Dicentric dic(7;9)(p11;p11): a new case in childhood ALL

Elvira D Rodrigues Pereira Velloso, Carolina Kassab, Silvia Helena A Figueira, Denise Tiemi Noguchi, Eliana Carla Armelin Benites, Cristóvão L P Mangueira, Fábio Morato de Oliveira

Clinical Laboratory, Hospital Israelita Albert Einstein, Sao Paulo, Brazil (EDRPV, CK, SHAF, CLPM); Instituto de Clinicas Pediatricas Bolivar Risso - GRENDAAC, Sao Paulo, Brazil (DTN, ECAB); Cytogenetics and Onco-Hematology, FMRP USP, Brazil (FMDO)

Published in Atlas Database: September 2009
DOI: 10.4267/2042/44836
This work is licensed under a Creative Commons Attribution-Noncommercial-No Derivative Works 2.0 France Licence. © 2010 Atlas of Genetics and Cytogenetics in Oncology and Haematology

Clinics

Age and sex
13 years old female patient.

Previous history
No preleukemia. No previous malignancy. No inborn condition of note.

Organomegaly
Hepatomegaly, splenomegaly, enlarged lymph nodes, no central nervous system involvement.

Blood

WBC: 28.5X 10^9/l
HB: 6.6g/dl
Platelets: 110X 10^9/l
Blasts: 64%
Bone marrow: 72.4% of lymphoid blast cells.

Survival

Date of diagnosis: 02-2009
Treatment: Chemotherapy with BFM95 for intermediate risk.
Complete remission: remission was obtained after the first induction cycle.
Treatment related death: no
Relapse: no
Status: Alive. Last follow up: 04-2009
Survival: 1 month

Karyotype

Sample: Bone marrow
Culture time: 24h and 48 h without stimulating agents
Banding: GAW- band
Results:
Karyotype at Relapse: not applied.
Other molecular cytogenetics technics:
Spectral Karyotyping (SKY) using SkyPaint ASR (Applied Spectral Imaging). The marker seen in the first clone was elucidated as a derivative chromosome 9

The study confirms the der(7;9) seen in the second clone (figure 4).

Other Molecular Studies

Technical: Not done.

Figure 1: G-banding karyotype showing the first clone with monosomy 7 and one marker (recognized as a der(9) after sky study).

Figure 2: Partial G-banding karyotypes showing the second clone with dic(7;9)(p11;p11).

Figure 3: Sky study showing the first clone with monosomy 7 and a der(9).
Comments

The case presented here is, to our knowledge, the 19th reported case of dic(7;9)(p11;p11) in acute lymphoblastic leukemia. From the literature review, 10 patients were less than 15 years old, seven with FAB L1 morphology, like our patient. She presented an enlarged liver and spleen, as ten and six of the cases reported in the studies, but without hyperleukocytosis, which was common for ALL patients with simultaneous dic(7;9) and t(9;22), present in 9 of 18 cases.

The prognostic significance of this abnormality remains controversial. Russo et al. suggested that the deletion of tumor suppressor genes located on 7p is associated with an adverse prognostic factor in ALL.

Heerema et al. related that abnormalities in chromosome 9p are associated with increased relapse in children with ALL, probably because the inactivation of the tumor suppressor genes CDKN2 and CDKN1, mapped to 9p21-22. In the series of Pan et al. (2006), the prognosis of the patients with dic(7;9) and t(9;22) is worse than that of those with isolated dic(7;9).

From the 18 cases, 11 have no data of survival, 4 achieved long term remission and 3 died. Our children remain in clinical remission 45 days after induction therapy with intermediate risk BFM 95 (Berlin-Frankfurt-Munster) protocol.

References


This article should be referenced as such: