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ACUTE LYMPHOBLASTIC LEUKEMIA IN A CHILD WITH HEMOGLOBINS S AND Q–IRAN *

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Summary: Gürgey A, Özsoylu Ş, Hiçsönmez G, Irken G, Altay Ç. (Pediatric Hematology Unit, Department of Pediatrics, Hacettepe Universty Faculty of Medicine, Ankara, Turkey). Acute lymphoblastic leukemia in a child with hemoglobins S and Q-Iran. Turk J Pediatr 32: 39-41, 1990.

A 13-year-old girl with acute lymphoblastic leukemia is presented. The peripheral smear showed, in addition to lymphoblasts, marked anisocytosis, poikilocytosis, and polychromasia. In vitro sickling test was positive. Hemoglobin electrophoresis at pH 9.0 on starch gel revealed the presence of hemoglobin A, hemoglobin S, and a band with a mobility of hemoglobin A_2. Structural analysis revealed the presence of hemoglobin S and an alpha-chain variant, hemoglobin Q-Iran. The patient attained remission with the initial therapy administered but a relapse occurred five months later. Our study indicates the need for detailed investigation of leukemia patients in which abnormal hemoglobins are prevalent. *Key words: acute lymphoblastic leukemia, hemoglobin S, hemoglobin Q-Iran.*

Recently, hematologic malignancies in patients with hemoglobinopathies have been reported[1,2]. However, to our knowledge, the association of acute lymphoblastic leukemia (ALL) with hemoglobins S and Q-Iran has not been reported in the literature. Therefore, the purpose of this paper is to present such a case with leukemia in whom hemoglobinopathy was detected.

Case Report

A twelve-year-old girl from Southern Turkey with a diagnosis of ALL was referred to the Hacettepe University Children's Hospital for treatment of the disease. On physical examination the child appeared wasted and had a generalized petechial rash. The liver and spleen were palpated three cm and six cm below the respective costal margins.

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Laboratory studies revealed a hemoglobin level of 5.85 g/dl, mean corpuscular volume 92 fl, white blood cell count 150,000/ml. The peripheral smear showed 26 % blasts and thrombocytopenia with marked anisocytosis, polychromasia and poikilocytosis of red blood cells. The sickling test was positive and glucose -6-dehydrogenase activity was normal. The diagnosis of ALL (L 3 by FAB classification) was made by examination of the bone marrow. Since the patient showed signs of hemolysis, hemoglobin electrophoresis was performed which revealed hemoglobin A, hemoglobin S, and a variant with a mobility similar to that of hemoglobin A_2 on starch gel at pH 9.0. The diagnosis of hemoglobin Q-Iran ($\alpha^{75}{}_{B_2}$ Asp → His) and hemoglobin S was made by the structural analysis of abnormal hemoglobins. The child had obtained the hemoglobin S gene from her father, and the hemoglobin Q-Iran gene from her mother (Fig. 1). Remission was achieved with combination chemotherapy of prednisone, vincristine and L-asparaginase followed by cranial x-ray prophylaxis. However, she had a relapse five months later.

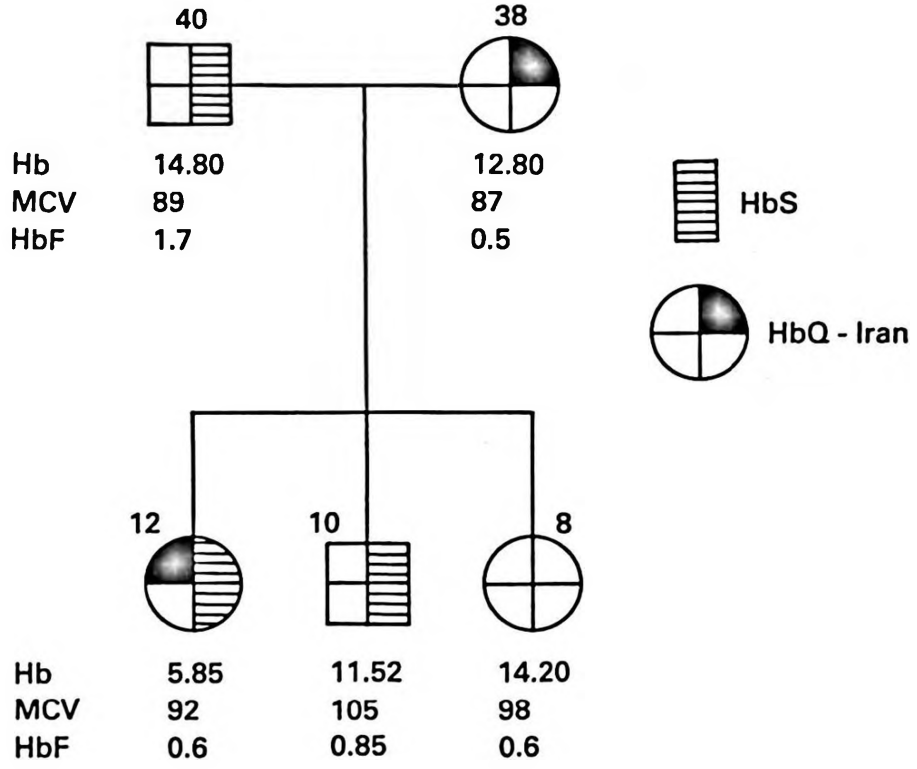

Fig 1: The family pedigree: hemoglobin (Hb; g/dl), mean corpuscular volume (MVC), hemoglobin F (HbF; %).

Discussion

Hemoglobin S and other hemoglobin abnormalities have been encountered in Southern Turkey[3]. Recently, two other families with hemoglobin Q-Iran were also reported from this part of the country[4].

Between 1975 and 1987, 1062 children were diagnosed as having acute leukemia at the Hacettepe University Children's Hospital. Hemoglobin electrophoresis was performed in twenty-two cases, and hemoglobinopathy was noted in three patients; one had Hb-E Saskatoon, another had sickle cell trait, and the third Hb H disease[5,6]. Acquired hemoglobin H disease may be observed in patients with leukemia[7]. In a two-year-old patient that we studied with hemoglobin H, it was impossible to ascertain whether this hemoglobin variant was related to her disease since a family study could not be performed. There are few reports in the literature concerning patients with a hemoglobin abnormality who have developed leukemia.

In the future, routine screening of patients for hemoglobinopathies may disclose whether or not subjects with hemoglobinopathies are more susceptible to developing leukemia than normal children.

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