



Sickle-cell disease in pregnancy: importance of prenatal diagnosis and associated maternofetal morbidity and mortality

Rodrigues M, Marques P, Ferreira C, Meira M, Reis L, Sepúlveda F, Costa A, Nazaré A
Hospital Prof. Doutor Fernando Fonseca, Amadora, Portugal

Objective

Drepanocytosis is a haemoglobinopathy characterized by a change in the structure of the beta chain: replacement of glutamate by valine at position 6 of the chain - Haemoglobin S. It is an inherited disease with autosomal recessive transmission, in which it acquires importance before pregnancy making it necessary to look at both the parents and, eventually, prenatal diagnosis. It has a higher prevalence in black population. On the one hand, pregnancy favours vaso-occlusive crises (VOC) and haemolytic anaemia, which are typical for drepanocytosis. On the other hand, this condition causes a high rate of maternofetal morbidity and mortality: abortion, hypertension, preeclampsia, premature detachment of normally inserted membranes, fetal growth restriction (FGR) and threat of preterm delivery. The objective is to relate the drepanocytosis in pregnancy to maternofetal morbidity and mortality.

Methods

This is a case report.

Results

Pregnant 26-year-old black woman, G0P0, with a personal history of drepanocytosis with necrosis of the left femoral head and hepatitis B carrier status. Drepanocytosis was diagnosed in infancy and had sporadic VOCs controlled with Hydroxyurea, which suspended before becoming pregnant. The pregnant woman was sent to the obstetrics clinic hospital, where the diagnosis of drepanocytosis was proposed. She refused any examination of prenatal diagnosis. Low levels of haemoglobin (6.5-8.4g/dL) were observed throughout pregnancy, as well as frequent VOCs requiring hospitalization and transfusions of erythrocyte concentrates. At 34 weeks of gestation, she was hospitalized for VOC in the setting of acute pyelonephritis and premature rupture of membranes. An emergency caesarean section was determined due to a non-reassuring cardiogram, with the delivery of a newborn female, 1920 grams, Apgar Index 10/9/10, with Drepanocytosis. Puerperium took place without intercurrents.

Conclusion

A typical case of drepanocytosis in pregnancy was described. Anaemia and VOC were present and led to a preterm birth with FGR, as described in the current literature. Drepanocytosis is very important in pregnant women due to the high maternofetal morbidity and mortality.