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Continuing Medical Education (CME) Questions

Genetic characterization of B-PLL

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Chapiro E, Pramil E, Diop M, Roos-Weil D, Dillard C, Gabillaud C, Maloum K, Settegrana C, Baseggio L, Lesesve J-F, Yon M, Jondreville L, Lesty C, Davi F, Le Garff-Tavernier M, Droin N, Dessen P, Algrin C, Leblond V, Gabarre J, Bouzy S, Eclache V, Gaillard B, Callet-Bauchu E, Muller M, Lefebvre C, Nadal N, Ittel A, Struski S, Collonge-Rame M-A, Quilichini B, Fert-Ferrer S, Auger N, Radford-Weiss I, Wagner L, Scheinost S, Zenz T, Susin SA, Bernard OA, Nguyen-Khac F; the Groupe Francophone de Cytogénétique Hématologique (GFCH) and the French Innovative Leukemia Organization (FILO). Genetic characterization of B-cell prolymphocytic leukemia: a prognostic model involving *MYC* and *TP53*. *Blood*. 2019;134(21):1821-1831.

1. Your patient is a 72-year-old man with B-cell prolymphocytic leukemia (B-PLL). According to the case series by Chapiro et al, which of the following statements about the genetic portrait of B-PLL is correct?

- ☐ B-PLL is highly associated with *MYC* deletion (del) and 17p (*TP53*) aberrations (translocation or gain)
- ☐ The karyotype was complex (≥3 abnormalities) in 23% of the patients and highly complex (≥5 abnormalities) in 15%
- ☐ The most frequent chromosomal aberrations were translocations involving *MYC* [t(*MYC*)] (62%), del17p (38%), trisomy (tri)18 (30%), and del13q (29%)
- ☐ Whole-exome sequencing showed rare mutations in *TP53* and *MYD88*

2. According to the case series by Chapiro et al, which of the following statements about correlations between cytogenetic and molecular findings in B-PLL and the patients' clinical outcomes is correct?

- ☐ Patients with t(*MYC*) had significantly higher overall survival than patients with *MYC* gain
- ☐ Cases with *MYC* aberration and 17p (*TP53*) deletion had the worst prognosis
- ☐ Only 2 distinct cytogenetic risk groups were identified
- ☐ Cytogenetic analysis was useful only for diagnosis but not for prognosis

3. According to the case series by Chapiro et al, which of the following statements about primary B-PLL cells' in vitro response to novel targeted therapeutics is correct?

- ☐ Findings of this study show that targeting del17p is likely the most useful treatment option in B-PLL
- ☐ Cases with *MYC* gain were highly susceptible to the combination of ibrutinib or idelalisib with OTX015 (a bromodomain and extra-terminal motif inhibitor [iBET])
- ☐ The ATP assay and flow cytometry assay showed conflicting results
- ☐ Drugs targeting the B-cell receptor and *BCL2* in combination with an iBET may be a treatment option for patients with B-PLL

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