Chronic neutrophilic leukemia evolving from polycythemia vera with multiple chromosome rearrangements: a case report

Chronic neutrophilic leukemia (CNL) is a rare disorder defined by a persistent increase of mature peripheral neutrophils in the absence of monocytosis, basophilia, eosinophilia, Philadelphia chromosome, occult infection or malignancy. Although CNL is considered a distinct entity, it was recently reported in association with polycythemia vera (PV). We report one case of CNL associated with chromosomal abnormalities evolving from a long history of PV.

A female patient with PV, diagnosed in 1984 at the age of 55 years, was treated with hydroxyurea and phlebotomy until 1998 when splenectomy was performed because of massive splenomegaly. At that time, bone marrow examination revealed severe fibrosis associated with chronic myeloproliferative syndrome without signs of blast transformation. After two months, despite treatment with hydroxyurea, progressive neutrophilic leukocytosis in the range of 120,000/mm³ developed.

The diagnosis of CNL was confirmed by the presence of granulocytic proliferation without morphologic dysplasia in a bone marrow biopsy, elevation of leukocyte alkaline phosphatase and absence of BCR/ABL hybrid gene transcripts of p210 and p185. Cytogenetic study performed on the only 5 available metaphases showed in 3 reciprocal translocations between the long arm of chromosome 8 and the long arm of chromosome 11: t(8;11)(q24.3;q147), t(15;19)(p11.2;q12), and t(15;19)(p12;q12), loss of one chromosome X and presence of a marker chromosome of unknown origin (Figure 1). Southern blot analysis of the MLL gene did not reveal any rearrangement. Despite chemotherapy with etoposide and busulfan the patient died of bronchopneumonia.

Figure 1. Karyotype of the patient: 46, X,-X, t(8;11)(q24.37;q147), t(15;19)(p11.27;p127)+ mar.

The clonal nature of CNL has been the subject of controversy, particularly when hyperleukocytosis is associated with other clonal processes. In our case, in accordance with the findings of Elliott et al., the extent and the progression of the leukocyte count argue reasonably against a reactive process.

In conclusion, we describe a case of aggressive and refractory hyperleukocytosis compatible with CNL evolving from PV. The documentation of the associated karyotypic abnormalities may add some clues about the cytogenetic profile in PV-derived CNL.

Key words: chronic neutrophilic leukemia, polycythemia vera.

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References


