

Solid Tumour Section

Short Communication

t(2;2)(p23;q13)

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Clinics and pathology

Disease

Inflammatory myofibroblastic tumors (IMT).

Clinics

Rare soft tissue tumour found in children and young adults.

Pathology

Spindle cell proliferation with myofibroblastic differentiation and an inflammatory infiltrate; staining of ALK-positive cases, representing 60% of IMT cases, is usually confined to the cytoplasm, but it showed a nuclear membran pattern in one of the two t(2;2) available cases.

Prognosis

Inflammatory myofibroblastic tumors have a low malignant potential and good prognosis; however, the 2 cases with a t(2;2) showed (relatively) aggressive clinical course with tumor recurrence and round cell transformation.

Genes involved and proteins

ALK

Location

2p23

Protein

1620 amino acids; 177 kDa; glycoprotein (200 kDa mature protein); membrane associated tyrosine kinase receptor.

RANBP2

Location

2q13

Protein

3224 amino acids; nuclear pore protein involved in nuclear export.

Result of the chromosomal anomaly

Hybrid Gene

Description

5' RANBP2 - 3' ALK

Fusion Protein

Description

1430 amino acids; 6867 N-term amino acid from RANBP2, fused to the 562 C-term amino acids from ALK (i.e. the leucine-rich domain, including the leucine zipper, of RANBP2 and the entire cytoplasmic portion of ALK with the tyrosine kinase domain).

Oncogenesis

The leucine zipper of RANBP2 could mediate oligomerization of RANBP2-ALK.

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