Current status of diagnosis and treatment of langerhans cell histiocytosis in Korea

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Langerhans cell histiocytosis (LCH) has a wide range of clinical presentations, and its clinical course varies widely from spontaneous regression to severe disseminated disease with the risk of permanent consequences. Although the pathogenesis of LCH has been a conundrum, recent advances have led to a better understanding of the molecular pathogenesis and ontogeny of the disease. Advanced genomic analyses have suggested that LCH is a disorder of MAPK pathway mutations. Due to the rarity of LCH, collaborative, cooperative group trials, especially those of Histiocyte Society, have contributed to the development of the treatment protocol. However, optimal treatment for recurrent or high-risk LCH has not yet been established.

The Korea Histiocytosis Working Party (KHWP) reported the outcome of a large, retrospective multicenter study in Korean children with LCH. A nationwide, multicenter, prospective registry of LCH was launched in 2013 with the support of the Korean Centers for Disease Control to overcome the limitation of the retrospective study and better understand its pathobiology and clinical course. The registry has collected patients' information with LCH, using iCReaT (internet-based clinical research and trial management system), a web-based data entry system. The goal of the registry is to produce epidemiologic and clinical data to reveal the current status of diagnosis and treatment of LCH, leading to developing an optimal treatment strategy for LCH in Korea. The KHWP is also trying to establish Korean guidelines for the diagnosis and treatment of LCH. Furthermore, the introduction of genomic sequencing and targeted agents is changing the landscape of diagnosis and treatment of LCH in real-world clinical practice.

This lecture will cover the current status and future perspectives in multicenter efforts to establish better diagnostic and therapeutic strategies for LCH in Korea. In addition, data on genomic alterations of Korean LCH patients will be presented.