To the Editor:

Paroxysmal nocturnal hemoglobinuria (PNH) is a rare acquired hemolytic anemia characterized by an unusual susceptibility of erythrocytes to complement activities. The abnormal phenomena are now known to be caused by the deficiency of the complement regulatory protein, i.e., decay-accelerating factor (DAF), which prevents the assembly of the C3 and C5 convertases of both the classic and alternative complement pathways on erythrocytes.1,2

The present case, a 20-year-old male, was initially sent to our...
analyses of his erythrocytes and granulocytes obtained at hemolytic and nonhemolytic phases disclosed the same expressions and distributions of DAF (Fig 1) and Ach-E as normal subjects.

The present case showed signs of PNH in the results of both clinical and laboratory studies, while the results of the FACS analysis on erythrocytes and granulocytes revealed no deficiency of DAF or Ach-E. If defined as PNH, this would be, to our knowledge, the first report of a case of PNH without the deficiency of DAF on erythrocytes and granulocytes. Recently, numerous studies of PNH indicated that Ach-E, homologous restriction factor (HRF), and lymphocyte function-associated antigen 3 (LFA-3), as well as DAF, were deficient in erythrocytes. Moreover, DAF, HRF, LFA-3, and Ach-E were determined to be linked to the membrane of normal erythrocytes by phosphatidylinositol. Therefore, it has been suggested that the biosynthetic pathway generating the anchoring mechanisms was defective in PNH. We suggest that there may be different biosynthetic pathways generating the anchoring mechanisms in each membrane protein, or that hemolysis in this case may be caused by unknown mechanism(s). The mechanism(s) of hemolysis in this case is unclear, and further examinations of more cases are necessary.

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REFERENCES


A case of paroxysmal nocturnal hemoglobinuria without deficiency of decay-accelerating factor on erythrocytes [letter]

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