BIRC3 (baculoviral IAP repeat-containing 3)

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Identity

Other names: API2; c-IAP2; HIAP1; MIHC; HAIP1
HGNC (Hugo): BIRC3
Location: 11q21

DNA/RNA

Description
Centromere to telomere orientation; 10 exons; large 5’ untranslated region, start codon in exon 3, stop codon in exon 10.

Transcription
6.5 kb mRNA.

Protein

BIRC3 protein - Baens Mathijs, Peter Marynen.

Description
68 kDa; 604 amino acids. The protein contains three ‘baculovirus inhibition of apoptosis’ (BIR) repeats, a caspase recruitment domain or CARD and a carboxyterminal ‘Zn-binding’ RING domain.

Expression
Wide, most abundantly in kidney, liver, small intestine and lung.

Function
Member of the 'inhibitor of apoptosis' (IAP) protein family. Binds to the tumor necrosis factor receptor via its associated factors TRAF1 and TRAF2 and inhibits apoptosis, probably by interfering with activation of caspases.

Homology
With Baculovirus IAP proteins, Drosophila DIAP1 and DIAP2.

Implicated in

t(11;18)(q21;q21) / marginal zone B-cell lymphoma of MALT-type --> BIRC3 - MALT1

Disease
B-cell non Hodgkin lymphoma (NHL); marginal zone B-cell lymphoma (MZBCL) of mucosa-associated lymphoid tissue (MALT); found in extranodal MZBCL or MALT-type, absent in splenic and nodal MZBCL.

Prognosis
For gastric MALT-type lymphomas, t(11;18) is a clonal marker for resistance to Helicobacter pylori eradication therapy and antigen independent growth.

Cytogenetics
Hybrid/Mutated gene
5’ API2 on chromosome 11q21 translocated on
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chromosome 18 in frame with 3’ MALT1. Deletions often exclude the expression of the reciprocal 5’ MLT ‘API2 3’ transcript.

Abnormal protein
All MALT-type lymphomas reported with a t(11;18) express an ‘in frame’ API2-MLT fusion protein with consistently the three BIR domains of API2 fused to the caspase p20 domain and VDJ4-like domain of MLT.

Oncogenesis
Transient expression of the API2-MLT fusion protein activates an NF-KB luciferase reporter construct, suggesting the involvement of this signaling cascade in MALT lymphomagenesis.

References


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