8p11 Stem Cell Leukemia/Lymphoma Syndrome without Myeloproliferation: A Rare Clinical Entity

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Received: 27 July 2017 / Accepted: 25 September 2017 / Published online: 27 September 2017
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Patient was a 13 year old boy presenting with bilateral large neck lymphadenopathy. Imaging workup showed no mediastinal widening or abdominal organomegaly (spleen largest diameter was 10 cm). Complete blood count was as the following: white blood cell = 6800/ μ L (neutrophil = 46%, lymphocyte = 34%, Monocyte = 16%, Eosinophil = 4%), red blood cell = 4.42000/ μ L, hemoglobin = 13.3 gr/dL, hematocrit = 38%, platelet = 332,000/ μ L. Although bone marrow aspiration was normocellular, synchronized bone marrow culture revealed t(8;13) (p11;q12) (Fig. 1a). Cervical lymph node biopsy showed infiltration of small to medium lymphoid blasts

with dusty chromatin extending to perinodal fat (Fig. 1B). Immunohistochemistry exhibited positive reaction for CD3, tdt and Ki67 which was compatible with T cell lymphoblastic lymphoma. Final diagnosis was 8p11 stem cell leukemia/lymphoma syndrome.

8p11 stem cell leukemia/lymphoma syndrome is a clinicopathologic entity with less than 100 reports that is characterized by the following features: (1) 8p11 reciprocal translocation, (2) myeloproliferative disorder with eosinophilia, (3) T lymphoblastic lymphoma/leukemia presenting with lymphadenopathy, (4) rapid transformation to acute myeloid leukemia [1]. This syndrome can happen at

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Fig. 1 **a** Bone marrow karyotype with Giemsa banding showed t(8;13)(p11;q12). **b** Cervical lymph node histologic sections revealed infiltration of lymphoid blasts with extension to perinodal fat

any age ranging from 3 to 84 years with male to female ratio of 1.5. Lymphoma and myeloproliferation can occur simultaneously or sequentially in this syndrome [1, 2].

Compliance with ethical standards

Conflict of interest Authors declares that he/she has no conflict of interest.

Ethical approval All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Informed consent Informed consent was obtained from individual participant included in the study.

References

1. Jackson CC, Medeiros LJ, Miranda RN (2010) 8p11 myeloproliferative syndrome: a review. *Human Pathol* 41:461–476
2. Kuskonmaz B, Kafalı C, Akcoren Z, Karabulut HG, Akalin İ, Tuncer MA (2008) The 8p11 myeloproliferative syndrome in a 3-year-old child. *Leuk Res* 32:198–199