

Erythrophagocytosis in Cold Agglutinin Disease

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An 8-year-old boy with B precursor acute lymphoblastic leukemia (ALL) presented with a 1-week history of cough and mild fever during the 107th week of St. Jude total XIII high-risk protocol. Physical examination was normal except rales on auscultation. His hemoglobin level was 11.7 g/dL, white blood count was $2.6 \times 10^9/L$ (absolute neutrophil count 1560), and platelet count was $208 \times 10^9/L$. Peripheral blood film showed several erythrophagocytosis by monocytes (Fig. 1). Direct Coombs' test was positive (2+) with complement (C3d). Cold agglutinin titer was 1:64 and 10 days later it was 1:128. Urine analysis and bilirubin levels were normal. Although thorax radiography revealed pneumonic infiltration, serologic tests for mycoplasma pneumonia, parainfluenza virus, respiratory syncytial virus and also Epstein-Barr virus, rubella, cytomegalovirus, adenovirus were all negative. At follow-up hemoglobin level decreased to a nadir of 9.2 g/L and cold agglutinins disappeared within 3 months.

Cold agglutinin disease (CAD) is an uncommon form of autoimmune hemolytic anemia. The primary form typically affects older adults; however, it is also a secondary disorder in association with a number of different underlying disorders, such as certain infectious diseases (eg, mycoplasma infection and infectious mononucleosis) and lymphoproliferative diseases, especially mature B cell origin such as lymphoma, chronic lymphocytic leukemia, and Waldenström's macroglobulinemia.^{1,2} Very rarely it was described in patients with ALL.^{3,4} Although infection-associated CAD is transient, lymphoproliferative disorders produce a more chronic course. Peripheral smear may show agglutination, polychromasia, anisocytosis, poikilocytosis, and occasionally spherocytosis.^{1,5} Cold agglutinin disease should be borne in mind in patients with erythrophagocytosis on peripheral blood smear.

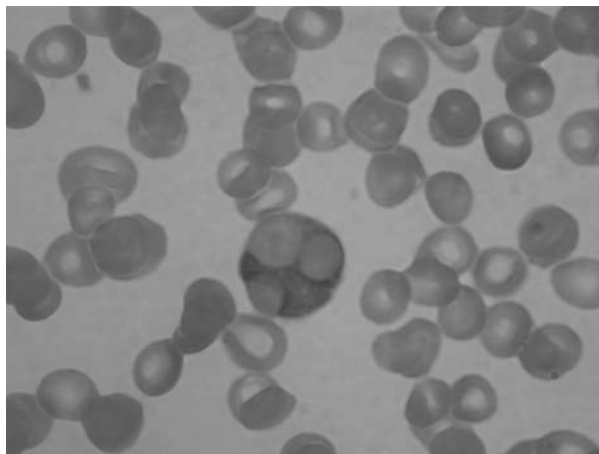


FIGURE 1. Erythrophagocytosis by monocyte.

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