

Malignant transformation of a benign soft tissue tumor is extremely rare, with the exception that malignant peripheral nerve sheath tumors (neurofibrosarcoma, malignant schwannoma) can arise from neurofibromas in patients with neurofibromatosis. Several etiologic factors have been implicated in soft tissue sarcomas.

Environmental Factors Trauma or previous injury is rarely involved, but sarcomas can arise in scar tissue resulting from a prior operation, burn, fracture, or foreign body implantation. Chemical carcinogens such as polycyclic hydrocarbons, asbestos, and dioxin may be involved in the pathogenesis.

Iatrogenic Factors Sarcomas in bone or soft tissues occur in patients who are treated with radiation therapy. The tumor nearly always arises in the irradiated field. The risk increases with time.

Viruses Kaposi's sarcoma (KS) in patients with HIV type 1, classic KS, and KS in HIV-negative homosexual men is caused by human herpesvirus (HHV) 8 ([Chap. 195](#)). No other sarcomas are associated with viruses.

Immunologic Factors Congenital or acquired immunodeficiency, including therapeutic immunosuppression, increases the risk of sarcoma.

■ GENETIC CONSIDERATIONS



Li-Fraumeni syndrome is a familial cancer syndrome in

which affected individuals have germline abnormalities of the tumor-suppressor gene *p53* and an increased incidence of soft tissue sarcomas and other malignancies, including breast cancer, osteosarcoma, brain tumor, leukemia, and adrenal carcinoma ([Chap. 71](#)). Neurofibromatosis 1 (NF-1, peripheral form, von Recklinghausen's disease) is characterized by multiple