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NCCN Clinical Practice Guidelines in Oncology
(NCCN Guidelines®)

Soft Tissue Sarcoma

Overall management of Soft Tissue Sarcoma is described in the full NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®) for Soft Tissue Sarcoma. Visit NCCN.org to view the complete library of NCCN Guidelines®.

NCCN National Comprehensive Cancer Network®

SYSTEMIC THERAPY AGENTS AND REGIMENS WITH ACTIVITY IN SOFT TISSUE SARCOMA SUBTYPES AND AGGRESSIVE SOFT TISSUE NEOPLASMS

Inflammatory Myofibroblastic Tumor (IMT) with Anaplastic Lymphoma Kinase (ALK) Translocation	Malignant Perivascular Epithelioid Cell Tumor (PEComa) (for locally advanced unresectable or metastatic disease)	Recurrent Angiomyolipoma, Lymphangioleiomyomatosis
Preferred regimens • Alectinib[87] • Brigatinib[88,89] • Ceritinib[90] • Crizotinib[91] • Lorlatinib[92]	**Preferred regimens** • Albumin-bound sirolimus[93,94] **Other recommended regimens** • Sirolimus[95-98] • Everolimus[99] • Temsirolimus[100,101]	**Preferred regimens** • Sirolimus[95-98] • Everolimus[99] • Temsirolimus[100,101]

Footnotes and references see SARC-G, 7 of 12

Note: All recommendations are category 2A unless otherwise indicated.
Clinical Trials: NCCN believes that the best management of any patient with cancer is in a clinical trial. Participation in clinical trials is especially encouraged.

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