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Case Report

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A case of sickle cell disease during pregnancy: by the eyes of an internist

Abstract

Sickle cell disease is an inherited monogenic disease characterized by misshapen red blood cells that causes vaso-occlusive disease, vasculopathy, and systemic inflammation. We report a case of a 24 years old black female with sickle cell disease (SCD), (hemoglobin S 30%), at her twelfth week of pregnancy, with fever, myalgia, productive cough and leucorrhea. Chest radiography demonstrated bilateral infiltrates and a microcytic anemia of 7.4g/dL. The patient was hospitalized with the diagnostic of acute chest syndrome and treatment with ceftriaxone 1g/day, hydration and analgesic therapeutic to pain relief. However, because of resistant abdominal pain to paracetamol and tramadol and laboratory studies with hemoglobin value of 7g/dL, a compatible blood transfusion was done. Despite the better recognition and diagnosis, pregnancy in SCD is associated with higher clinical and obstetric complications compared to healthy individuals. So, this case demonstrates the importance of an access to multidisciplinary care team to decrease significantly the morbidity and mortality of these cases.

Keywords: pregnancy, sickle cell disease, anemia in pregnancy, blood transfusion, abdominal pain

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Introduction

Sickle cell disease (SCD) is the most common inherited hemoglobinopathy, counting almost 300.000 neonates each year, predominantly from Nigeria, India and Democratic Republic of Congo.[1] With the present immigration flow, it stared to be seen worldwide with increased prevalence. The term sickle cell disease includes different genotypes of homozygous Hemoglobin S sickle cell anemia (SCA) and a variety of double heterozygote states of sickle hemoglobin disease. SCA is cause by homozygous mutation and presents as chronic anemia with painful episodes throughout life due to impaired microcirculation by sickling of erythrocytes. Despite the better recognition and diagnosis, pregnancy in SCD is associated with higher clinical and obstetric complications compared to healthy individuals.[2,3]

Case report

We describe a case of a 24 years old black female with sickle cell trait (hemoglobin S 30%) with no other previous medical record. Without chronic medication or allergies described. She went to the emergency department during her twelfth week of pregnancy because of fever (max 101ºF), myalgia, productive cough and leucorrhoea with five days of evolution. Two days before has started to felt abdominal pain predominantly in the left iliac and lumbar region. On examination she was pale, hemodynamically stable and with no fever. No abdominal pain was observed. The gynecologic exam showed thick and curdy white leucorrhoea compatible with fungal infection. Her cervix was closed and painless. The fetal echography showed good cardiac and corporeal vitality with normal amniotic fluid. She presented a microcytic anemia of 7.4g/dL with no other remarkable analytic alterations. She was admitted and hours later complained of chest pain. The chest radiography showed bilateral infiltrates and was decided to treat with ceftriaxone 1g/day for 7 days. On the third day of hospitalization she had resistant abdominal pain to paracetamol and tramadol and her hemoglobin was 7g/dL. Was done a compatible blood transfusion with relief of pain in hours after transfusion. The fungal infection was treated with topical Clotrimazole. No fever was observed from the second day of hospitalization, pain was solved with no need for analgesics on discharge and no hemolysis was seen during the all stay in hospital. She was closely followed in routine consultations and ended given birth at 37weeks of gestation with no associated complications.

Discussion

Both Mother and child, during pregnancy are at increased risk of morbidity and mortality because of increased metabolic demand, increased blood viscosity and aggravated hypercoagulability. Those factors lead to increased incidence of acute chest pain syndrome, vasoocclusive crisis, hepatic and osteonecrosis, thromboembolic events and ulcers. The vasoocclusion can also occur in the placental microcirculation leading to infarction and necrosis causing fetal hypoxia and adverse fetal outcomes due to impaired uteroplacental circulation.[4–6] Abortion is observed up to 26%, maternal death up to 11.4%,[7] severe anemia in 73% of cases[8] and preeclampsia up to 56%.[9] The incidence of pre-eclampsia and eclampsia is higher in SCD patients.[10] Occlusive vessel crisis usually lead to hospitalization due to bed rest and adequate fluid intake. Pain management with acetaminophen and nonsteroidal anti-inflammatory drugs (NSAIDs) can be given.[11] Pethidine should not be used due to higher incidence of seizures.[12]

Acute chest syndrome should be suspected when pregnant women complain of severe cough. The chest x-ray will show bilateral infiltrates and respiratory infection should be ruled out. Antibiotics, oxygen support, adequate hydration, analgesics and even blood transfusions can be given.[11] When the pregnant SCD women presents

with chest pain and respiratory distress but no infiltrates showed on chest radiography, pulmonary thromboembolism must be a diagnostic to exclude. Ddimer are not confirmatory of diagnosis, and treatment with low- molecular-weight heparin should be started promptly.[11] In case of neurological impairment, stroke must be ruled out. The treatment goes through emergency blood support.[11] In case of sepsis or infection, anemia is expected to worsen due to fever and acidosis. Prompt antibiotics should be given, depending on the source of infection.

It is known that anemia is the most common complication during pregnancy, but blood transfusion is only indicated when hemoglobin falls down of 7g/dl. Each hospital should have a protocol for the management of anemia during pregnancy in SCD including the use of transfusion therapy. The standard of care for blood supply therapy during pregnancy should be as it is to the general population.[11,12] Information should be given to all SCD women during reproductive age about complications during pregnancy and what should be done to minimize them. Vaccination against all encapsulated organisms should be given.[12] Folic acid and iron should be supplemented.[13] To prevent pre eclampsia, the use of low-dose aspirin 75mg once daily from 12 weeks of gestation should be given.[12] Hydroxyurea and angiotensin converting enzyme inhibitors should be discontinued at least 3 months prior to conception due to teratogenic effects.[14] During the first two quarters, women should undergo a screening for urinary tract infection and pre-eclampsia. From 28 weeks of pregnancy, a monthly echography and weekly cardiotocography should be performed.

Conclusion

Although most pregnancies complicated by maternal SCD are likely to result in healthy newborns, these pregnancies are at increased risk of obstetrical/fetal complications. These risks occur due to the metabolic demands, hypercoagulable state, and vascular stasis associated with pregnancy. With the present immigration flow, the prevalence of pregnant with SCD is increasing which demands an access to a multidisciplinary care. The role of close follow up and knowledge in this disease and its complications is decreasing morbidity and mortality in both mother and newborn.

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Conflicts of interest

The author declares there are no conflicts of interest.

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