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High-grade B-cell lymphomas in children: Focus on new entities created in the 2017 revision of the World Health Organization classification of tumors of hematopoietic and lymphoid tissues

The 2017 revision of the World Health Organization Classification of Tumors of Hematopoietic and Lymphoid Tissues (2017 WHO) has created several new provisional entities of high-grade B-cell lymphomas, including Burkitt-like lymphoma with 11q aberration, high-grade B-cell lymphoma, not otherwise specified (NOS), and high grade B-cell lymphoma with C-MYC and BCL-2 and/or BCL-6 rearrangements (double hit or triple hit lymphomas). Little is known on the clinicopathologic features, diagnostic approaches, as well as the prevalence of these new entities in pediatric patients. We collected pathologic and clinical data from the medical record on all pediatric high-grade B-cell lymphoma (HGBL) cases diagnosed in the past 10 years at our institution (2007-2017). Nine cases that did not meet criteria for either BL or diffuse large B-cell lymphoma (DLBCL) underwent FISH for MYC, BCL-2, and BCL-6, as well as array comparative genomic hybridization (CGH). A total of 52 cases of HGBL were identified. These included 23 cases of classical Burkitt lymphoma (BL) as defined by classic morphology and MYC rearrangement, 20 cases of diffuse large B-cell lymphoma (DLBCL), and 9 other cases. Chromosome 11q aberrations were identified in 5 out of the 9 non-DLBCL, non-BL HGBL cases. The other 4 cases were classified as HGBL, not otherwise specified (NOS.) We did not identify any cases of HGBL with MYC and BCL-2 and/or BCL-6 rearrangements. Morphologically, all 5 cases of Burkitt-like lymphoma with 11q aberration showed typical pathological features as described in 2016 WHO. All 5 of these cases occurred in the head/neck region. Four of these cases were localized (stage II), with the remaining case also involving a few metabolically active but non-enlarged lymph nodes in the chest and abdomen (stage III). All 5 patients achieved complete remission with standard therapy for mature B-cell lymphoma. All patients were alive with no clinical evidence of disease at a median follow-up time of 34 months (range 12-95 months). Our results suggest that the majority of pediatric non-Burkitt, non-DLBCL cases of HGBL carry 11q aberrations. In addition, patients with 11q aberrations appear to be more likely to present with lower stage disease, thus requiring less intensive therapy, and also tend to have the primary disease in the head/neck. While HGBL-NOS does happen, double hit or triple hit lymphoma is almost never seen in Children. These findings further support the classification of Burkitt-like lymphoma with 11q aberration as a distinct pathologic and clinical entity.

Biography

Shunyou Gong is the director of Hematopathology at the Ann and Robert H Lurie Children's Hospital of Chicago and assistant professor at Northwestern University Feinberg School of Medicine. Gong's clinical interests and areas of expertise include pediatric hematopoietic malignancies, inherited bone marrow failure syndromes, and bleeding or thrombotic disorders. As first-author and corresponding author he has published many landmark papers on prestigious journals including Cell, Blood, and American Journal of Surgical Pathology.

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