ALK-positive diffuse large B-cell lymphoma

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Clinics and pathology

Disease
Anaplastic lymphoma kinase (ALK) -positive diffuse large B-cell lymphoma (DLBCL) is a variant of DLBCL, expressing the ALK kinase (Delsol et al., 2008).

Phenotype/cell stem origin
The postulated normal counterpart is post-germinal centre B cell with features of plasma cell differentiation.

The immunophenotype typically show ALK+ (cytoplasmic staining). Plasma cell markers such as CD138 test positive, whereas B-cell markers such as CD20 and CD79a are negative, CD30 is usually negative. Cytoplasmic staining for IgA and, less frequently, IgG is typically seen in the majority of cases.

Clinics
The disease runs an aggressive course (Laurent et al., 2010). Long term survival was reported in some children.

Pathology
The lymph node sections show large immunoblastic cells with nucleoli.

Treatment
Chemotherapy using CHOP or CHOP-like regimens is the standard of care. Due to CD20 negativity, the monoclonal antibody rituximab is ineffective.

Prognosis
Median overall survival was 20 months in a study (Laurent et al., 2010).

Cytogenetics

Cytogenetics molecular
The Ig gene is clonally rearranged.
The founder lesion involves the ALK gene on chromosome 2 (Lee et al., 2008).
The t(2;17)(p23;q23) determining the clathrin/ALK fusion is the most frequent aberration (Chikatsu et al., 2003), accounting for approximately 70% of the cases. The classical t(2;5)(p23;q35) was reported in 10% of the cases (Adam et al., 2003).

Other aberrations, mostly reported in individual cases include:
- cryptic insertion of the ALK gene into chromosome 4q22 (Stachurski et al., 2007),
- cryptic SEC31A-ALK fusion generated by an insertion of the 5' end of SEC31A (4q21) upstream of the 3' end of ALK. An in-frame fusion transcript in which exon 24 of SEC31A (4q21) was fused to exon 20 of ALK is generated by this rearrangement (Van Roosbroeck et al., 2010).

References


This article should be referenced as such:
