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A MULTICENTER EXPERIENCE FROM LEBANON IN CHILDHOOD AND ADOLESCENTS ACUTE MYELOID LEUKEMIA: HIGH RATE OF EARLY DEATH IN CHILDHOOD APL

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Background: Lebanese Social & Health Facts

- Population:
  - 4 million Lebanese
  - 2 million non-Lebanese refugees
- 10,452 km^2
- High level of education
- Health care system mostly private - cancer patients partially covered by MOH

Lebanon Social & Health Facts

- No regular Cancer statistics. Incidence of AML and therefore APL unknown
- Advanced Lab Tests & Transfusion Services available in tertiary care centers but not ideal in small centers or rural areas
- ATRA available upon request: not always present in Hospital stocks – coverage of ATRA varies

Background: AML & APL

- AML is a disease with marked heterogeneity in clinical and biologic features, response to therapy and survival.
- Despite major achievements in the treatment of AML, long term survival remains poor. Half of pediatric AML patients relapse and die internationally.
- APL is a unique subtype with distinct biologic and molecular characteristics, from highly fatal 50 years ago to highly curable nowadays but still associated with high rates of early death and delays in diagnosis in the “real world”.
- There is no published data on pediatric AML or APL in Lebanon

Objectives

- Identify: Clinical, Cytogenetic, Molecular Findings and Outcome data of pediatric AML in the Lebanese population in comparison to the International and Regional data available.

- Focus the study on Childhood and Adolescent APL in Lebanon.

Methods

- Retrospective chart review of children with AML and APL diagnosed at 3 tertiary care centers in Beirut over the past ten years.

- Data collection sheets were filled and analyzed.

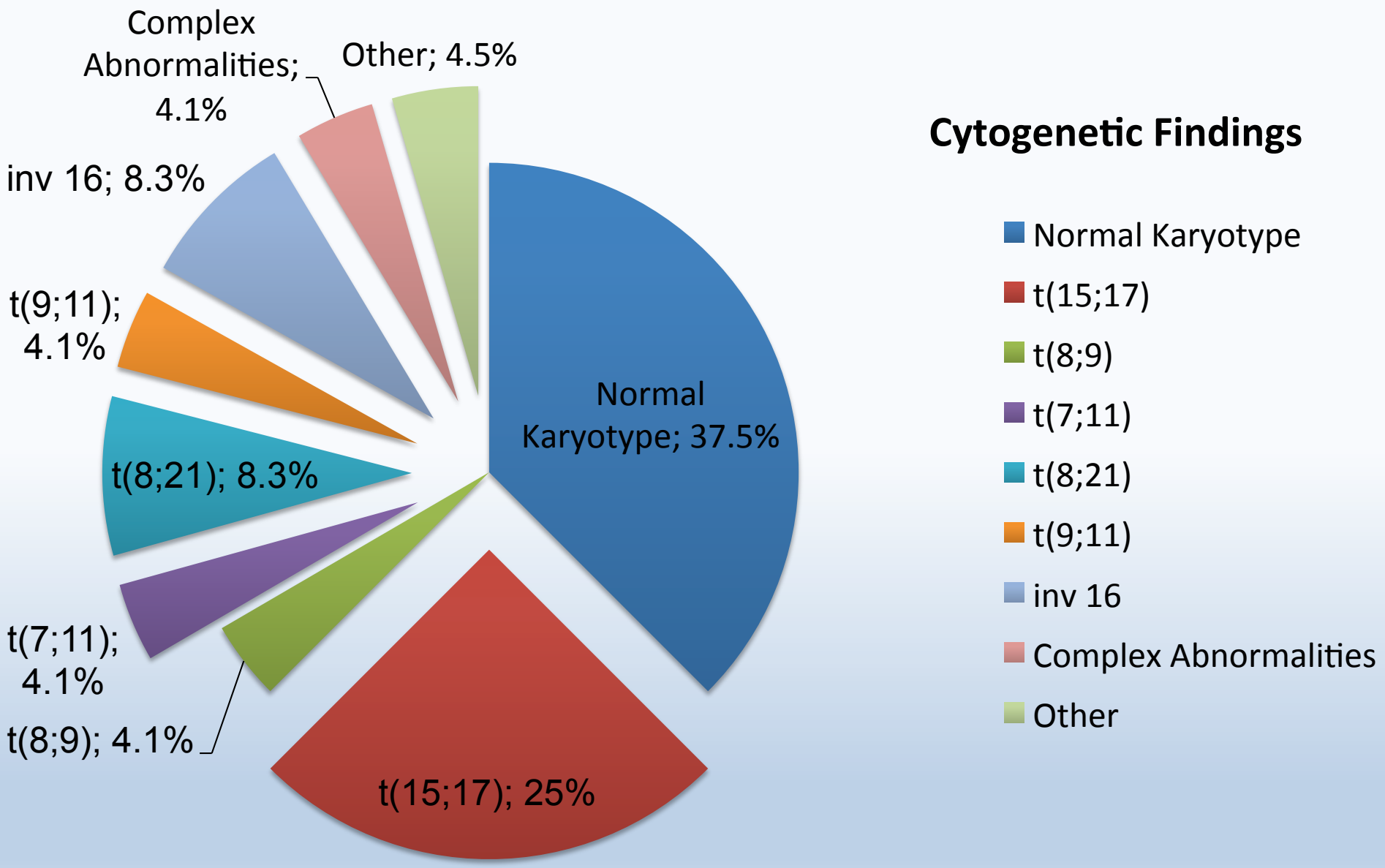
DATA Collection Sheets

- Demographic Information
- Date of Diagnosis and Presenting Symptoms
- Blood Analysis, Bone Marrow Aspirate, Cytogenetic Features, Molecular Biology Analysis, HLA typing
- Treatment and Outcome
- Data on HSCT and Long-Term Follow-up

Results: Total AML & APL Patients

- Twenty four patients were diagnosed with AML from 2002-2010 , 12 girls and 12 boys.
- Two had pre-existing Fanconi anemia, 1 Down syndrome, 1 MDS with monosomy 7, 1 secondary AML after treatment for Burkitt's lymphoma.
- 6 (25%) had *de novo* APL with t(15;17)
- Mean age was 8.6 years (range, 1-24)
- Median WBC at dx = 31 x 10^9/L (range, 2.1-376); PLT:46 x 10^9/L (range, 10-164)

Results


Complex Abnormalities; 4.1%
Other; 4.5%
inv 16; 8.3%
t(9;11); 4.1%
Normal Karyotype; 37.5%
t(8;21); 8.3%
t(7;11); 4.1%
t(8;9); 4.1%
t(15;17); 25%
Cytogenetic Findings
Normal Karyotype
t(15;17)
t(8;9)
t(7;11)
t(8;21)
t(9;11)
inv 16
Complex Abnormalities
Other

Results: Molecular Studies not Performed on all Children

- FLT-3 (Internal Tandem Duplications) were detected in 3 AML patients and was associated with high WBC at presentation and a poor outcome

- NPM1 was screened in one patient, mutations were not found.

Results

- 71% of patients developed AML at an age < 10 years with the youngest is 1 year old.
- This age group carried a 50% survival rate compared to 0% survival in patients > 10 yrs.

Results: Early Death in APL

- Death in induction was observed in 3/6 patients with APL (50%). All 3 patients had an initial WBC >10 x 10^9/L

- Two died during induction due to DIC and CNS bleed despite a quick diagnosis and early start of ATRA; 1 died from CNS bleed 2 days after diagnosis and before starting treatment.

HSCT in AML

- Indicated in 15 patients with AML and was performed on 9 children in Europe or the United States: 5 MUD and 4 matched related sibling.
- Survival after transplant was 37.5%.
- Two AML children died: 1 with M6 AML and 1 AML post-Fanconi anemia.

Results: All AML Patients

- Median survival for patients who died from disease progression was 25.8 months.
- Overall disease–free survival was 30.4%.

Conclusions

- APL represented 25% of all AML in our series
- Early death in APL was 50%.
- Overall survival in the AML cohort was 30.4% .
- Early death in all AML patients was 20%.

Areas of Improvement for APL

- More rapid availability of ATRA in hospitals
- Start ATRA at first morphologic suspicion
- Aggressive early transfusions of PLTs and FFP or Cryoprecipitates
- Increase awareness among general practitioners for early referral to specialized centers
- Further data collection to include the entire country

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Thank you
for your attention