Epidemiology and Etiology of Sarcomas

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KEYWORDS

• Sarcoma • Epidemiology • Etiology • Li-Fraumeni • Radiation

KEY POINTS

- Sarcomas are rare malignant tumors of mesenchymal origin, accounting for less than 1% of all new cancer diagnoses.
- The extremity (particularly the thigh) is the most common location for soft tissue sarcoma. Bone sarcomas are rare and are more commonly seen in the pediatric population.
- Most sarcomas are sporadic and idiopathic, with no associated inherited genetic defect or environmental factor identified as the cause.
- Genetic syndromes associated with sarcoma development include Li-Fraumeni syndrome, retinoblastoma, neurofibromatosis type 1, and familial adenomatous polyposis syndrome.
- Ionizing radiation is strongly linked to subsequent development of bone sarcoma and soft tissue sarcoma. In cases of prior radiation therapy, the secondary sarcoma develops within the radiation field.

INTRODUCTION

Sarcomas make up a broad group of malignant neoplasms of mesenchymal origin. More than 70 histologic subtypes have been identified. However, sarcomas can be classified into 2 broad categories: (1) soft tissue sarcomas (STS), and (2) sarcomas of the bone. In the former group, sarcomas that have histologic resemblance to fat, muscle, nerve sheath, and blood vessels are included and are named accordingly.

EPIDEMIOLOGY

Sarcomas are rare, making up less than 1% of all new cancer diagnoses. There will have been an estimated 1.66 million new cancer diagnoses in 2015 in the United States, of which, only 11,930 cases will have been STS, and 2970 cases, bone sarcomas.[1]

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Soft Tissue Sarcoma

According to the Surveillance, Epidemiology, and End Results Program of the National Cancer Institute, the incidence of STS is approximately 3.4 per 100,000.[2] The true incidence of STS is likely somewhat underestimated, as some visceral sarcomas are likely counted with their organ of origin rather than with STS. There is a slight male preponderance of 1.4:1.[2] The median age at diagnosis is 59,[2] with a bimodal distribution that peaks in the fifth and eighth decades.[3]

STS occur most commonly on the extremities; upper and lower extremity STS account for 12% and 28%,[4] respectively, of all STS. The thigh is the most common site of STS, accounting for 44% of all extremity STS.[4] The most common type of extremity STS is liposarcoma (LPS).[4] Visceral STS account for 22% of all STS[4] and include gastrointestinal stromal tumors (GIST) and uterine leiomyosarcoma (LMS). GISTs are most commonly located in the stomach (59%), followed by small intestine (31%), with rectal (3.3%), colonic (2.7%), and esophageal (0.6%) locations being rare.[5] The median age at diagnosis for GIST is 62.[5] Retroperitoneal sarcomas account for 16% of all STS, whereas trunk and other sites (including the head and neck) account for 10% and 12%, respectively.[4] Retroperitoneal sarcomas are typically LPS and LMS.

Overall, LPS is the most common type of STS, accounting for approximately 20% to 25% of all STS.[2,4,6] LPS can be further subdivided into well-differentiated LPS (also called *atypical lipomatous tumor*), dedifferentiated LPS, myxoid LPS, and pleomorphic LPS.[7] Other common STS histologic subtypes include LMS (14%) and undifferentiated pleomorphic sarcoma (14%),[4] formerly known as *malignant fibrous histiocytoma*. The histologic distribution of STS among the various sites is found in **Fig. 1**.

Bone Sarcoma

Bone sarcomas are even more uncommon, accounting for 0.2% of all new cancer diagnoses.[2] This disease tends to affect the younger population, most frequently diagnosed in those 20 years or younger.[2] The age at diagnosis also varies with the histologic subtype. Osteosarcoma is the most common bone sarcoma overall and is more frequently seen in adolescents than in adults.[8] Similarly, Ewing sarcoma is more common in children and adolescents[8] but can also be seen in adults. The median age at diagnosis is 15.[8] Although any bone (or even soft tissue) can be involved,

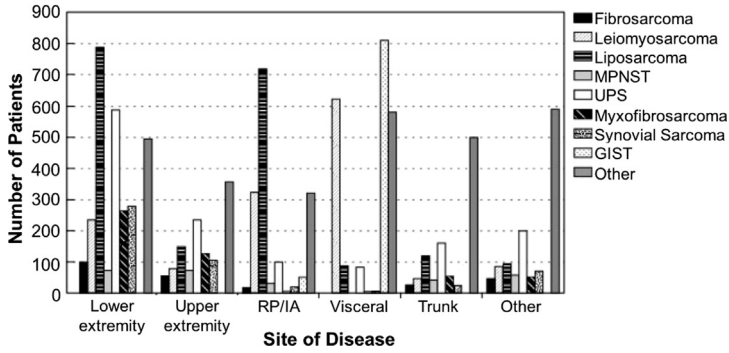

Fig. 1. Histopathology of soft tissue sarcomas by site of disease, N = 10,000. IA, intraabdominal; RP, retroperitoneum; UPS, undifferentiated pleomorphic sarcoma. (*From* Brennan MF, Antonescu CR, Moraco N, et al. Lessons learned from the study of 10,000 patients with soft tissue sarcoma. Ann Surg 2014;260(3):419; with permission.)