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A rare condition: subgroup incompatibility due to anti-E

To the Editor

While immune hydrops occurs mostly in relation with ABO and Rh incompatibility (95-97%), it occurs rarely in relation with subgroup incompatibility (3-5%) (1).

Subgroup incompatibilities lead to neonatal hemolytic anemia with varying degrees, prolonged jaundice and rarely immune hydrops. The most common subgroup incompatibilities which lead to blood incompatibility between the mother and the baby include C, c, E, e antigens in the Rh system and K antigene in the Kell system. 14% of the subgroup incompatibilities are related to anti-E antibody (2). The clinical picture related to anti-E mostly consists of mild-moderate anemia and prolonged jaundice; hydrops fetalis is observed rarely (3).

In the literature, cases of hemolytic anemia related to anti-K, anti-E, anti-C subgroup incompatibility have been rarely reported (4,5,6,7,8,9).

In this letter, we wished to report a rare case of hydrops fetalis which developed in the neonatal period as a result of anti-E subgroup incompatibility.

Our patient was born by cesarean section at the 34th gestational week because of pleural fluid and fetal distress with a birth weight of 2200 g as the first live birth from the first pregnancy of a 26-year-old mother (G1A0P1). The patient who appeared pale and edematous was intubated in the delivery room and was internalized in the neonatal intensive care unit. Diffuse edema was found on physical examination. The apical heart beat was found to be 190/min and the blood pressure was found to be 70/40 mmHg. The patient had hepatosplenomegaly and a 3/6 systolic murmur. Neurologic examination revealed marked hypotonia and weak newborn reflexes. The laboratory tests were as follows: the blood type of the baby: 0Rh(-), the blood type of the mother: ARh (+), direct Coombs test: negative. Hemoglobin: 11 mg dL, hematocrite: 34%, lökosit: 9050 /mm^3, platelets: 17000/mm^3 and reticulocyte: 8%. Peripheral smear revealed 55% neutrophils, 33% lymphocytes, 12% normoblasts and fragmented erythrocytes and quartet platalet aggregations. Pathological biochemical tests were as follows: total protein: 2.5 g/dL, albumin: 1 g/dL, Na: 128 mEq/L, total bilirubin: 4 mg/dL. On the postero-anterior lung graphy, fluid was found in bilateral fissures. On thoracal ultrasonography (USG), pleural fluid (13 mm in the right and 11 mm in the left) was observed. On abdominal USG, a small amount of freee fluid was found. Cranial USG was normal. Thoracentesis was performed and transudate fluid was evacuated. Fluid was restricted because of hyponatremia and increased weight and phototherapy was started.

The results of the tests performed to determine non-immune causes were as follows: TORCH-S (-), Parvovirüs B19 (-), EBV IgM (-) and reducing substance in urine (-). Chromosome analysis, metabolic screening tests, hemoglobin electrophoresis, glucose-6-P-dehydrogenase, echocardiography and electrocardiography were found to be normal.

Direct Coomb test was repeated because of exclusion of non-immune causes of hydrops, hemolysis findings and indirect hyperbilirubinemia and was found to be mildly positive. Thereupon, subgroup analysis was performed and anti-E antigen incompatibility was found. Intravenous immunoglobulin (1 g/kg) was administered to the patient.

On the 5th day, ventilatory support and phototherapy were discontinued. On the 13th day, full enteral nutrition was started. During the treatment, albumin was administered for two times, erythrocyte suspension was administered for one time and thrombocyte suspension was administered for two times. On the 21st day, the patient was discharged without any problem. No problem was observed in the outpatient follow-up.

Joy et al. (3) found 283 anti-E incompatibilities in the study they performed between 1959 and 2004. In 32 of these pregancies, anemia in the newborn was found and hydrops was found only in one. In the literature, the rate of anti-E positivity was found to be 0,12% in 43 000 women between 1994 and 1990. In only 5% of these, findings of anemia developed in the baby (10).

In the study performed recently by Karagol et al. (11) in our country, anti-E incompatibility was found in 28.3% of 106

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subgroup incompatibility cases, but hydrops was not found in these cases.

In subgroup incompatibilities, direct Coombs test is found positive with a rate of 33%. A negative direct Coomb test is not a definite indicator of abscence of incompatibility.

Since subgroup incompatibility generally leads to mild anemia in the newborn baby, it can be frequently overlooked. However, it should be kept in mind that subgroup incompatibility may lead to severe anemia and severe hyperbilirubinemia requiring exchange transfusion and rarely to a clinical picture of hydrops fetalis. We wanted to remind of the necessity of further investigations to determine subgroup incompatibility in cases of hydrops fetalis.

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