

Leukaemia Section

Mini Review

Polyclonal B Lymphocytosis with Binucleated Lymphocytes (PPBL)

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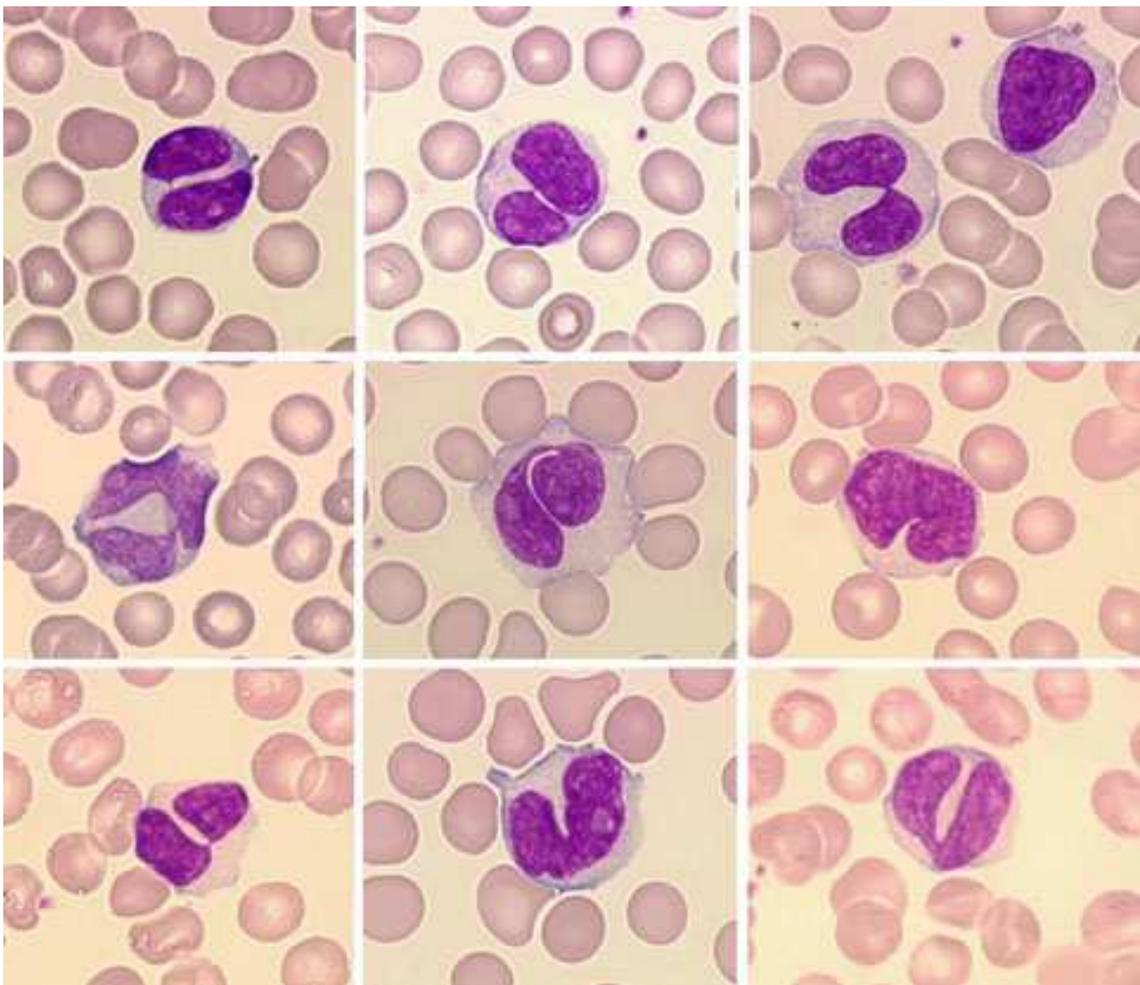
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Clinics and pathology



Morphologic features showing typical binucleated cells.

Phenotype/cell stem origin

Unknown. The polyclonal expansion of B-cells fit into the peripheral CD27+IgM+IgD+ B cell population. Cloning and sequencing of VH genes of PPBL IgVH genes showed a mutated profile suggesting like CD27 expression an expansion of memory B cells.

Etiology

The etiology of polyclonal B lymphocytosis with binucleated lymphocytes (PPBL) remains unknown. An association with cigarette smoking was initially suggested. However PPBL was observed in non smokers patients. The morphology of binucleated lymphoid B-cells could suggest an association with viral infections, such as Epstein - Barr virus. Biologic studies are not completely achieved to exclude and/or to confirm definitely the role of EBV in the pathogenesis of PPBL. The presence of characteristic binucleated lymphoid B-cells in asymptomatic family members and the description of familial PPBL cases suggest a genetic predisposition as a more likely possibility.

Epidemiology

PPBL was first reported in 1982. We have no epidemiological data on the incidence of PPBL.

Clinics

In a large series we reported on forty-three patients (9 males, 34 females: median age: 40 years, range 28-65), the clinical characteristics were spleno-megaly in 16%, hepatomegaly in 0.5% and lymph nodes in 11.5% cases. An absolute lymphocytosis $> 4 \times 10^9/l$ is present in 80% of PPBL patients. A persistent, stable and polyclonal increase of IgM levels is usual and most PPBL patients express HLA-DR7.

Cytology

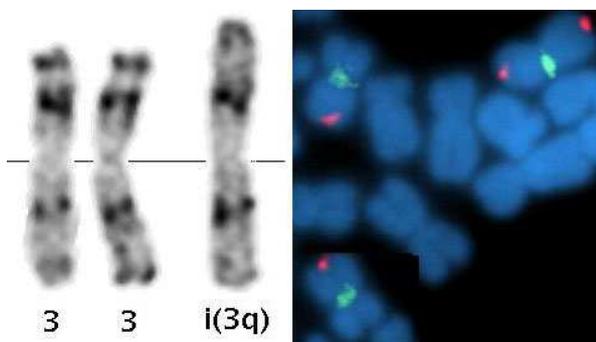
PPBL is identified in all cases by the presence of a variable (1.5 to 9%) number of binucleated peripheral lymphoid cells (Fig 1). The majority of lymphoid cells are large with abundant faintly and basophilic cytoplasm. Characteristic nuclei with a rounded or more commonly irregular form are observed.

Immunologic markers: Both kappa and lambda light-chain are expressed, indicating a polyclonal expansion of the lymphocyte pool. The lymphocytosis is of the B-cell type: the lymphocytes react with CD19, CD20, CD22 and FMC7 antigens.

Prognosis

After a median follow-up of 5.5 years without treatment, 45 PPBL patients are alive.

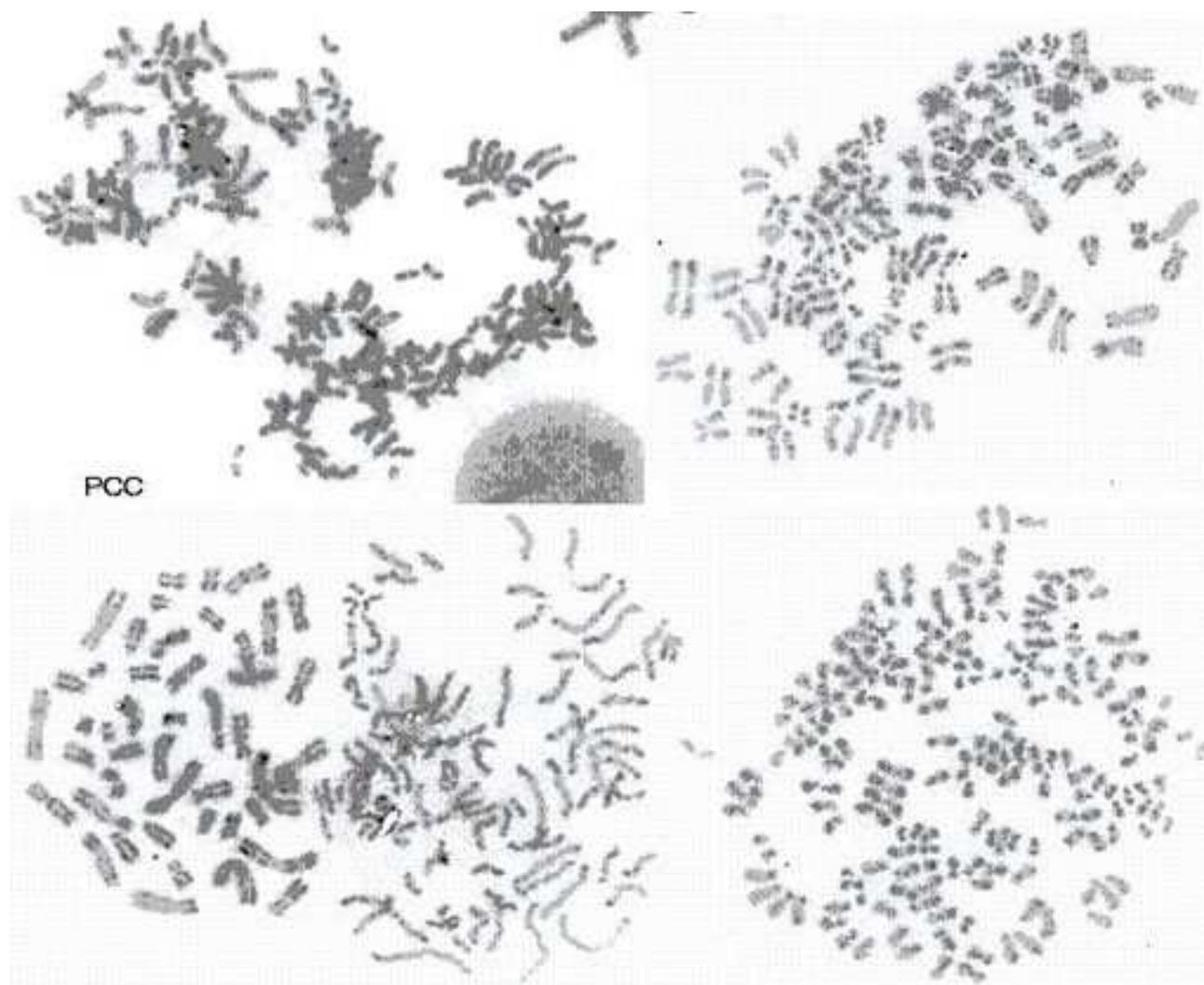
Cytogenetics



Partial karyotype showing +i(3q) -R-banding (left); Detection of i(3q) with telomere chromosome 3q and alpha satellite specific DNA probes (right).

Cytogenetics morphological

+i(3q) (Fig 2) is the most common abnormality and is observed in 70% cases, occurring as a single aberration in only a few patients. PCC (Fig 3) is observed in 40% cases and occur rarely as a sole abnormality. Both abnormalities associating +i(3q) and PCC are present in 37% cases. Using alpha-satellite and telomere chromosome 3 specific probes, +i(3q) is more frequently detected by metaphase FISH studies. (Fig 2). +i(3q) is rarely described as a recurrent cytogenetic abnormality in patients with hematologic malignancy. Trisomy 3 is reported to be associated with marginal zone B-cell lymphoma. Gain of chromosome 3 or 3q was described in patients with typical clonal b-cell chronic lymphoproliferative disorders, chronic lymphocytic leukemia, prolymphocytic leukemia or Waldenström macroglobulinemia. A chromosomal instability is present in 67.5% patients. These patients present various clonal [del(6q), +der(8) or +8 or polyploid karyotype] and non clonal chromosomal abnormalities with structural and numerical abnormalities. This chromosomal instability is variable over time but persist in most cases. In spite of genomic instability, a long follow-up of PPBL patients remains essential and chemo-therapy unnecessary.



Premature Chromosome Condensation (PCC).

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