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Review

Head and neck soft-tissue sarcoma in adults

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ABSTRACT

Adult soft-tissue sarcoma is rare but aggressive, with incidence around 5 per 100,000 per year. Head and neck locations are infrequent. Genetic disease and irradiation are risk factors. The diagnosis needs to be known in order to avoid treatment delay. There are about 50 histologic subtypes, with different patterns and prognoses. Pathologic review and the development of molecular techniques are therefore essential. Prognosis in adult head and neck soft-tissue sarcoma (HNSTS) is poor: 5-year overall survival, about 60%. Recurrence is most often local. Prognostic factors are: tumor size and local extension, histologic grade and margin status. There are few targeted management guidelines. Surgical resection with negative margins is the primary treatment. Postoperative radiation therapy can improve prognosis. The role of chemotherapy is not well established. HNSTS should be treated in a reference center, with multidisciplinary staff following national network guidelines. Several factors are still unknown. The purpose of this article is to summarize the state of knowledge in adult HNSTS.

1. Introduction

Head and neck soft-tissue sarcoma (HNSTS) is rare, aggressive and heterogeneous. There have been few large series and treatment guidelines therefore fail to target HNSTS specifically. Several factors remain unknown or are under assessment. The present article updates the state of knowledge, studying anatomopathologic, diagnostic, prognostic and therapeutic aspects.

2. Discussion

2.1. Epidemiology and diagnosis

Soft-tissue sarcoma is rare, with annual incidence around 5 per 100,000 [1].

Head and neck locations are the rarest [1,2], at 5–15% of sarcomas and about 1% of malignant head and neck tumors in adults [3,4].

There are certain known risk factors. Genetic diseases such as Li-Fraumeni syndrome or type-1 neurofibromatosis may induce certain sarcomas; genomic abnormalities, and notably specific reciprocal translocations, are frequently reported on molecular analysis [5]. Irradiation is associated with elevated incidence [6]; Mark et al. estimated the long-term risk in irradiated body areas at 0.03 and 0.8%, with onset at a mean 12 years [7]. Diagnosis and causality are hard to establish, with criteria of progression over time; the terms "radiation-induced sarcoma" and "sarcoma in irradiated tissue" are a matter of discussion [8,9].

There are some 50 histologic subtypes of sarcoma, classified by the WHO [10]. In the head and neck, the most frequent forms, taking all series together, are pleomorphic sarcoma (or malignant fibrous histiocytoma: MFH), fibrosarcoma, angiosarcoma, malignant peripheral nerve sheath tumor and non-classified/non-differentiated sarcoma [3,11–19] (Fig. 1).

Given the anatomic and functional specificities of the head and neck region, tumor site is an important therapeutic decision factor, influencing surgical options, the feasibility of negative margins, and functional and esthetic prognosis. The most frequent sites are superficial areas of the neck and parotid, nasal sinuses and cavities, and lastly the visceral spaces of the neck (pharynx, larynx) [3,11–21] (Fig. 2).

Diagnostic imaging is on CT and MRI. Although there are no specific criteria for sarcoma, certain points may be suggestive: progressive lesion, >5 cm, sub-aponeurotic, irregular contours, irregular intra-tumoral septa, necrotic areas, heterogeneity on T1- and T2-weighted MRI, and prolonged intense contrast uptake. Fine-needle aspiration cytology is not recommended, and biopsy should adhere to precise rules. There is no role for diagnostic frozen-section biopsy [22]. Pathology seeks to establish diagnosis of malignant mesenchymatous tumor and to classify and grade the

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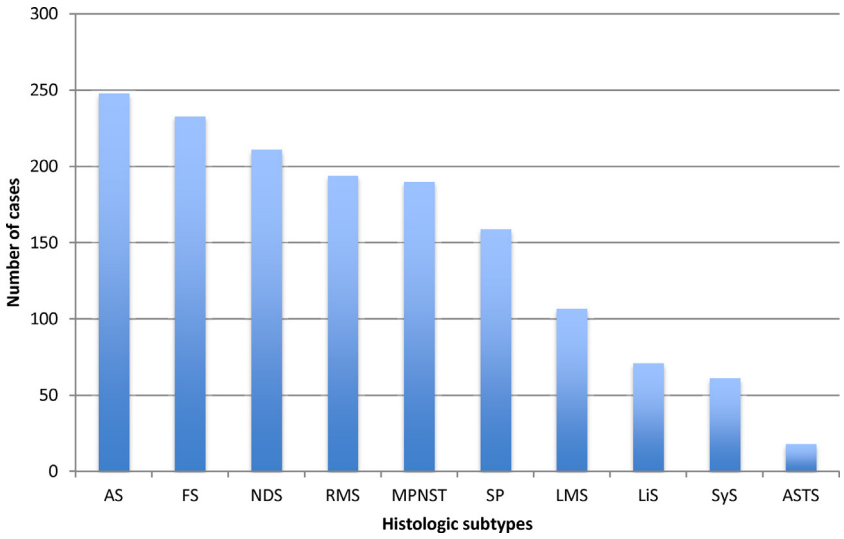

Fig. 1. Histologic subtypes: 1579 cases of HNSTS [3,11–19]. AS: angiosarcoma; FS: fibrosarcoma; NDS: unclassified/non-differentiated sarcoma; RMS: rhabdomyosarcoma; MPNST: malignant peripheral nerve sheath tumor; SP: pleomorphic non-differentiated sarcoma (formerly; MFH); LMS: leiomyosarcoma; LiS: liposarcoma; SyS: synovial sarcoma; ASTS: alveolar soft-tissue sarcoma.

sarcoma. Histologic diagnosis of sarcoma is difficult, due the rarity and great histologic heterogeneity of the entity. Cross-reading by different pathologists can make diagnosis more robust [23]. Several histologic grading systems have been described; that of the French Federation of Cancer Centers Sarcoma Group (FFCCSG) seems to be the most effective in predicting overall survival and onset of metastasis [24].

Chest CT is recommended for extension assessment [2].

The most widely used clinical classification is that of the American Joint Committee on Cancer (AJCC) Union for International Cancer Control (UICC), with 3 grades summarizing primary size and extension, regional lymph-node invasion, metastasis and sarcoma grade.

2.2. Survival and prognostic factors

Local control at 5 years is the principal assessment criterion, as recurrence is most often local in HNSTS [19].

Five-year local control varies greatly between series, from 47 to 78% [3,11–13,15,25,26]. As the pathology is rare, some series are old and did not benefit from more recent surgical and oncological progress; large series sometimes adopted long inclusion periods. Inclusion criteria sometimes allowed tumors of differing natural history and prognosis, such as desmoid tumor, solitary fibrous tumor or dermatofibrosarcoma protuberans. Onset of local recurrence is usually within 24 months [13].

Ten to 40% of HNSTS patients develop remote metastases during follow-up, mainly in the lung [19,27]. Series reporting high rates of metastasis, such as that of Tejani et al., with 37%, mainly included high-grade sarcomas [27].

Overall survival in HNSTS is poorer than for other locations. This may partly be due to the distribution of histologic subtypes, with more frequent angiosarcoma and fibrosarcoma, and to the anatomic difficulty of ensuring negative margins in the head and neck. Five-year overall survival ranges from 31 to 80%, for a mean around 60%, versus around 80% for trunk and limb sarcoma [3,11–13,15–19,26,28–33] (Table 1). The inclusion of pediatric cases in some series and reclassification to intermediate malignancy categories may explain differences in results [10]. Finally, the low incidence of sarcoma means that most large series covered cases arising over several decades, with the consequent changes in treatment modalities.

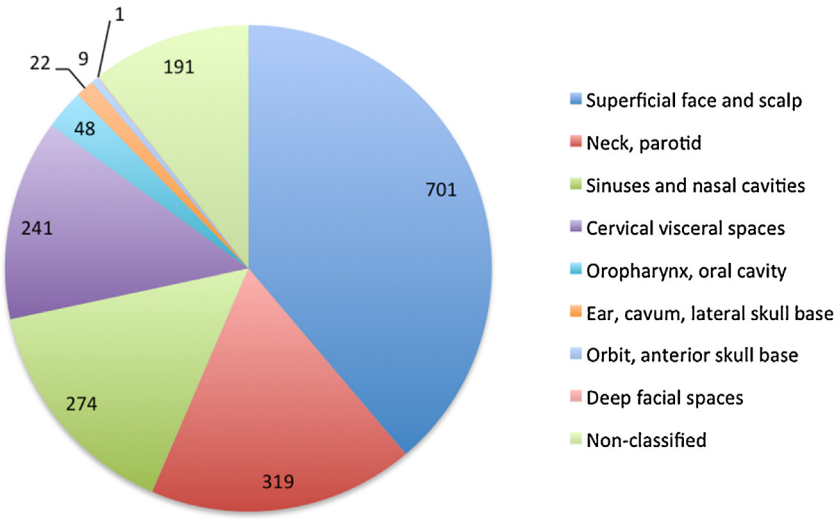

Fig. 2. Tumor locations in main published series [3,11–21].