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Leukaemia Section
Short Communication

+13 or trisomy 13

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Identity

Other names

+13

Trisomy 13

Clinics and pathology

Disease

- Minimally differentiated acute myeloid leukemia (AML) (FAB Type M0)
- Acute myeloblastic leukemia, without maturation (FAB Type M1)
- Acute monoblastic/monocytic leukemia (FAB Type M5)
- Acute erythroid leukemia (FAB Type M6)
- B-cell acute lymphoblastic leukemia (B-ALL)
- Myelodysplastic syndrome
- Idiopathic myelofibrosis
- Atypical chronic myeloid leukemia (CML)

Epidemiology

Trisomy 13 (when the sole cytogenetic abnormality) in AML manifests most commonly as minimally differentiated AML (FAB Type M0), and has a predilection for older men over 70.

Mesa et al. found that the incidence rate of trisomy 13 was 0.7% of all AML in their respective study. Other associated diseases have been, to date, described rarely.

Trisomy 13 in acute myeloblastic leukemia, without maturation (FAB Type M1) has been described in two older men from India (Trivedi et al., 2009).

Trisomy 13 in atypical CML was described in 1 case report from China in 2011 (Guo-Yu et al., 2011).

Pathology

According to Mehta et al. (1998), characteristic small hand-mirror blasts with cytoplasmic blebs and tails and scanty small granules were seen in 13/24 cases and 18/25 cases had small blasts would could easily be mistaken for lymphoblasts. Morphologic findings in bone marrow, according to Mesa et al. were: median of 80% cellularity, 21% median bone marrow blasts (range 1%-94%), myelodysplastic changes in 56%, reticulin fibrosis in 11%, and ringed sideroblasts in 11% of cases.

Treatment

One recent study (Fehniger et al., 2009) reported induction of sustained morphologic and cytogenetic complete remission in 2 older patients with AML with trisomy 13 (as the sole cytogenetic abnormality) treated with lenalidomide.

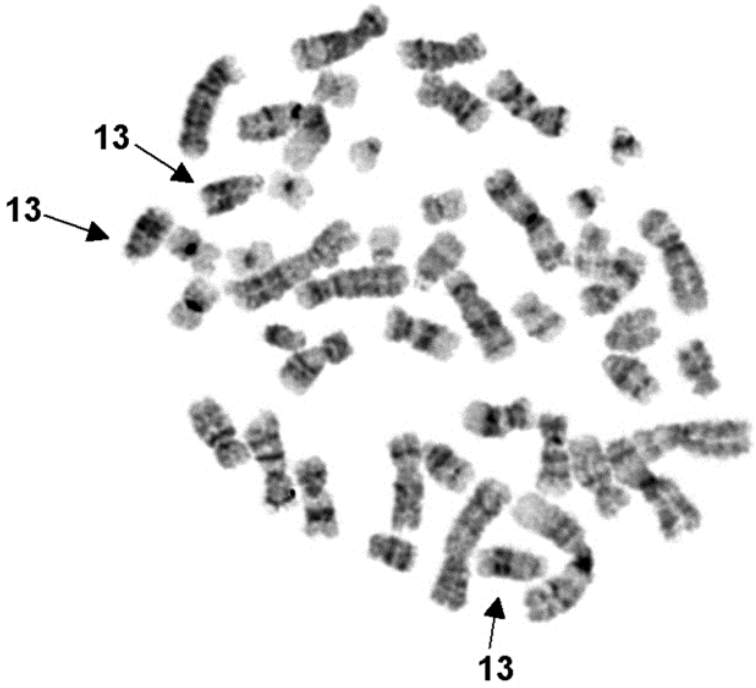
However, in regards to a 2011 Fehniger article re: clinical trials of high-dose lenalidomide for older male AML patients, the Swedish AML group responded via an online published letter that lenalidomide has not been shown to benefit older men with AML.

Prognosis

Trisomy 13 in AML is considered a poor prognostic factor, with low complete remission rate and brief remission duration.

Per Mehta et al. (1998), median patient survival was 3 months. The paper by Dohner et al. described 8 individuals with trisomy 13 as the sole cytogenetic abnormality, with survival ranging from 0.5 to 14.7 months.

Mesa et al. report from a retrospective study a median survival of 6.1 months.



GTG-banded metaphase spread showing the isolated trisomy 13.

Genetics

Note

AML with trisomy 13 is strongly associated with presence of RUNX1 mutations and a high expression of FLT3 mRNA. Because FLT3 is localized on chromosome 13, Dicker et al. (2007) hypothesized that RUNX1 mutations may cooperate with trisomy 13 in leukemogenesis by increasing FLT3 transcript levels. They found that increasing FLT3 transcript levels revealed a highly significant ($P < 0.001$) ~5-fold increase in AML with RUNX1 mutations and trisomy 13 compared with samples without trisomy 13.

Cytogenetics

Note

Trisomy 13 occurs as a single clone in the majority of the cases so far reported. However, additional chromosome aberrations were observed in a second related clone. A few cases of tetrasomy 13 have been reported as second abnormal clone.

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