Hydrops fetalis caused by a blood group antibody usually undetected in routine screening

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Abstract
A case of isoimmunisation in pregnancy caused by antibodies to the Kp* red blood cell antigen is described. The preceding pregnancy had resulted in fetal hydrops for which no cause was found as the antibody screening cells used to investigate the fetal hydrops were Kp* negative. This case emphasises the importance of serological screening at a reference laboratory for low frequency red cell antigens before a diagnosis of non-immune hydrops is made.

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The Penny (Kp*) and Rautenberg (Kp9) antigens are part of the Kell blood group system. The incidence of Kp* is 2% in the white population.1 Anti-Kp* is known to cause haemolytic disease of the newborn but is not usually associated with hydrops fetalis or haemolysis severe enough to require intratuerine transfusion.2

Case report
The serum of a 33 year old woman in her fourth pregnancy was found to contain anti-Kp* (Penny) during her routine antenatal serological screening at 17 weeks' gestation. Her first pregnancy seven years previously resulted in a normal vaginal delivery. Five years later she had a spontaneous abortion at 7 weeks' gestation. In the third pregnancy, intrauterine death of the fetus occurred at 17 weeks' gestation. The fetus was hydropic but no fetal cells were obtained for red cell grouping or antibody identification. No irregular antibodies were found in the maternal serum, and no other cause for the hydrops was identified. She had no history of previous blood transfusion.

Her red cells were phenotyped as group A Rh D positive, Kp(a–b–). Her husband's red cells were phenotyped as group O Rh D positive, Kp(a+b+); being heterozygous for the Kp* antigen, and therefore giving the baby a 50% chance of being Kp* positive. The titre of anti-Kp* was 256 at 17 weeks' gestation, and conservative management was advised with weekly ultrasound scanning for fetal hydrops and maternal blood sampling every two weeks. At 24 weeks' gestation the titre of anti-Kp* had risen to 1000 and at 25 weeks, early fetal ascites was noted by ultrasound scanning. Intrauterine fetal blood sampling revealed a severely anaemic fetus with a haemoglobin concentration of 40 g/l. The direct Coombs test (direct antiglobulin test) was positive and the fetal cells were typed as group O Rh D positive, Kp(a+b+). Spectrophotometric measurement of the deviation in optical density of the amniotic fluid at 450 nm (A450) was unhelpful in predicting the severity of the fetal anaemia.

The pregnancy was managed with regular intratuerine transfusions up to 32 weeks'
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Labour was induced at 35 weeks' gestation resulting in a live male infant.

The cord blood was group O Rh D positive, direct Coombs test positive. The Kleihauer test showed 18% fetal cells and the haemoglobin was 136 g/l. A month after delivery the direct Coombs test was positive and a top-up transfusion was given. The infant required a further top-up transfusion at 2 months of age.

Discussion

The low incidence of the Kp\(^a\) antigen in the population means that red cell screening reagents for routine antenatal screening do not usually carry the Kp\(^a\) antigen. It was fortuitous that the cells in use at the time of this patient’s serological screening at 17 weeks’ gestation expressed the Kp\(^a\) antigen and that the antibody was detected early enough to allow planned management of the pregnancy.

The screening red cells in use at the time of the previously affected pregnancy were Kp(a−b+) and the antibody would therefore not have been identified during this investigation.

The measurement of optical density of amniotic fluid is a measure of the bilirubin content of the fluid. The fact that the ΔA50 of the amniotic fluid was unhelpful in predicting the severity of anaemia was not unexpected as antibodies to the Kell antigens are thought to have a disproportionate effect on the fetal red cell precursors\(^3\) rather than causing haemolysis of red blood cells.

We conclude that in cases of hydrops fetalis where no serological cause is found, serum should be sent to a reference centre for investigation using a panel of red cells expressing low frequency antigens. Identification of antibodies to these antigens allows improved management, as in this case, and counselling regarding future pregnancies. Furthermore, such referral would be likely to identify cases which are otherwise labelled as non-immune hydrops of unknown cause.\(^4\)\(^5\)

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