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Disease
Inflammatory myofibroblastic tumors.

Clinics
Rare soft tissue tumor found in children and young adults.

Pathology
Spindle cell proliferation with myofibroblastic differentiation and an inflammatory infiltrate.

Prognosis
Low malignant potential and good prognosis.

Genes involved and proteins

ALK
Location
2p23
Protein
1620 amino acids; 177 kDa; glycoprotein (200 kDa mature protein); membrane associated tyrosine kinase receptor.

TPM4 (tropomyosin fibroblast, non muscle type)
Location
19p13.1
Protein
248 amino acids, 29 kDa; coiled coil structure

Result of the chromosomal anomaly

Hybrid Gene
Description
5’ TPM4 - 3’ ALK

Fusion Protein
Description
783 amino acids. N-term TPM4 containing all but its 27 C-term amino acids, including the coiled coil domain, fused to the 562 C-term amino acids from ALK (i.e. the entire cytoplasmic portion of ALK with the tyrosine kinase domain).

Expression / Localisation
Predominantly cytoplasmic.

Oncogenesis
TPM4-ALK is continuously activated.

References


This article should be referenced as such: