

## Solid Tumour Section

### Short Communication

# t(1;2)(q25;p23)

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## Clinics and pathology

### Disease

Inflammatory myofibroblastic tumors.

### Clinics

Rare soft tissue tumour found in children and young adults.

### Pathology

Spindle cell proliferation with myofibroblastic differentiation and an inflammatory infiltrate.

### Prognosis

Low malignant potential and good prognosis.

### Disease

Anaplastic large cell lymphoma: translocations involving 2p23 are found in more than half cases of anaplastic large cell lymphoma (ALCL), a high grade non Hodgkin lymphoma (NHL). They involve ALK, and are therefore called ALK+ ALCL.

The most frequent ALK+ ALCL being the the t(2;5)(p23;q35) with NPM1 -ALK fusion protein, which localises both in the cytoplasm and in the nucleus.

The t(1;2)(q25;p23) is very rare., and, like other t(2;Var) involving various partners and ALK, the fusion protein has a cytoplasmic localization; they are therefore called "cytoplasm only" ALK+ ALCL.

### Epidemiology

A very few (four) cases known so far.

### Clinics

ALK+ ALCL without the t(2;5) (so called cytoplasmic only ALK cases) show clinical features similar to those of classical ALK+ ALCL: young age, male predominance, presentation with advanced disease, systemic symptoms, frequent involvement of

extranodal sites, and a good prognosis. Nothing in particular is known concerning t(1;2) cases, as cases are not documented.

### Cytogenetics

Complex karyotypes and/or hidden translocation in the 2 cases with cytogenetic data; FISH analyses are essential.

## Genes involved and proteins

### TPM3 (tropomyosin alpha chain)

#### Location

1q25

#### Protein

284 amino acids, 33 kDa; coiled coil structure; role in Calcium dependant actin-myosin interaction.

### ALK

#### Location

2p23

#### Protein

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

## Result of the chromosomal anomaly

### Hybrid Gene

#### Description

5' TPM3 - 3' ALK.

### Fusion Protein

#### Description

104 kDa ; 221 (?) N-term amino acids from TPM3 fused to the 562 C-term amino acids from ALK (i.e. the

entire cytoplasmic portion of ALK with the tyrosine kinase domain); homodimerization of the fusion protein.

### Expression / Localisation

Cytoplasmic localisation (in contrast with the t(2;5)(p23;q35) with NPM1-ALK, which localizes both in the cytoplasm and in the nucleus).

### Oncogenesis

TPM3-ALK is constitutively activated.

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