Referral patterns, prognostic models and treatment in soft tissue sarcomas

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Chapter 8

Angiosarcoma

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The term angiosarcoma applies to a range of malignant endothelial vascular neoplasms, mimicking the morphologic and functional features of normal endothelium, which can affect a variety of sites. Soft tissue angiosarcoma consists for the majority out of cutaneous tumors, and less than one quarter present as deep soft tissue tumor. [1] Angiosarcomas usually occur in adulthood, with a peak incidence in the seventh decade. The tumor is very uncommon and has a low prevalence worldwide. Several conditions are associated with the development of angiosarcomas, including neurofibromatosis (NF1) [2,3], adjacent to synthetic vascular grafts [4] and following radiation therapy.

Among angiosarcomas, the location at the breast forms a special subgroup. Two main groups are described: primary angiosarcomas and secondary angiosarcomas of the breast. Primary angiosarcomas account for less than 1% of all breast malignancies [5] and their peak incidence is in the third and fourth decade of life. Secondary angiosarcomas are further defined into two subgroups: angiosarcomas following longstanding lymphedema, known as Stewart Treves syndrome, and angiosarcomas following radiation therapy. Stewart Treves syndrome can develop in the lymph edematous arm as a consequence of breast cancer treatment with axillary nodal dissection and was firstly described by Stewart and Treves in 1948. [6] Since the treatment for early stage breast cancer has changed to a more conservative approach with lumpectomy and adjuvant radiation, less cases of Stewart Treves have been observed and more angiosarcomas in the radiated field. Patients treated for breast cancer with radiation have a five fold higher risk for angiosarcoma than patients not receiving radiation. [7] The clinical presentation differs between these two subgroups; angiosarcomas following radiation develop earlier and have shorter symptom duration than the Stewart Treves tumors. [8] Radiation associated angiosarcomas of the breast will be further described in the next chapter.

A systematic review published in 2012, reported all literature about radiation associated sarcoma since 1970. [9] Focusing on the radiation associated sarcoma of the breast, the incidence is particularly low considering the long time span of the studies. The radiation dose to the breast as part of the conservative breast cancer treatment was median 53Gy. Previous studies have shown a dose related

risk of radiation induced sarcomas, starting upon as much as 14Gy. [10,11] The age at the time of diagnosis of radiation associated sarcoma was median 68 years, and the latency period between the treatment of breast cancer and the diagnosis of radiation induced sarcoma was median 9 years. The relative long latency period at which radiation associated sarcoma of the breast develop implies a long follow up in the hospital or at least a good self examination by the patient.

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