Simultaneous finding of hereditary spherocytosis in a mother and her daughter following parvovirus B19 infection

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Introduction :

Acute parvovirus B19 infection causes a transient inhibition of erythroid cell formation, which may induce an erythroblastopenia crisis in patients suffering from chronic hemolytic anemia.

We report here the cases of a mother and her daughter who developed at the same time acute erythroblastopenia crisis and severe anemia following parvovirus B19 infection. Both were suffering from hereditary spherocytosis which was not diagnosed before.

Cases :

A 6-year-old girl presented with regenerative hemolytic anemia (hemoglobin level of 4 g/dL, reticulocytes count of 276,000/mm^3) whereas her 32-year-old mother was admitted five days later with aregenerative hemolytic anemia (hemoglobin level of 3.6 g/dL, reticulocytes count of 21,000/mm^3). Both patients presented with a negative direct coombs test.

No history of hemolytic anemia or cholelithiasis was reported in the family.

The peripheral blood smear examinations mainly showed the presence of spherocytes (Fig. 1). Main laboratory findings comprised normal mean cell volume, high LDH, high bilirubin and haptoglobin < 0.2 g/L).

Recent parvovirus infection was showed by the detection of serum parvovirus B19-specific IgM antibodies and viral DNA in the mother and her daughter.

A precautionary bone marrow aspirate taken from the mother confirmed typical parvovirus B19-induced erythroblastopenia with giant proerythroblasts.

An abdominal CT scan showed cholelithiases in the mother that indicated additional chronic hemolysis, along with the splenomegaly and laboratory results of both patients.

Diagnosis of hereditary spherocytosis was confirmed two months later by the screening tests positivity and the electrophoretic analysis of erythrocyte membrane proteins which showed a deficiency of protein 4.2 in the mother and her daughter.

Outcome was good in both patients. Hemoglobin level normalized two weeks after admission without blood transfusion in the daughter. The mother required six erythrocyte concentrates and will undergo splenectomy and cholecystectomy.

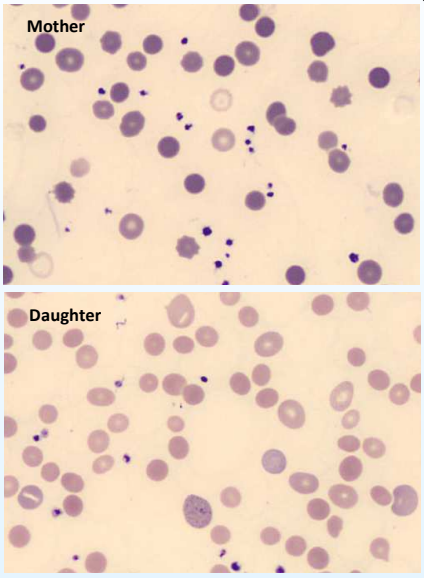

Fig. 1. Peripheral blood smears at admission showed numerous spherocytes in both patients, polychromatophilia, basophilic stippling and stomatocytes in the daughter, then anisochromia and echinocytes in the mother (May-Grünwald Giemsa).

Discussion :

The mother presented during the parvovirus B19-induced erythroblastopenia crisis (low reticulocyte count) whereas the daughter was probably admitted a few days after the crisis when the bone marrow was in a compensatory regeneration phase (high reticulocyte count).

However, such profound anemia was surprising following parvovirus B19 infection alone and erythroblastopenia crises generally occur in patients suffering from chronic anemia.

Furthermore, the fact that the mother presented a few days after her daughter with the similar clinical picture indicated a likely hereditary pathology which needed to be investigated further.

Despite characteristic – but not specific – erythrocytes morphology, hereditary spherocytosis diagnosis was delayed because of negative results obtained with the cryohemolysis screening test in both patients at admission. This was probably due to the profound anemia. The cryohemolysis test was finally positive aloof from the erythroblastopenia crisis.