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Case Report

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β –THALASSEMIA AND PREGNANCY ABOUT 1 CASE

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BACKGROUND

Beta-thalassemia major is a hemoglobinopathy of genetic origin, more widespread in the Mediterranean basin. Patients have a longer life expectancy but are often infertile. Pregnancies are very rare but preimplantation diagnosis is possible. They are then considered as high risk and must be managed by a multidisciplinary team.

Case presentation

Thanks to therapeutic advances, the life expectancy of patients with b-thalassemia major is now 40 years. We report the case of one of our patients followed in our department first for secondary infertility and then for her pregnancy and we propose to review the literature on the management of these patients.

Conclusion

The importance of the preconception visit is reaffirmed in order to educate the population in a preventive manner before pregnancy to avoid complications.

KEYWORDS: thalassemia, pregnancy, infertility, complications.

CASE REPORT

31-year-old Mediterranean female patient, with a personal pathological history of an appendicectomy, a cholecystectomy and a $\beta -$ thalassemia since 12years old, for which she has been regularly treated with hydroxyurea (500 mg) 3 tablets/day and iron supplementation, presenting repeated vasoocclusive crises. The current obstetric history of the pregnant woman during her first admission to the hospital is as follows: G4 P 2 (2 spontaneous abortions), with date of last menstrual period on November 8, 2019, for a gestational age at hospital admission of 37 weeks. She refers that three weeks earlier she had two episodes of syncope so she went to the on-call department of the regional hospital in rabat and it was decided to admit her to the Hematology ward, during her stay she had 2 transfusions and analgesics for her pain in the extremities. A second partial exchange transfusion was performed one week after but she continues to have pain of variable intensity, an ultrasound was performed which showed a splenic infarct with hepatomegaly, and a normal heart function.

Results of biological analysis of her admission

Regenerative hypochromic microcytic anemia with pseudopolyglobulia and hemolysis

hg=9,8g/l → hg=9,8g/dl
hematocrit=28%
Reticulocyte= 120,9 $10^3/mm^3$
VGM= 70 fl
CCMH= 22,5g/dl
Red cells= red cells=4,6. $10^{/}/mm^{3.}$→4,6. $10^{6/}/mm^3$
White cells =18 $000/mm^3$
monocyte = $5/mm^3$
Eosinophil=0,9%
Lymphocyte= $3500/mm^3$
TP=100%
Fibrinogen=3,4g/l
LDH=406mmol/l→LDH=406U/lBilirubin total=32mg/l
Bilirubin indirect=23mg/l
Electrophoresis of the hemoglobin post transfusion (figure 1): HbA : 0% , Hb F : 94%, Hb A2 : 6%-->HbA: 95%
HbF: 2,2% HbA2= 2,7%

Target cells were found in the blood frosting

Painful crises are maintained attenuated with different treatments of oral analgesics until she is transferred to our department, then, throughout her admission an obstetrical ultrasound was performed which revealed an amniotic fluid index at 8 with a fetal anasarque (figure 2-3-4) and a high systolic velocity ratio of the ACM (cerebral artery).

For this reason, it was decided to terminate the pregnancy by a c-section which resulted in a live newborn of 3500 grams with Apgar 8-9, who evolved satisfactorily. The patient underwent surgical sterilization and evolved well, a change in treatment with heparin to oral anticoagulants was indicated, and she was discharged with hematology control. The new born was admitted to the neonatology department for more investigations.

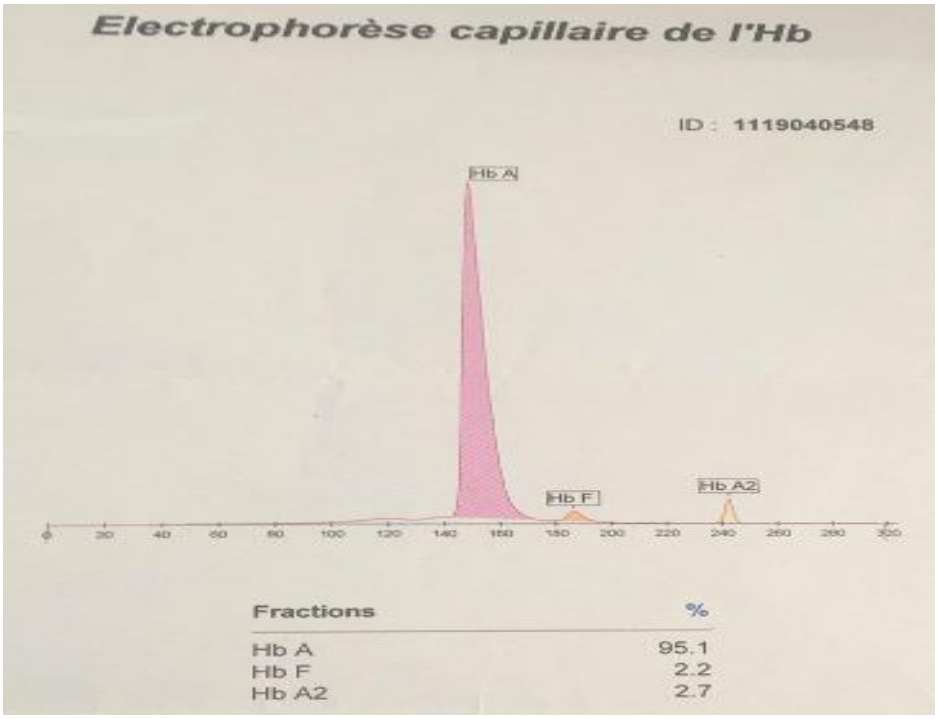

Figure 1.

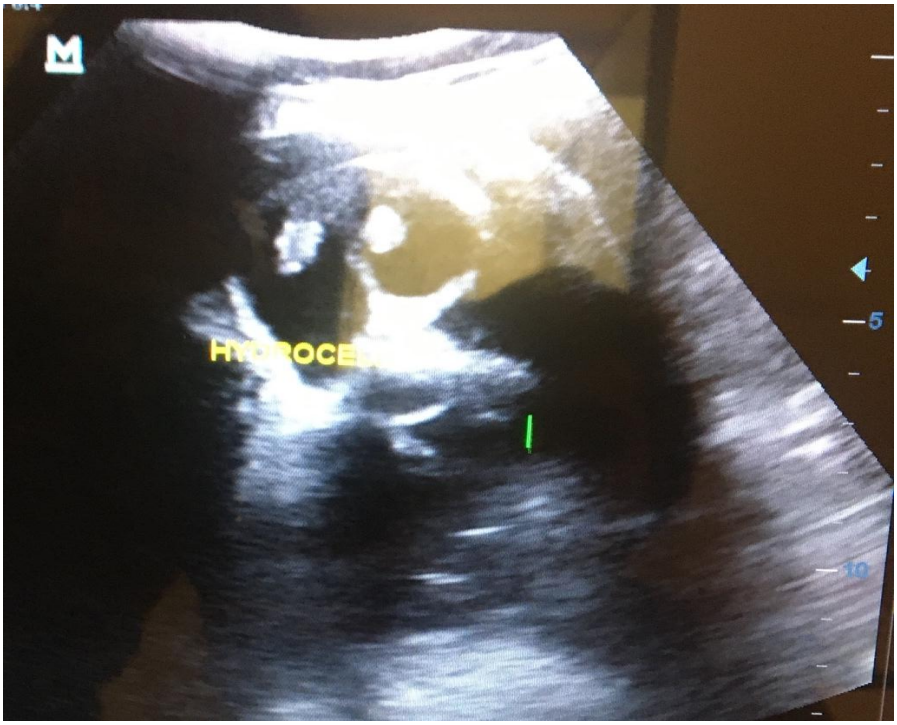

Figure 2.

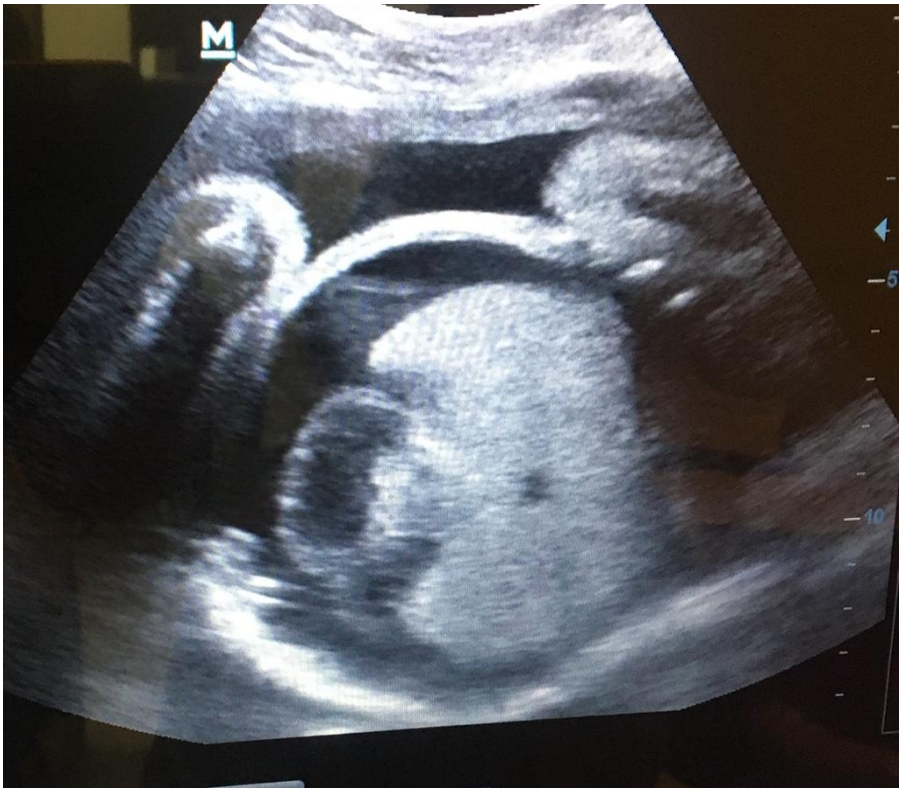

Figure 3.

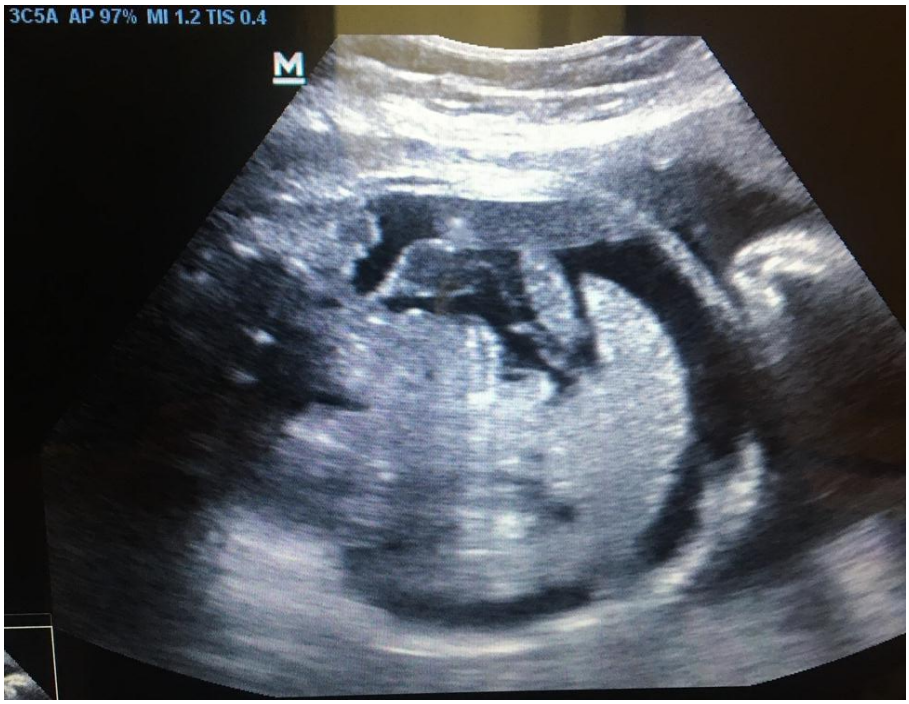

Figure 4.

DISCUSSION

During pregnancy, hemoglobinopathies can lead to maternal and perinatal complications that's why they are considered as high-risk pregnancies.

The variations in the type of presentation and complications that can occur in each case of thalassemia depends on the number of genes that are concerned[1-2] it results in the destruction of red blood cells, which leads to the frequent occurrence of anemia and obstructions.[4-10]

It is estimated that two hundred million people worldwide are born annually with this heterozygotes type and three hundred thousand people with the homozygote

type, these are of higher incidence than other serious congenital diseases such as congenital hypothyroidism and phenylcetonurisme.[9]

Thalassemia major is rarely a problem for pregnancy, since the iron overload associated with this pathology usually results in pubertal failure to thrive and delayed sexual development.

Hemoglobin electrophoresis is of interest to detect this disease in the heterozygous state in patients from these regions. In the homozygous state, there is a delay in height and weight and a mongoloid aspect of the face due to the increase in the volume of the bones of the skull and the face (medullary hyperplasia), and hepatosplenomegaly. Spontaneous evolution leads to death within two to five years.

The main complication in a study that interests 14 pregnancies is fetal losses (five deaths) and it had concluded that pregnancy may precipitate in heart failure in these cases[15] but it is not only dangerous to the fetus but also to the mother who may suffer significant sequels.[13-7]

Complications are currently related to post-transfusion martial overload. It affects the heart, liver and endocrine glands. The use of (deferoxamine-Desferalt) has increased the life expectancy of these patients. It was 15 to 20 years in 1970, it is currently 40 years.

A wide range of laboratory tests have provided solid evidence of the existence of a chronic hypercoagulable state in thalassemia, particularly in splenectomies patients. The presence of a persistent hypercoagulable state with an infrequent occurrence of thrombotic events suggests that this phenomenon in thalassemia is subclinical and has been associated with autopsy findings of thrombosis in the pulmonary and cerebral vessels.[11]

Our patient had to undergo exchange transfusion in early pregnancy. The prophylactic use of transfusion or exchange transfusion is debated. Those who support this treatment claim that decreasing the percentage of Hb S decreases placental vaso-occlusive complications, and maternal and fetal morbidity and mortality, the others find no difference. On the other hand, 22% of transfused women develop anti-erythrocyte allo-antibodies, some of which may cause hemolytic anemia of the newborn as we sow it in our case.

The purpose of this presentation is to highlight the importance of monitoring this group of women with hematological conditions, mainly in primary health care.

Preconception consultation is of great importance for the adequate genetic counseling The typical picture of the impact of the disease on fertility is hypogonadism due to hemosiderin deposits on the hypothalamus and pituitary gland so Patients have anovulation and primary or secondary amenorrhea[1]

Besides, spontaneous pregnancies are rare. Studies published from 1981 to 2001 report 83 pregnancies in 75 women and 25% of these pregnancies are obtained by medically assisted reproduction[2-8] These pregnancies are high risk and are authorized after a multidisciplinary clinical and in particular cardiological assessment.

The association of folate treatment is recommended to avoid additional megaloblastic anemia. Trans-fusions should maintain a hemoglobin of 10 g, but there is a risk of sudden volume overload and heart failure.

Advances in genetics open up new horizons and a very important field of preventive action for Public Health. In the United States, Public Health Genetics has been defined as the application of advances in genetics and molecular biotechnology to improve public health and prevent diseases.[12-5]

Despite the benefits that the periconceptional consultations would bring, we know they are hardly attended, since more than half of pregnancies are not scheduled, so we insist on the importance of systematic preconception visits to educate the population on the importance of prevention in mother-child health.

CONCLUSION

Thanks to the progress in medicine, the life expectancy of patients with thalassemia can only increase, Moreover, pre-implantation diagnosis is possible, the children born will not be sick. Pregnancies are at high medical risk for the mother and the child. They must be programmed if possible and require multidisciplinary care.

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