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Pre-B-cell colony-enhancing factor is markedly elevated in childhood hemophagocytic lymphohistiocytosis

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ABSTRACT. Hemophagocytic lymphohistiocytosis (HLH) is a life-threatening syndrome involving a final common pathway of hypercytokinemia, in which tumor necrosis factor (TNF)-α, interferon (IFN)-γ, and soluble interleukin 2-receptor-alpha (sIL-2Rα) are the key cytokines. Pre-B-cell colony-enhancing factor (PBEF) is an inflammatory cytokine involved in several inflammatory diseases. However, its role in HLH is unknown. In this study, we examined the role of PBEF in HLH. Plasma was collected from 22 children with HLH and 14 healthy children. The concentrations of plasma PBEF, TNF-α, IFN-γ, and sIL-2Rα were determined using an enzyme-linked immunosorbent assay. All clinical data were derived from medical records. In the acute phase,

children with HLH had much higher PBEF, TNF-α, IFN-γ, and sIL-2Rα levels than did healthy children ($P < 0.05$). After treatment, 13 HLH children improved and PBEF, TNF-α, and IFN-γ levels decreased to normal levels ($P < 0.05$); sIL-2Rα levels also decreased ($P < 0.05$), but remained above the normal level ($P < 0.05$). Two patients were lost to follow-up, while 7 patients showed a bad response to therapy and eventually died, showing high PBEF levels above those of the survivors ($P < 0.01$). PBEF level was significantly positively correlated with TNF-α, IFN-γ, sIL-2Rα, serum ferritin, and triglycerides (all $P < 0.05$), and was negatively correlated with fibrin ($P < 0.05$). PBEF appears to be involved in the inflammatory process of HLH, and elevated PBEF is related to disease activity. We are currently evaluating the role of PBEF as a marker for the diagnosis and management of patients.

Key words: Childhood; Cytokine; Hemophagocytic lymphohistiocytosis; Inflammatory process; Pre-B-cell colony-enhancing factor