To the editor:

Development of Hodgkin lymphoma in homozygotic triplets with constitutional deletion in MKL1

First-degree relatives of patients with Hodgkin lymphoma (HL) have a threefold to fourfold increased risk of developing HL, which is stronger when the proband is <40 years at diagnosis, in males and in siblings. Monozygotic twins carry an even higher risk than first-degree relatives. It is currently not known whether (or how) extrinsic factors interact with genetic predisposition though a susceptibility gene on chromosome 4 has been suggested. We herein report, to our knowledge, the first cases of HL in 2 male homozygotic triplets occurring in middle adulthood and with a 23-year interval. The first triplet was diagnosed in 1985 at age 40 with stage IIIA HL of Epstein-Barr virus (EBV)-positive mixed cellularity subtype. He remains in remission after mustargen, oncovin, procarbazine, prednisone/adriamycin, bleomycin, vinblastine, and dacarbazine chemotherapy. The second triplet developed stage IIIB HL in 2008 at the age of 63. His tumor was also EBV-positive but of nodular sclerosis (NS) subtype. A complete and lasting remission was achieved following adriamycin, bleomycin, vinblastine and dacarbazine chemotherapy.

Considering the epidemiology of the different HL subtypes in developed countries, with mixed cellularity and NS predominantly arising in older and younger patients, respectively, the reciprocal presentation in these patients is somewhat unusual. Similarly, the second triplet developed stage IIIB HL in 2008 at the age of 63. His tumor was also EBV-positive but of nodular sclerosis (NS) subtype. A complete and lasting remission was achieved following adriamycin, bleomycin, vinblastine and dacarbazine chemotherapy.

In summary, we report the first constitutional mutation of MKL1 in homozygotic triplets of whom 2 so far have developed HL. The deletion may represent an accidental finding, but potentially it could have contributed to the pathogenesis of HL.

References

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