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

**Clinics**
- **Age and sex:** 13 year(s) old female patient.
- **Previous History:** no preleukemia
  - no previous malignant disease
  - no inborn condition of note
- **Organomegaly:** hepatomegaly; splenomegaly; enlarged lymph nodes; no central nervous system involvement

**Blood**
- WBC: $28.5 \times 10^9$/l; Hb: 6.6 g/dl; platelets: $110 \times 10^9$/l; blasts: 64%
- Bone marrow: 72.4% (of lymphoid blast cells)

**Cyto pathology classification**
- Cytology: ALL-L1
- Immunophenotype: Blast cells positivity for: CD 34, HLA, CD10, CD19, CD22 and cCD79a.
- Rearranged Ig Tcr: not done
- Pathology: not done
- Electron microscopy: not done
- Precise diagnosis: Common B (B-II) ALL at diagnosis.

**Survival**
- **Date of diagnosis:** 02-2009
- **Treatment:** Chemotherapy with BFM95 for intermediate risk
- **Complete remission was obtained**
- **Comments:** remission was obtained after the first induction cycle
- **Treatment related death:** -
- **Relapse:** -
- **Status:** Alive 04-2009
- **Survival:** 1 month(s)

**Karyotype**
- **Sample:** Bone marrow; culture time: 24 h, and 48 h without stimulating agents; banding: GAW-band
- **Results:** 45,XX,-7,+mar[11]/45,XX,dic(7;9)(p11;p11)[9]/46,XX[4]
- **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 (figure 3). The study confirm the der(7;9) seen in the second clone (figure 4).
- **Other molecular studies technics:** 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).
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.

**Bibliography**

- *Three adults with acute lymphoblastic leukemia and dic(7;9)(p11.2; p11).* Smith A, Das P, O'Reilly J, Patsouris C, Campbell LJ.