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Drepanocytosis and Pregnancy: About a Clinical Case

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Abstract | **Case Report**

Sickle cell disease is a genetic disorder characterized by the presence of abnormal hemoglobin: Hb S whose polymerization in the deoxygenated state is responsible for the clinical manifestations of the disease: anemia and vaso-occlusive seizures. The occurrence of a pregnancy in a sickle cell woman is a risk situation for both the mother and the fetus, in view of the worsening clinical signs of the disease, to which are added obstetric complications. We report, a case of sickle cell disease in a woman of 22 years, G1P1, without significant pathological antecedents, followed at the gynecology department of the Military Hospital Moulay Ismail MEKNES with a gestational age of 12 weeks of amenorrhea. In her systematic pregnancy assessment, the blood count showed leukocytosis at 31000 / mm3, normochromic regenerative normocytic anemia (Hb: 8 g / dl, VGM: 80fl, TCMH: 29.2 pg, MCHC: 30.1 g reticulocytes: 145,000 / mm3). In front of this leukocytosis, a colored blood smear with MGG is carried out and showed the presence of 70% of circulating erythroblasts and the presence of numerous sickle cells. A sickling test is done and found to be positive. The diagnosis of sickle cell disease is confirmed by electrophoresis of hemoglobin in an alkaline medium and in an acid medium. It is a homozygous S / S sickle cell disease. The delivery was conducted under strict vagal supervision without complications. The association sickle cell and pregnancy is a risky situation, and the fortuitous discovery (our case) shows the interest of the biolologist both in the diagnosis and in the follow-up of the disease, hence the interest of close collaboration between clinician and biologist.

Key words: Sickle cell disease, pregnancy, Hb S, electrophoresis of hemoglobin.

INTRODUCTION

Sickle cell disease is a genetic disorder of hemoglobin, characterized by the presence of abnormal hemoglobin S at high concentrations in red blood cells [1]. It is responsible for the clinical manifestations of the disease: anemia and vaso-occlusive attacks [2]. Pregnancy in a sickle cell is a maternal-fetal risk because of the aggravation of the clinical signs of the disease, to which obstetric complications are added [3,4].

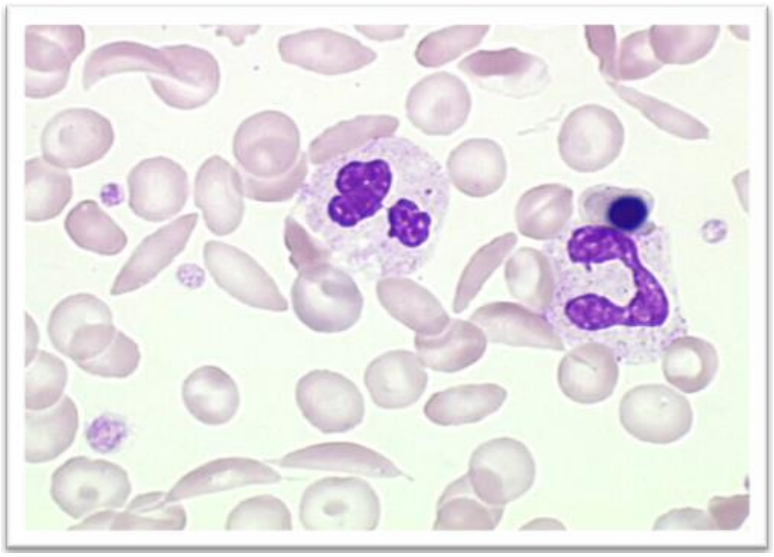
The objective of our work is to show the interest of the biologist in the diagnosis and the follow-up of these pregnancies in collaboration with the clinician.

OBSERVATION

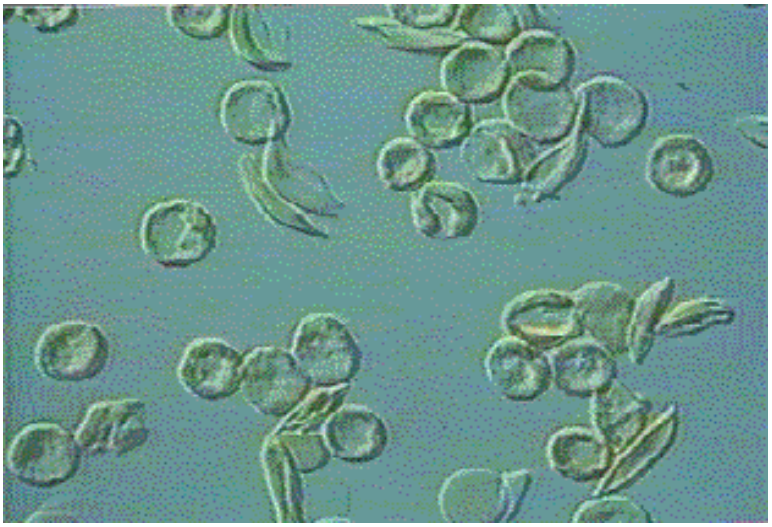
This is a 22-year-old woman, G1P1, with no notable pathological antecedents, followed at the Gynecology Department of the Moulay Ismail Military Hospital MEKNES with a gestational age of 12 weeks of amenorrhea.

In her routine pregnancy checkup, the blood count showed leukocytosis at 31 103 / mm³, normochromic regenerative normocytic anemia (Hb: 8 g / dl, VGM: 80fl, TCMH: 32 pg, MCHC: 31 g / dl reticulocytes: 145,000 / mm³).

In front of this leukocytosis, a blood smear stained with MGG is carried out and showed the presence of 70% of circulating erythroblasts (picture1) and the presence of numerous sickle cells (picture1). The corrected leukocyte count was 7.1 103 / mm3. A sickling test was done and found to be positive (picture2).



Picture-1: blood smear stained with MGG showing circulating erythroblasts and sickle cells



Picture-2: Positive sickling test

The diagnosis of sickle cell disease is confirmed by electrophoresis of hemoglobin in an alkaline medium and in an acid medium. It is a homozygous S / S sickle cell disease. The delivery took place under strict vaginal monitoring without complications

DISCUSSION

Sickle cell disease, homozygous (Hb SS) or heterozygous composite (Hb S / B thalassemia, Hb S / C, Hb S / O Arab ...), is a genetic disease of hemoglobin. It is characterized by the presence of abnormal hemoglobin S at high concentration in red blood cells. It represents, with thalassemias, the most common genetic pathologies of the planet [5]. It is an autosomal recessive disorder [1] of variable expression. It results from the mutation on chromosome 11 of the sixth codon of the beta globulin chain of hemoglobin (substitution of a glutamic acid by a valine) with formation of Hb S [6, 7].

Maternal complications of sickle cell women during pregnancy are common, especially during the last trimester, during labor and in the postnatal period [3]. Anemia is constant, and may be increased by haemodilution and haemolytic seizures of infectious origin. Thromboses are frequent in late pregnancy and predominate in areas of microvasculature. Bone manifestations of vaso-occlusive seizures are all the more serious as they can be complicated by acute thoracic syndromes, renal manifestations ranging from simple microscopic hematuria to vasculorenal syndrome (1/4 of cases), and preeclampsia. Infections are common [8, 9].

Placental hypoxia favored by chronic anemia, placental microthromboses reducing maternal-fetal exchanges, are the cause of intrauterine growth retardation and fetal death. Late miscarriages and prematurity are most often due to infections [8, 9].

Due to these multiple maternal-fetal complications, the pregnancy of sickle cell women requires special care within a multidisciplinary team (gynecologist, biologist, midwife, pediatrician, dietician) with a specific biological monitoring and narrow comprising other elements that add to normal pregnancy monitoring parameters [2-4, 10].

Anemia: the fortuitous and delayed discovery of sickle cell disease in our patient required transfusion to correct anemia and reduce blood viscosity thus limiting the occurrence of vaso-occlusive attacks. The purpose of this transfusion is to have a Hb S less than 35-50%. Monitoring is by hemoglobin level, capillary electrophoresis in alkaline and acidic medium, and by high performance liquid chromatography (HPLC). The monitoring of this anemia continues postpartum.

Obstetrical complications: Sickle cell pregnant woman is likely to develop complications; usually anemic attacks, painful seizures, urinary tract infections, lung infections, proteinuria, high blood pressure, and even maternal death [11- 15]. This explains the benefits of 24-hour proteinuria screening, evaluation of renal and hepatic function (creatinine, transaminase, GGT and ALP), monitoring of thrombocytopenia and serum uric acid, as well as urinary tract infection testing.

Couples at risk: The electrophoresis of hemoglobin in the husband is essential in order to identify couples at risk, to be able to offer genetic counseling [16] and the use of antenatal diagnosis.

Antenatal diagnosis: It can be performed in couples of heterozygotes. Molecular biology allows diagnosis at the 10th week of pregnancy, from a chorionic villous biopsy, or sometimes by fetal cell isolation in the maternal blood (by flow cytometry). Molecular techniques study DNA. The mutation of the gene carries on a cleavage site of the restriction enzyme Mst II: the site is no longer cleaved after enzymatic digestion, an abnormally long DNA fragment is obtained, which can be amplified with specific oligonucleotides. of the PCR mutation region [17] (this was not done in our case).

Diagnosis at birth: Allows management before the appearance of clinical signs. Is carried out on drop of blood recovered on blotting paper and recovery of the hemolysate. Iso-electro-focusing techniques are more efficient at identifying very moderate fractions with less than 10% HbS [17].

CONCLUSION

The association sickle cell and pregnancy is a high-risk maternal-fetal situation, and the incidental discovery (our case) shows the interest of the hematologist both in the diagnosis and in the follow-up of the disease. Hence the interest in close collaboration between clinician and biologist.

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