Enhanced orbital CT scan in axial sections: locally advanced right intra-orbital tumoral process (red star), heterogeneously enhanced after contrast, deforming and pushing back the eye (blue arrow) causing Grade III Proptosis.

[Reference: Mother-child radiology department/ Hassan II university hospital of Fez]

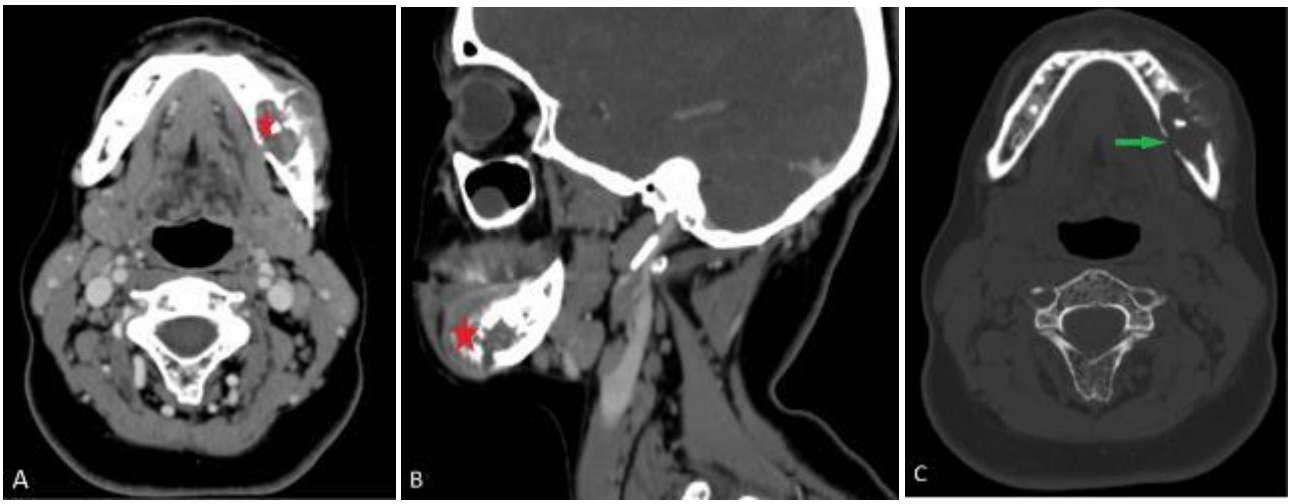

Figure 3 Facial CT scan after contrast injection, in axial sections(A), with sagittal reconstructions(B) and bone window (C).

Osteolytic lesion process involving the horizontal ramus of the left mandible (Red star), heterogeneously enhanced after contrast, responsible for a cortical disruption (green arrow) and infiltration of the adjacent soft parts.

[Reference: Mother-child radiology department/ Hassan II university hospital of Fez]

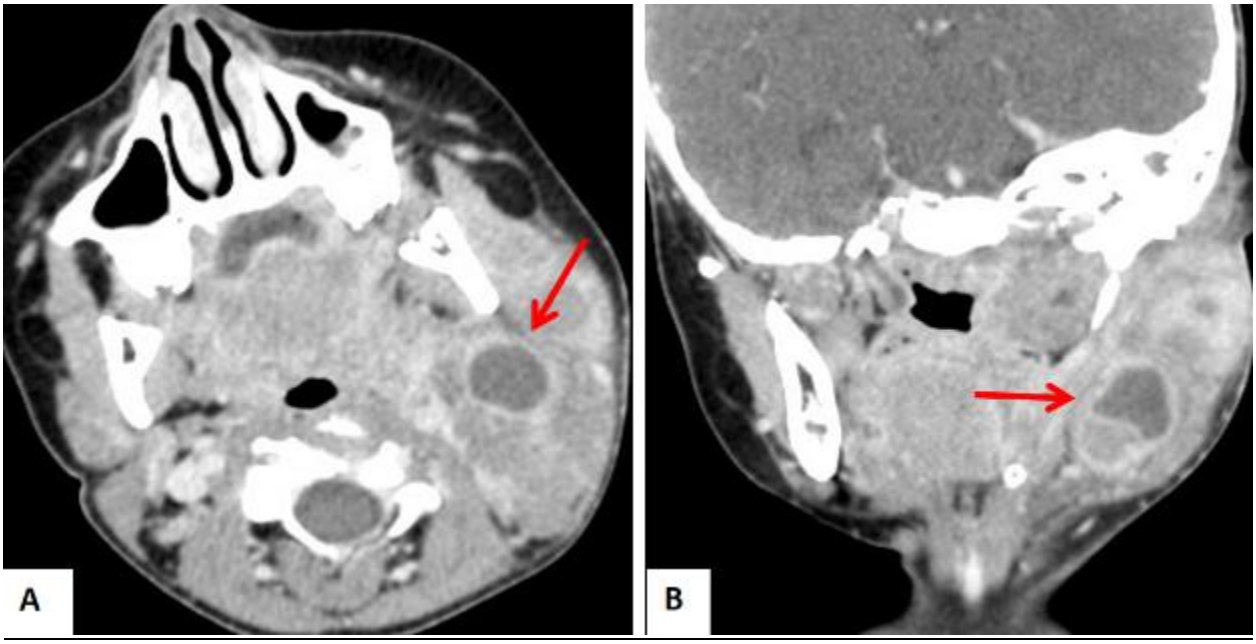

Figure 4 Craniofacial CT scan in axial (A) and coronal (B) sections with contrast agent administration: Large tumoral process (red arrow) centered on the left parotid, poorly limited, poly-lobed in outline, showing intense, heterogeneous enhancement, delineating areas of necrosis.

[Reference: Mother-child radiology department/ Hassan II university hospital of Fez]

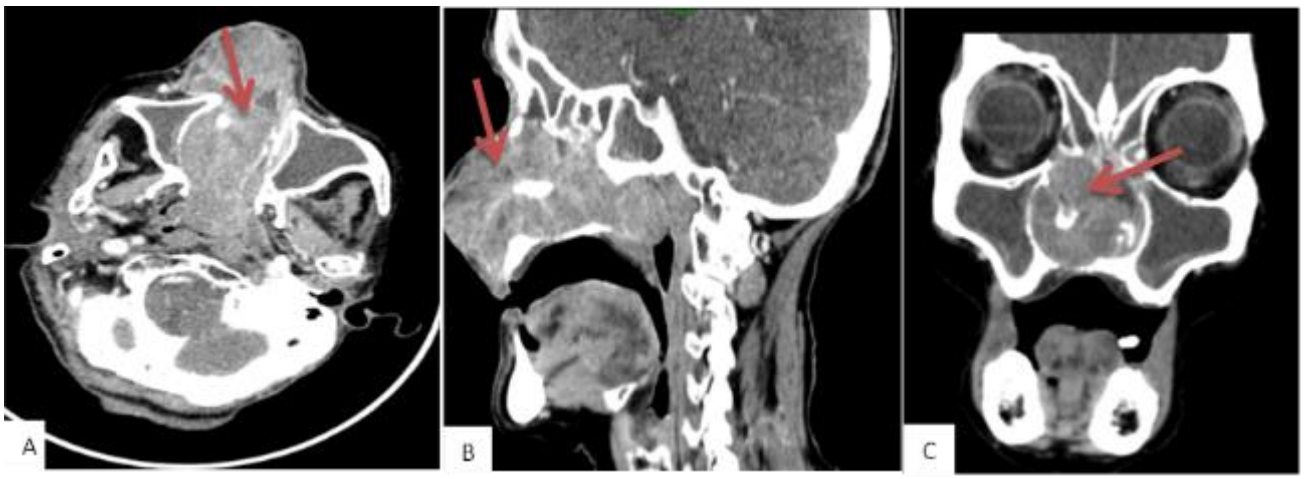

Figure 5 Facial CT scan after contrast injection, in axial sections(A),with sagittal (B) and coronal reconstructions(C) :Locally advanced lytic tissular mass centred on the nasal cavity (red arrow), with irregular margins, enhancing significantly and heterogeneously after contrast, with lysis of the nasal septum and the walls of the 02 maxillary sinuses, and extends towards the oropharynx, invading the soft palate inferiorly.

[Reference: Mother-child radiology department/ Hassan II university hospital of Fez]

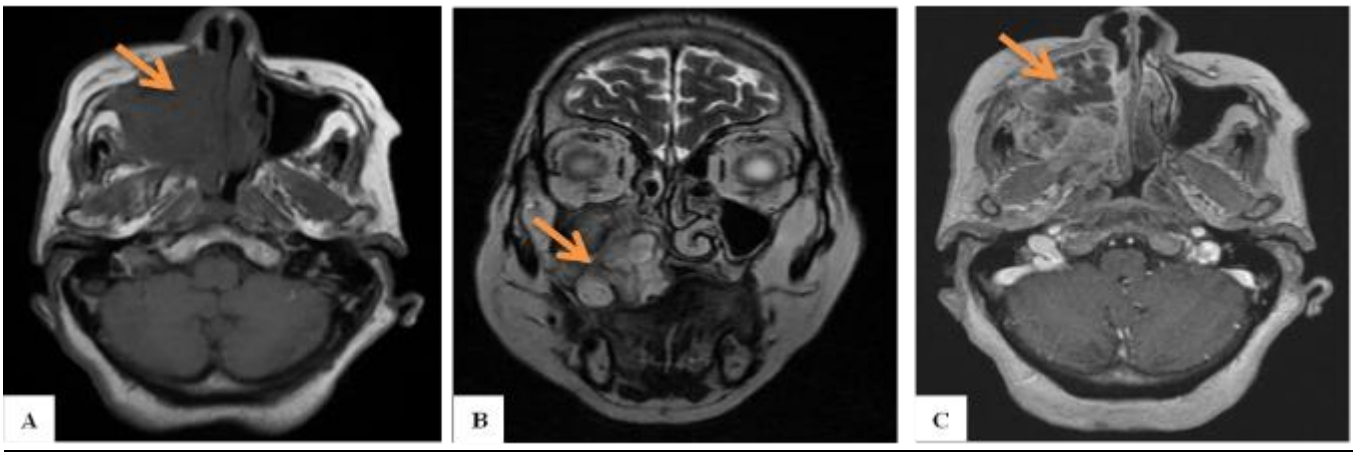

Figure 6 Facial MR images on axial T1-weighted (A), on coronal T2 -weighted (B) and axial T1 -weighted after injection of gadolinium (C) demonstrate voluminous process centered on the right maxillary sinus (orange arrow), multiloculated, poorly limited, exibiting a low T1 signal , with high signal T2 areas, heterogeneously enhanced after injection of Gadolinium.

[Reference: Mother-child radiology department/ Hassan II university hospital of Fez]

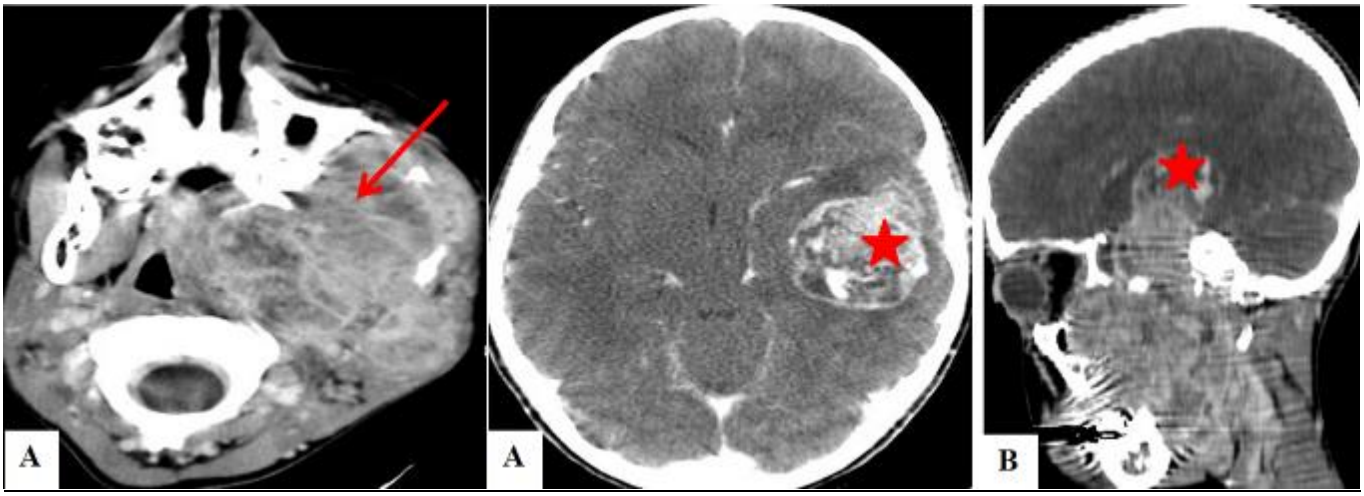

Figure 7 Craniofacial CT scan in axial sections (A), with sagittal (B) reconstructions after contrast injection Voluminous tumour process (Red arrow) centred on the left infra-temporal space, poorly limited, with irregular, lobulated contours, showing intense, heterogeneous enhancement after contrast, resulting in lysis of the ascending ramus of the mandible and the zygomatic arch laterally, as well as lysis of the temporal bone superiorly. It extends upwards towards the left temporal lobe (Red star).

[Reference: Mother-child radiology department/ Hassan II university hospital of Fez]

All patients underwent thoracoabdominal and pelvic CT scans as part of the extension workup. Of all cases, 15 patients (75%) had significant locoregional extension, with endocranial extension in 15% of patients (Figure 7) and bone lysis in 10%. Lymph node invasion occurred in 06 patients in our study. Five patients had pulmonary metastases at the time of diagnosis.

In all cases, diagnosis was made histologically based on biopsy. The most common histological type was embryonal rhabdomyosarcoma, detected in 06 patients (30%). The different histological types are shown in **Table 2.**

The therapeutic protocol developed for each patient was determined based on the nature of the tumor, its extent, and the feasibility of resection. Treatment aimed at cure in 11 cases, involving surgery with a margin of healthy resection. External radiotherapy was administered to 11 patients, accounting for 55% of the series. It was adjuvant to surgery in

9 cases and was combined with chemotherapy for nasopharyngeal location. The average administered dose was 67 Gray, with extreme values ranging from 60 to 70 Gy, depending on the quality of macroscopic and microscopic resection.

Table 2 Various histological types of head and neck sarcomas

Histological types	Number of patients (%)
Pleomorphic sarcoma	3 (15)
Chondrosarcoma	4 (20)
Leiomyosarcoma	1 (5)
Osteosarcoma	1 (5)
Embryonal rhabdomyosarcoma	6 (30)
Sarcomatoid carcinoma	1 (5)
Hemangio endotheliosarcoma	1 (5)
Sinonasal sarcoma	1 (5)
Alveolar rhabdomyosarcoma	2 (10)

The treatment was palliative in 5 cases. This involved tumors with pulmonary metastases in three cases and an inoperable tumor due to its extent in two cases. For these patients, the treatment consisted of palliative chemotherapy.

Clinical and radiological monitoring during and at the end of treatment was carried out for all patients who received treatment. After an average follow-up of 48 months, 06 patients were in complete remission (30%) **(Figure 09),** partial regression with stabilization was observed in 03 cases (20%) **(Figure 10).** Tumor recurrence **(Figure 08)** was observed in 02 cases, one of which was associated with bone metastasis. Progressive disease was noted in 04 cases. Two patients died from their tumors, and 3 patients were lost to follow-up.

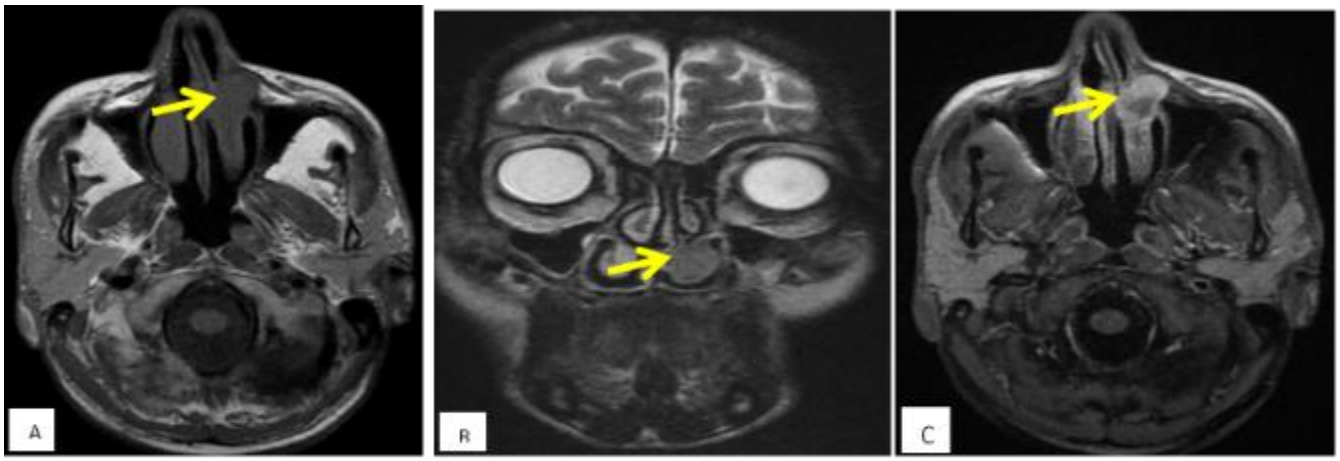

Figure 8 Facial MR images on axial T1-weighted (A), on coronal T2 -weighted (B) and axial T1 -weighted after injection of gadolinium (C): A history of surgically treated leiomyosarcoma of the nasal fossa.

Well-limited lobulated lesional process, in the anterior part of the left inferior turbinate (yellow arrow) presenting as iso-T1, hypo-T2 heterogeneously enhanced after contrast, in relation to a recurrence of leiomyosarcoma.

[Reference: Mother-child radiology department/ Hassan II university hospital of Fez]

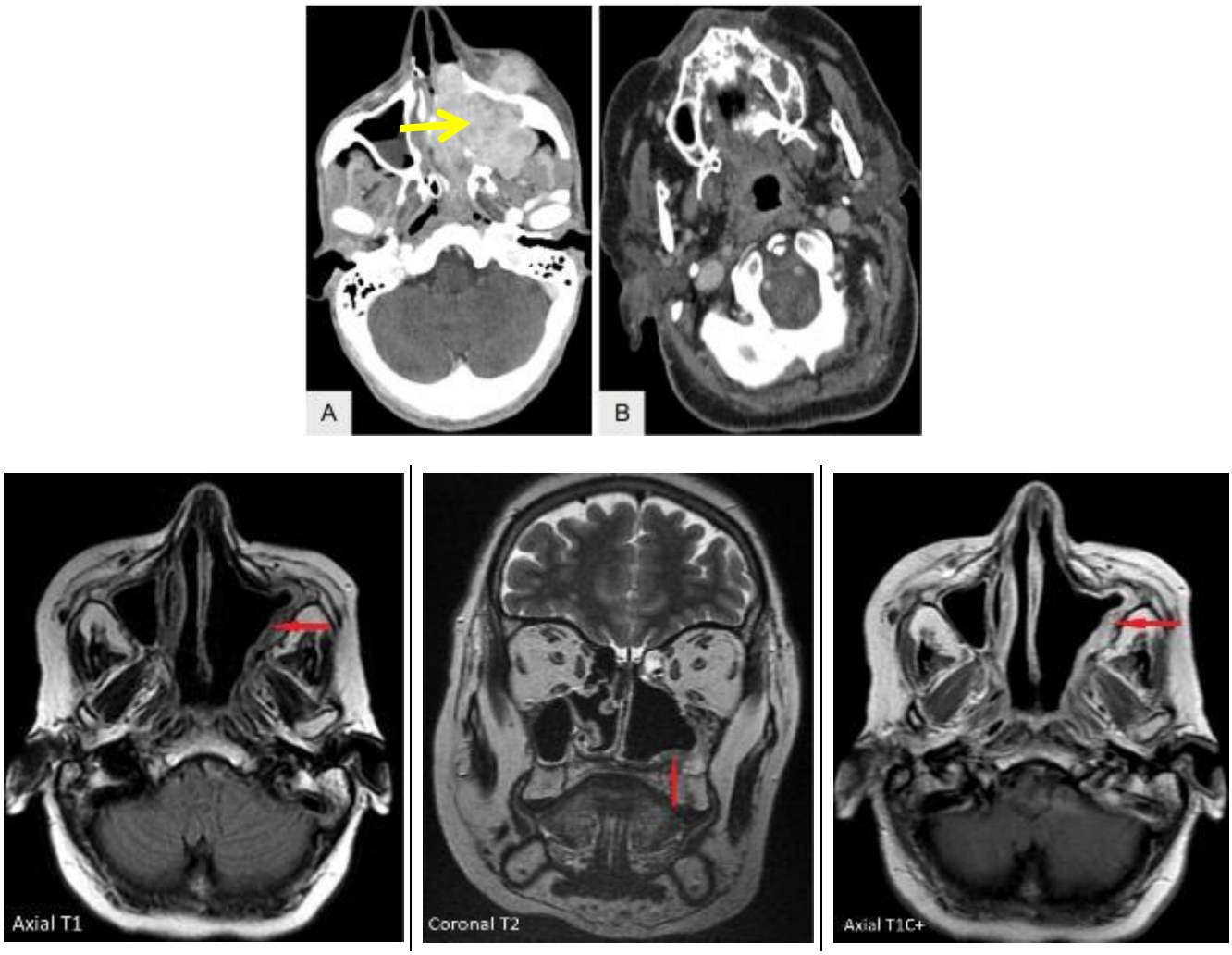

Figure 9 A: Contrast enhanced Facial CT axial section: tissue mass centered on the left maxillary sinus, with lobulated margins, heterogeneously enhanced after contrast (yellow arrow). B: Contrast enhanced Facial CT axial section : Absence of tumour residue in the left maxillary sinus region, with inflammatory thickening in the hard palate related to postoperative remodelling. Facial MR images : Tissue infiltrate centered on the left hard palate, presenting as low signal on T2 and T1, progressively enhanced after contrast, with regular, symmetrical thickening of the outer and anterior wall of the maxillary sinus, with a post-radiation appearance.

[Reference: Mother-child radiology department/ Hassan II university hospital of Fez]

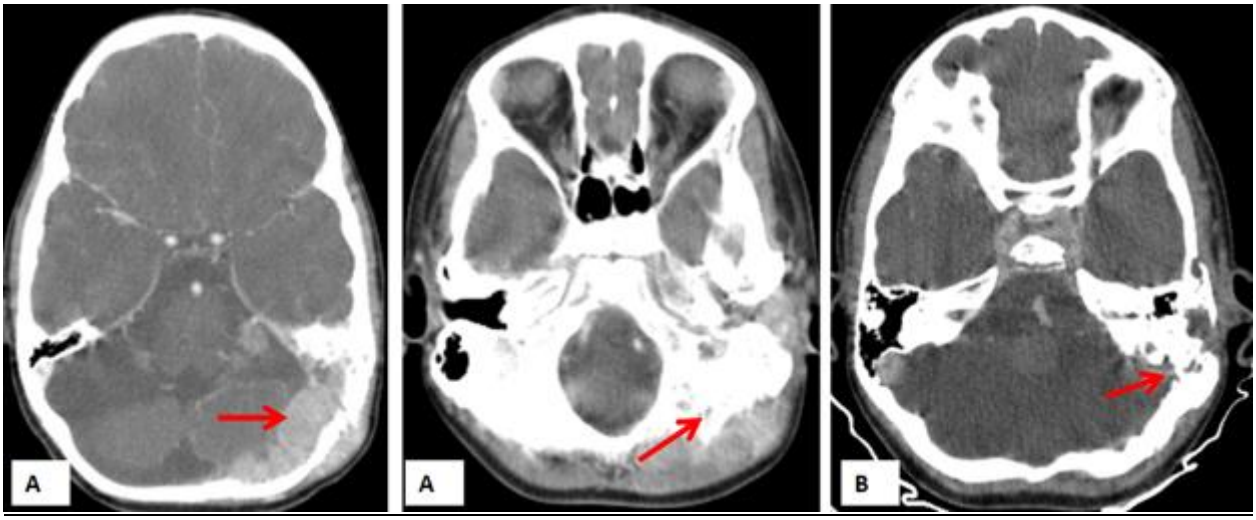

Figure 10 Contrast enhanced CT brain scan in axial sections before (A) and after (B) chemotherapy :

A: locally advanced tissue lesion process centered on the left mastoid extending into the homolateral cerebellar parenchyma. B: Clear regression of the previously described tissue lesion process.

[Reference: Mother-child radiology department/ Hassan II university hospital of Fez

4. Discussion

Soft tissue sarcoma is an uncommon disease, with an incidence of approximately 5 new instances annually per 100,000 individuals (1). While head and neck sarcomas exhibit significant diversity, they stand out as one of the less prevalent types, representing merely 1% of primary facial tumors (2) and comprising 4-10% of all sarcomas (3). Although soft tissue sarcomas can occur at any age, those affecting the head and neck tend to occur in young individuals (4). In our series, the average age was 33 years, with age ranging from 6 to 77 years.

Gender predominance varies from one series to another. In our series, a slight female predominance was observed (sex ratio 0.66).

Table 3 Illustrates the variation in average age and sex ratio in the literature

Table 3 Age and sex ratio in the literature

Authors	Number of cases	Average age (years)	Sex ration
Hammami (13)	15	38.4	0.66
Barosa (9)	29	45.9	2.22
Our series	20	33	0.66
Thomas Mucke (14)	74	Man / Woman 49 / 46.4	1.96
Van damne (15)	42	50	1.1

The etiopathogenesis of sarcomas is not completely understood (5), but several predisposing factors have been identified. Genetic diseases such as neurofibromatosis type I, Li-Fraumeni syndrome, Gardner syndrome, or Werner syndrome predispose individuals to certain sarcomas (6). Exposure to ionizing radiation or certain chemicals is associated with an increased incidence of sarcomas. Mark et al estimated the long-term risk of sarcoma in irradiated areas to be between 0.03 and 0.8% (7). In our series, the patient's medical history was unremarkable.

Clinically, the initial symptoms are nonspecific and depend on the location of the tumor. Most often, the tumor is discovered following the appearance of a mass, as was the case in our patients (40%). Sometimes, the tumor presents

with exophthalmos, nasal obstruction, facial pain, rhinorrhea, otalgia, and epistaxis. Stavrakas (8) made the same observation in his series where 80% of patients had a painless mass as the initial sign.

The location of the tumor is a crucial prognostic factor, as it determines the quality of the intervention. In our study, the most commonly affected sites were the maxillary sinuses (40% of cases), followed by the orbital cavity (20%), the nasal cavity (15%), and then, in equal percentage, the parotid, mandible, oral cavity, infratemporal fossa, and base of the skull.

In Stavrakas' series (8), the disease was most commonly located in the naso-sinusal region (23%), followed by the oral cavity (15.3%). The oropharynx, mandible, and salivary glands also frequently affected (10.2% each).

Regarding diagnosis, the radiological assessment involves a CT scan, often the initial imaging study for head and neck masses, and an MRI for comprehensive soft tissue evaluation. These imaging techniques are crucial for characterizing the mass, considering size, contour, architecture, vascularity, invasion of surrounding structures, or intracranial extension, and for ongoing monitoring. While there are no specific criteria for sarcoma, certain features may suggest this histology, including progressive disease, size exceeding 5 cm, subaponeurotic location, irregular contours, irregular intratumoral septa, presence of necrotic areas, heterogeneity on T1 and T2 MRI sequences, and intense contrast (9). To complement radiological evaluation, a thora-abdomino-pelvic CT scan is recommended to explore potential additional tumor locations. A definitive diagnosis requires an anatomopathological study of representative specimens, encompassing various tumor contingents.

The predominant category of head and neck sarcomas comprises soft tissue sarcomas, accounting for approximately 80%, while only 20% manifest as osseous or cartilaginous in nature (11). The World Health Organization (WHO) has recognized around fifty histological subtypes of sarcomas (12). In the head and neck region, pleomorphic sarcomas, angiosarcomas, fibrosarcomas, and malignant tumors of peripheral nerve sheaths are among the most frequently observed types (9).

The frequency of each histological type varies from one series to another. In Hammami's series (13), the most frequent histological type was rhabdomyosarcoma in 33%, followed equally by fibrosarcomas, chondrosarcoma, and leiomyosarcoma in 13.3%. In Barosa's series, osteosarcoma was the most frequent histological type, found in 34% of patients (10).

In our series, the most commonly found histological types were rhabdomyosarcoma (6 cases, 30% of the series), chondrosarcoma (4 cases, 20%), and pleomorphic sarcoma (3 cases, 15%).

Optimal treatment of patients with cervicofacial sarcomas should be performed in specialized units and multidisciplinary settings based on surgery, radiotherapy, and chemotherapy.

Treatment of cervicofacial sarcomas is primarily surgical, followed by adjuvant radiotherapy for sarcomas that are high stage, large in size, or with borderline or positive resection margins (14).

In our series, surgery was the most common treatment method. See Table 4 for details.

The role of chemotherapy in treating sarcoma is not well established. whether used as neoadjuvant or adjuvant treatment in combination with surgery, radiotherapy, or both. The chemotherapy protocol should be tailored to the specific subtype of sarcoma. Despite numerous trials, there is no evidence to suggest that chemotherapy improves overall survival in sarcoma patients (13).

Surgery and radiotherapy remain the most widely used therapeutic methods, while chemotherapy plays a less significant role in the treatment of head and neck sarcomas.

Prognostic factors for cervicofacial sarcomas include high histological malignancy grade, tumor size exceeding 5 cm, local extension to skin, bone, or major neurovascular structures, and positive surgical margins.

These factors are correlated with a high rate of local recurrence, distant therapeutic failure (tumor progression), and a reduced survival rate.

The overall survival rate for five years ranges from 44% to 80% (16). This variability is due to the heterogeneity of these tumors and the lack of standardization of treatment modalities.

Table 4 Comparative table of the number of patients benefiting from the different therapeutic modalities

	Surgery		**Radiotherapy**		**Chemotherapy**	
Our series	12	60%	11	55%	7	35%
Van Damne(15)	39	93%	18	43%	10	24%
Barosa(10)	24	83%	18	62%	14	48%
Hammami(13)	7	47%	8	53%	4	27%

5. Conclusion

Haed and neck sarcomas are a rare and aggressive tumor that are highly heterogeneous. Imaging is crucial for early diagnosis of these tumors at the primary or metastatic stage, guiding appropriate treatment.

Establishing an early diagnosis and intervention remains the ideal approach to increase the chances of cure.

Compliance with ethical standards

Disclosure of conflict of interest

The authors declare that they have no competing interests

Statement of informed consent

Written informed consent was obtained for all patients. Anonymity and confidentiality were respected for all participants.

Availability of data and material

The datasets used and/or analysed during the current study are available from the corresponding author on reasonable request.

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