Clinical Case of Clonal Hemopoiesis in Patient with Splenectomy in Childhood

Nadezhda N. Nemstsveridze MD, Irina S. Martynkevich PhD, Elizaveta V. Efremova MD, Ekaterina V. Motyko PhD, Anna N. Kirienko MD, Daria V. Kustova MD, Vasiliy A. Shuvaev PhD, MD, Sergey V. Sidorkevich PhD

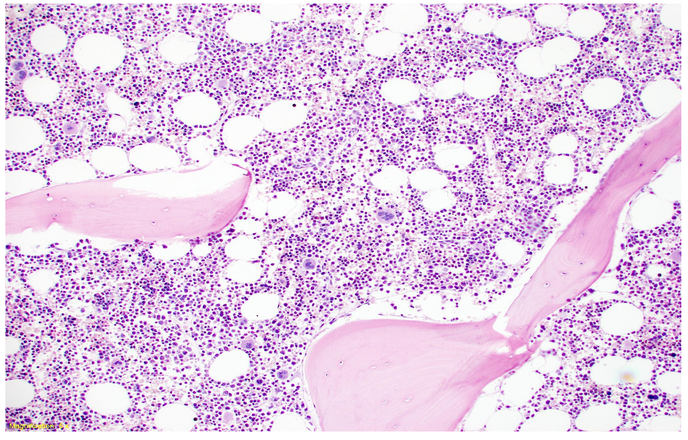
Russian Research Institute of Hematology and Transfusiology, Saint Petersburg, Russian Federation

Context

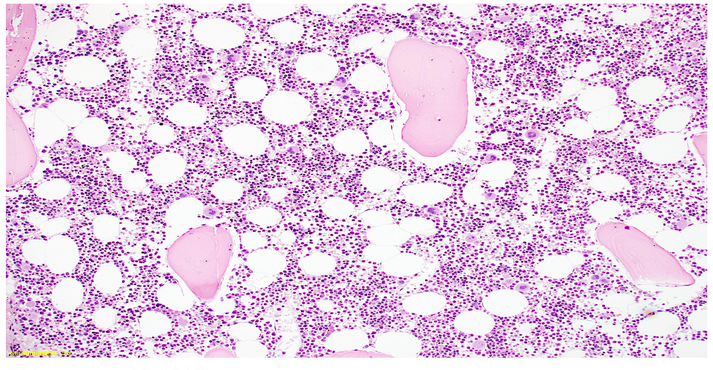
There is a subset of patients in the population who have undergone splenectomy for various reasons. Normally one of the spleen's functions is to eliminate defective blood cells and the impact of splenectomy on hematopoiesis is not clear yet.

Clinical Case

A patient born in 1993 with verified Minkowski-Chauffard's disease in 2005 is known to have a history of splenectomy at the age of 13 without any complications. The blood tests have been stable with minor deviations. In November 2016 patient consulted a hematologist due to complete blood count (CBC) which showed thrombocytosis (PLT –1101x109/L), leukocytosis (WBC-18,46, ANC-13,18) detected for the first time. Such an increase in PLT level could not easily be explained by the post-splenectomy syndrome, therefore the patient was further examined. No driver mutations were found according to molecular genetic tests (02.2017): JAK2V617F, BCR::ABL, CALR, MPL were not detected. The morphological examination of BM biopsy (05.2017) showed proliferation of mature megakaryocytes; erythrophagocytosis; MF–0; chronic myeloproliferative neoplasms? It was not possible to make an accurate diagnosis since the results did not meet the WHO diagnostic criteria for MPN. In subsequent monitoring, thrombocytosis was increasing (PLT–1857 09.2018), so next-generation sequencing (NGS) was performed to confirm the clonality of hematopoiesis. NGS (09.2018): polymorphisms in DNMT3A (4,8 exon), KIT (14 exon), BRAF, GNAS, BCOR, BCL6, CEBPA, PDGFRB, TP53 (3 exon) genes were detected. The BM biopsy was repeated on 03.2019: hyperplasia and dysplastic changes in the megakaryocytic germ of myeloid tissue; immature cells in granulocytic germ; MF–0. Despite the lack of MPN diagnostic criteria cytoreductive therapy, the combination of Interferon α2β and hydroxyurea had been started on 03/2019 due to the high risk of thrombohemorrhagic complications. Interferon therapy was ceased after a year of treatment due to intolerance, only hydroxyurea was continued but compliance to therapy was not good. Nevertheless, PLT level was stabilized [CBC (06.21): WBC-17,7, HB-158, PLT-973] without any thrombotic or hemorrhagic complications.



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