Journal of Visceral Surgery (2015) 152, 223—230

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REVIEW

Soft tissue sarcoma in France in 2015: Epidemiology, classification and organization of clinical care

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Available online 15 June 2015

KEYWORDS
Sarcoma;
Epidemiology;
Classification;
Recommendation;
Network

Summary Four thousand new cases of soft tissue sarcomas are diagnosed each year in France, 23% of which are localized in the abdomen and pelvis; the treatment of non-metastatic tumor is based on wide surgical resection, the quality of which determines the long-term outcome. To ensure appropriate care, the European Society of Medical Oncology (ESMO) recommends that any patient with an unexplained soft tissue mass (of any size for deep lesions or of >5 cm for superficial lesions) be referred to a specialized center with capacities for multidisciplinary team decision; appropriate imaging should be performed prior to treatment and a percutaneous image-guided needle biopsy should be routinely performed. In France, clinical and pathology networks (NetSarc and RRePS) currently offer patients a structured means to make a systematic diagnosis of soft tissue sarcoma and help to provide access to appropriate treatment in a specialized center.

Introduction

Sarcomas are rare malignant tumors of mesenchymal origin, that arise in connective tissue, in contrast to the more frequent and better-known carcinomas of epithelial origin [1]. Sarcomas have widely diverse pathologies with more than 70 histological subtypes and an ever-Increasing number of molecular subtypes. They may develop at any age including childhood, can occur anywhere anatomically from head to foot, and are of varying aggressiveness, even within the same histological subtype [2,3]. There are three principal kinds of sarcoma corresponding to different clinicopathological entities with individually specific progression, and specifically different management strategies: bone sarcomas, visceral

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http://dx.doi.org/10.1016/j.jviscsurg.2015.05.001
1878-7886/

sarcomas that develop in a specific organ (the most typical being gastrointestinal stromal tumors [GIST]), and soft tissue sarcomas (STS) arising in connective tissue and extraosseous connective tissue; these represent about 1% of all adult cancers [4—6]. No formal etiology has so far been defined, but several contributing factors have been identified (genetic mutations of the NF1RB1, WRN, p53, and APC genes, which are responsible respectively for type I neurofibromatosis, congenital retinoblastoma, and the syndromes of Li-Fraumeni, Gardner, and Werner) or extrinsic genetic damage (ionizing radiation, exposure to vinyl chloride, dioxin, chlorophenol, and certain viruses) [2—7]. This update aims to clarify recommendations for the diagnostic and therapeutic management of STS, which are infrequently encountered and poorly understood by most visceral surgeons.

Epidemiology

The exact annual incidence of STM is unknown. Several estimates based on retrospective analyses of cancer registries have been attempted [8—17]. These studies all suffer from methodological bias because the registries were set up to collect data based on the organ of origin, an appropriate methodology for the natural history of carcinoma but unsuitable for sarcomas that may arise in any part of the body. This is particularly the case for visceral sarcomas, which tend to be misclassified as digestive cancers based on the organ in which they arise. Adult registries are often separate from pediatric registries. This results in a systematic underestimation of the incidence of STS [8—17]. In addition, when a pathologist who is unfamiliar with these histological types of tumor performs the pathologic analysis, the risk of initial diagnostic error ranges from 10 to 25% [13—17]. The best estimates of incidence available today come from a French study; the authors, fully aware of diagnostic pitfalls, used a less biased methodology based on a systematic prospective re-analysis of all tumor specimens where a formal diagnosis or suspicion of sarcoma had been made, over a period of two years from 158 public and private practice pathologists in the Rhône-Alpes region of France. Tissue specimens were reviewed by two expert pathologists with additional systematic molecular analysis, and all samples were reclassified according to the 2002 WHO classification [18]. After review of 1287 tissue blocks, sarcoma was definitely diagnosed in 748 patients between 2005 and 2007 in an area with a population of about 6 million people. The overall and age-adjusted incidence of sarcoma was estimated at 6.2 and 4.8 cases per 100,000 population per year, respectively. The incidence of STS and visceral sarcoma were respectively 3.6 and 2.0 cases per 100,000 population per year [18]. The overall male to female ratio was 1.1/1, but there was a female preponderance of visceral sarcomas (1.4/1) and a male preponderance for STS (1.3/1). The median age at diagnosis was 60 years, with a range between 0 and 92 years. Eight percent of patients developed sarcoma before the age of 18, and 28% after the age of 70 years. A graphical representation of the evolution according to age of the incidence of STS is shown in Fig. 1. The median size of the lesion was 6 cm, with extremes ranging from 0.3—40 cm. Localization of STS was truncal in 40% of cases (17% thoracic, 9% retroperitoneal, 8% pelvic and 6% abdominal), while 60% of STS were peripheral (49% localized on a limb and 11% on the head and neck). Of the 433 diagnosed cases of STS, 25 (5.8%) arose in irradiated tissues [18].

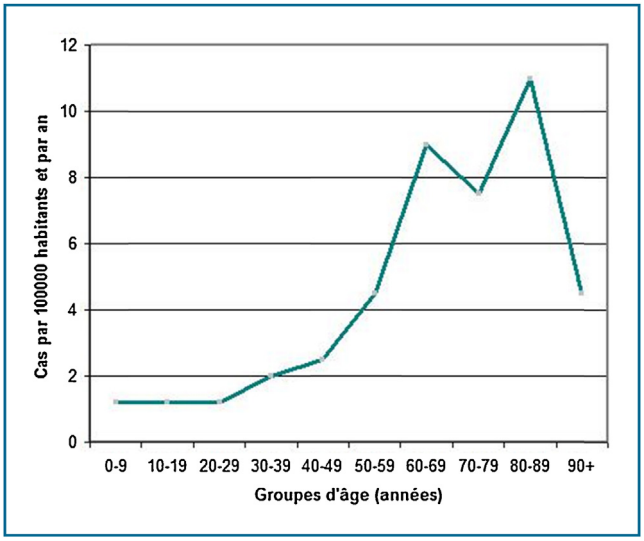

Figure 1. Incidence of soft-tissue sarcoma as a function of age in France. Cases per 100,000 inhabitants per year; age groups (years).

Table 1 Distribution of the principal histologic subtypes of soft-tissue sarcoma in France (2002 WHO Classification).

	n	%
Sarcomas		
Liposarcoma	1092	25.2
Undifferentiated sarcoma	947	21.8
Leiomyosarcoma	741	17.1
Myxofibrosarcoma	252	5.8
Angiosarcoma	219	5.0
Rhabdomyosarcoma	215	5.0
Synovial sarcoma	183	4.2
MPNST	115	2.6
Other	577	13.3
Mesenchymal tumors of intermediate malignancy		
Solitary fibrous tumor	119	
Desmoid tumor	363	

MPNST: malignant peripheral neural sheath tumors.

Extrapolation of these data to the overall French population led the authors to estimate that about 4000 new cases of STS were diagnosed annually in France [18]. An estimation of the distribution of histologic sub-types is illustrated in Table 1, based on the findings of the Network for Pathologic Registry of Sarcomas (RRePS), which has undertaken the systematic histopathologic review of all newly diagnosed cases of sarcoma, GIST, and desmoid tumors [19,20].

Classification of soft tissue sarcomas

Correct classification of STS is imperative from the very beginning of management. It informs and guides the