A 37-year-old man of Pakistani origin presented with a 5-week history of fever, night sweats, fatigue, nausea, vomiting, lymphadenopathy, and splenomegaly. He had similar symptoms 5 years prior with a cervical lymph node (LN) biopsy showing histiocytic necrotizing lymphadenitis or Kikuchi disease (KD). He had pancytopenia, with a neutrophil count of $0.52 \times 10^9/L$, hemoglobin count of 76 g/L, mean corpuscular volume of 82.2, platelet count of $96 \times 10^9/L$, elevated erythrocyte sedimentation rate of 51, lactate dehydrogenase of 531 U/L, ferritin of $10\,153 \,m\,g/L$, triglycerides of 4.12 mmol/L, alanine transaminase of 277 U/L, aspartate transaminase of 418 U/L, and $\gamma$-glutamyl transferase of 215 U/L. Repeat LN biopsy suggested KD with preserved architecture and patchy fibrinoid necrotizing foci with numerous apoptotic bodies and cellular debris admixed with lymphocytes and histiocytes (panel A; original magnification $\times 40$; hematoxylin and eosin stain). Extensive microbiological, rheumatologic, and hematological investigations, including immunohistochemistry, flow cytometry, molecular tests, etc, were noncontributory but ruled out lymphoma and the acute form of lupus. Marrow was reactive with increased histiocytes showing hemophagocytosis (panel B; original magnification $\times 100$, oil immersion; May-Grünwald Giemsa stain) and dyserythropoiesis including multinucletarity (panel C; original magnification $\times 100$, oil immersion; May-Grünwald Giemsa stain). Cytogenetics was normal. A diagnosis of hemophagocytic lymphohistiocytosis (HLH) with association of KD and dyserythropoiesis was rendered as he fulfilled 6 of 8 diagnostic criteria. The patient was treated with prednisone with complete recovery. He was doing well over 1 year after initial diagnosis.

The association of HLH and KD has been reported in limited cases, as has the association of HLH and reactive dysplasia. The coexistence of HLH, KD, and dysplasia is extremely rare but is worth further investigations.
Dyserythropoiesis due to reactive hemophagocytic lymphohistiocytosis

Zhaodong Xu and Richard Van der Jagt

Updated information and services can be found at: http://www.bloodjournal.org/content/128/17/2190.full.html

Articles on similar topics can be found in the following Blood collections

- BloodWork (573 articles)
- Free Research Articles (4756 articles)
- Immunobiology and Immunotherapy (5527 articles)
- Phagocytes, Granulocytes, and Myelopoiesis (639 articles)
- Red Cells, Iron, and Erythropoiesis (831 articles)

Information about reproducing this article in parts or in its entirety may be found online at: http://www.bloodjournal.org/site/misc/rights.xhtml#repub_requests

Information about ordering reprints may be found online at: http://www.bloodjournal.org/site/misc/rights.xhtml#reprints

Information about subscriptions and ASH membership may be found online at: http://www.bloodjournal.org/site/subscriptions/index.xhtml