



GESTATIONAL HEALTH PROBLEMS OF A PATIENT WITH SICKLE CELL ANEMIA: CLINICAL CASE REPORT

AGRAVOS GESTACIONAIS DE UMA PACIENTE COM ANEMIA FALCIFORME: RELATO DE CASO CLÍNICO

PROBLEMAS DE SALUD GESTACIONAL DE UNA PACIENTE CON ANEMIA DE CÉLULAS FALCIFORMES: REPORTE DE CASO CLÍNICO

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ABSTRACT

Objective: to investigate the intercurrent complications within the gestational period and the perinatal outcome of a patient with sickle cell anemia and demonstrate the importance of early and appropriate diagnosis of an inherited anemia during prenatal care. **Method:** this is an exploratory study, a clinical case report, involving the follow-up of a patient with sickle cell anemia by the high-risk pregnancy service of the University Hospital of Universidade Federal de Mato Grosso do Sul (UFMS). The study was approved by the Research Ethics Committee of UFMS, under the Protocol 1,781/2010. **Results:** pain and vaso-occlusive crises were the most common clinical problems, concomitant with severe anemia, premature labor due to rupture of membranes, and hospitalizations for urinary tract infection, and they culminated in delivery of low-birth-weight infant evolving to neonatal death. **Conclusion:** we highlight the importance of guidance for women with sickle cell anemia regarding their descendants and the effectiveness of monitoring by a specialized multidisciplinary team. **Descriptors:** Anemia, Sickle Cell; Hematological Complications in Pregnancy; Perinatal Mortality.

RESUMO

Objetivo: investigar as complicações intercorrentes no período gestacional e o resultado perinatal de uma paciente com anemia falciforme e demonstrar a importância do diagnóstico precoce e adequado de uma anemia hereditária durante o pré-natal. **Método:** trata-se de estudo exploratório, do tipo relato de caso clínico, com acompanhamento de uma paciente portadora de anemia falciforme pelo serviço de gestação de alto risco do Hospital Universitário da Universidade Federal de Mato Grosso do Sul (UFMS). O estudo foi aprovado pelo Comitê de Ética em Pesquisa da UFMS, sob o Protocolo n. 1.781/2010. **Resultados:** a dor e as crises de vaso-oclusão foram os agravos clínicos mais comuns, concomitantes com anemia severa, trabalho de parto prematuro por rotura de membranas e internações por infecção do trato urinário, e culminaram em parto de recém-nascido de baixo peso, com evolução para óbito neonatal. **Conclusão:** destaca-se a importância da orientação às portadoras de anemia falciforme quanto aos seus descendentes e a eficiência do acompanhamento por uma equipe multidisciplinar especializada. **Descritores:** Anemia Falciforme; Complicações Hematológicas na Gravidez; Mortalidade Perinatal.

RESUMEN

Objetivo: investigar las complicaciones intercurrentes en el período gestacional y el resultado perinatal de una paciente con anemia de células falciformes y demostrar la importancia del diagnóstico precoz y adecuado de una anemia hereditaria durante la atención prenatal. **Método:** se trata de un estudio exploratorio, un reporte de caso clínico, con seguimiento de una paciente con anemia de células falciformes por el servicio de embarazo de alto riesgo del Hospital Universitario de la Universidade Federal de Mato Grosso do Sul (UFMS). El estudio fue aprobado por el Comité de Ética en Investigación de la UFMS, bajo el Protocolo 1.781/2010. **Resultados:** el dolor y las crisis vaso-oclusivas fueron los problemas clínicos más comunes, concomitantes con anemia severa, trabajo de parto prematuro por rotura de membranas y hospitalizaciones por infección del tracto urinario, y culminaron en parto de recién nacido de bajo peso, con evolución a la muerte neonatal. **Conclusión:** se destaca la importancia de la orientación a las pacientes con anemia de células falciformes con relación a sus descendientes y la eficiencia del acompañamiento por un equipo multidisciplinario especializado. **Descriptores:** Anemia de Células Falciformes; Complicaciones Hematológicas en el Embarazo; Mortalidad Perinatal.

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INTRODUCTION

In Brazil, there is a variable prevalence of abnormal hemoglobins, due to the intense mixing of population. The sickle cell diseases, hemoglobin C, and alpha and beta thalassemias are the most common hemoglobinopathies.¹

Sickle cell disease is a term used to determine the group of genetic disorders characterized by the predominance of HbS, including sickle cell anemia, which is the homozygous HbS mutation (HbSS), thalassemia interactions with HbS, and the association of other hemoglobin varying with HbS.^{1,2}

Screening for hemoglobinopathies in prenatal and neonatal diagnosis is important to detect asymptomatic carriers, provide proper guidance in each case, in order to minimize clinical, psychosocial, and financial problems related to the disease.

Although pregnancy in patients with sickle cell anemia is not contraindicated, because the risks are not that high, despite the increase in maternal and infant morbimortality, this is a period which requires special attention from the multidisciplinary team, since there is a need to pay attention to the physiological changes and the relationships between these processes and the worsening of patient's condition.¹⁻³

This study aims to:

- Investigate intercurrent complications within the gestational period and the perinatal outcome of a patient with sickle cell anemia.
- Demonstrate the importance of early and appropriate diagnosis of an inherited anemia during prenatal care.

METHOD

Paper prepared through the dissertation **Intercurrent complications in pregnancies and the perinatal outcomes of women with hemoglobinopathies followed up by a high-risk pregnancy service**, presented to Universidade Federal de Mato Grosso do Sul, Campo Grande, Mato Grosso do Sul, Brazil. 2012.

This is a clinical case report, conducted by following up a pregnant patient with sickle cell anemia, cared for at the outpatient unit for high-risk pregnant women at a university hospital in Campo Grande, Mato Grosso do Sul, in 2007. The case was analyzed when conducting a cross-sectional study to investigate intercurrent complications in pregnancies and the perinatal outcomes of

women with hemoglobinopathies followed up by a high-risk pregnancy service.

The study was approved by the Research Ethics Committee of Universidade Federal de Mato Grosso do Sul (UFMS), under the Protocol 1,781/2010.

CASE REPORT

P.A.S., 28 years old, female, born in Campo Grande, Mato Grosso do Sul, Brazil, gestates 2, gives birth to 1, with 1 abortion. In 2007, with 4 weeks of pregnancy, she started her prenatal care in the outpatient gynecology and obstetrics unit at the University Hospital of Universidade Federal de Mato Grosso do Sul (HU/UFMS), in Campo Grande, which has a specialized care for high-risk pregnant women.

Her obstetric past reports a missed abortion at the 20th gestation week, by performing a uterine curettage. She treats a sickle cell anemia since childhood, with episodes followed by hospitalization due to vaso-occlusive crises, hemolytic anemia, heart failure for aortic murmur, with frequent blood transfusions and treatment using iron chelators. She reports not using or having used hydroxyurea within this period.

Among the clinical complications, the pain crisis was the most frequent, followed by vaso-occlusive phenomena, urinary tract infections, and severe hemolytic anemia requiring blood transfusion. Regarding the obstetric problems, there was pre-eclampsia, preterm labor due to rupture of membranes, followed by three hospitalizations for urinary tract infection, besides pain and severe anemia.

As for the postpartum outcomes, gestational age was 35 weeks and 6 days, interrupted by cesarean section due to severe pre-eclampsia. Hypertension and blood transfusion were the events observed.

Regarding the perinatal outcome data, the baby was born in March 2008, she was female and had the anthropometric measurements of head circumference (HC) of 32 cm, chest circumference (CC) of 29 cm, abdominal circumference (AC) of 28 cm, and height of 42 cm. The birth weight was 2,245 kg, characterized as low birth weight, with first minute Apgar score of 9 minutes and fifth minute of 10. She showed a dermatosis and was referred to the Neonatal Intensive Care Unit, where she evolved to fetal death at the seventh day of life due to infection.

The patient was discharged from hospital at the fourth day, returning to the immediate medical treatment sector five days after

discharge, with mediate postpartum complications due to bleeding in the surgical incision, followed by an extensive hematoma around the abdomen and a subaponeurotic hematoma, when she underwent drainage and blood transfusion with two units of packed red blood cells. She was referred to the obstetric clinic for treatment, where she was discharged after seven hospitalization days.

DISCUSSION

The occurrence of maternal and perinatal mortality has declined in recent decades due to the public health programs deployed, although there is a high risk of multiple maternal and fetal complications associated to hemoglobinopathies, especially SS.

Hemoglobinopathies may be aggravated by pregnancy, especially with the worsening of anemia and increased frequency of painful crises and infectious processes, besides adversely affecting the evolution of pregnancy.³

In Jamaica, a study with pregnant women with sickle cell anemia showed that only 57% of pregnancies had a good development, with live births, compared to 89% in controls, when miscarriage was the most frequent cause of fetal loss in the group under study.⁴

Another study involved 95 pregnancies of 43 patients with SC hemoglobin, 94 pregnancies of 52 women with SS hemoglobinopathy, and 157 pregnancies of 68 controls; when we observed a more benign clinical course for the SC pregnant women when compared to the SS ones. However, pregnancy in patients with SC hemoglobinopathy can precipitate the onset of disease complications, which, until then, were oligosymptomatic or even asymptomatic.⁵

Studies have shown that these complications can be effectively minimized through an adequate prenatal care. In a study conducted for 20 years at the Grady Memorial Hospital (Atlanta, Georgia, USA), it was verified by observation of patients with SS and SC hemoglobinopathies the following gestational complications: pre-eclampsia, prematurity, preterm premature rupture of membranes, preterm labor, intrauterine growth restriction, low birth weight, and increased risk for fetal losses (miscarriages and stillbirths).^{5,6}

Epidemiological studies provide a broader understanding of the occurrence and the course of sickle cell anemia, besides allowing us to assess direct and indirect consequences

of the disease, such as impaired individual, family, and social functioning.

Pregnant women suffering from the severe forms of S hemoglobinopathy must be followed up on frequent prenatal consultations, beginning at the first quarter, and, preferably, by a good multidisciplinary team.⁷

CONCLUSION

The gestation period of the female patient with hemoglobin SS was complicated both by complications related to primary disease and by obstetric problems. We highlight the vaso-occlusive crises, the infections, and the hypertensive disease specific to pregnancy. The worst perinatal outcome was characterized by the incidence of prematurity and low weight associated to the neonatal infection leading to fetal death.

Within the gestational period, despite the carriers of SS hemoglobinopathy show complications, we may expect a moderate gestational outcome both for the mother and the fetus when there is follow-up and treatment with an adequate approach during pregnancy and puerperium, with decreased morbimortality of pregnant women and newborn infants, something which might have been expected in this case.

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