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Case Study

Hemoglobin Electrophoresis - A Stitch in Time Saved Beta thalassemia Mother and Newborn

Preeta Kurvattigoudar[1]* and Kishanrao Suresh[2]

[1]Karnataka State Rural Development and Panchya Raj University (KSRDPRU), Gadag, Karnataka, India

[2]Visiting Professor-MPH, KSRDPRU, Karnataka State Rural Development and Panchya Raj University (KSRDPRU), Gadag, Karnataka, India

***Corresponding Author:** Preeta Kurvattigoudar, Karnataka State Rural Development and Panchya Raj University (KSRDPRU), Gadag, Karnataka, India.

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Abstract

Hemoglobinopathies are inherited disorders of red blood cells,an important cause of morbidity and mortality, as they impose a heavy burden on families in the absence of a antional program. People with Beta thalassemia minor exibit mild anemia needing no medical treatment but if the client be a lady may fall short of Hb% during pregnancies. We report one such case of Beta-thalassemia. An young female of 30 yrs, married for 4 years a housewife reported happily with amenorrhea for 2.5 months and a positive pregnancy test using a rapid test kit at home for routine check-up, follow-up, expecting normal outcome. She revealed normal menstrual history but the exact LMP was not known. Her past history indicated delayed developmental milestones following her mother's death an obstructed delivery by outlet forceps.

Physical examination revealed a uterus of 16 weeks size, urinary pregnancy test reconfirmed the pregnancy and all other findings were normal. Gestational diabetes was ruled out by a GTT. Investigation reports showed that Hb was 8.2 gm% with MCV count 69fL, WBC count was 7400 cells/ cumm and Platelet count was 301000 cells/cumm, Red blood cells count was 3.7 million. HIV and HbsAg were also negative and the patient blood group was "O" positive. Cardiotocography showed FHS (Fetal Heart Sound) ranging from 150 - 160 beets/min. An anomaly scan reported that a single live intrauterine fetus of gestational age 17wks 2 days +- 8 days with no obvious anomalies. And EDD (Expected Date of Delivery) was given on 28 September 2020. T. Iron and a multivitamin -Bd for 30 days. And T. calcium carbonate 600 mg + T. Vitamin D3 400 Iu - od for 30 days was given. Same follow-up was repeated, but Hb% did not improve after 3 more months. An USG was normal. Hemoglobin electrophoresis confirmed that she had Beta thalassemia trait in the 7th month of the pregnancy. She was given Inj. OROFER FCM 500 mg IV that improved the Hb% level to 10 gm%. As a result she underwent a full term lower segment Cessarian Section (LSCS) on 21st September 2020, No complications durng the entire process of LSCS. Both mother and baby with birth weight of about 2900 gms were healthy. No further peurperal complications were observed. Now the baby turned to 8th month with no such issues and Hb% of the mother mentained same as 10 gm% and the growth and development of the newborn are normal.

Keywords: Hemoglobin Electrophoresis; Hereditary; Diabetes Mellitus

Introduction

Beta-thalassemias are a group of hereditary blood disorders characterized by anomalies in the synthesis of the beta chains of hemoglobin resulting in variable phenotypes ranging from severe anemia to clinically asymptomatic individuals. The total annual incidence of symptomatic individuals is estimated at 1 in 100000 throughout the world. The overall prevalence of β-thalassemia in India is 3-4% with an estimate that around 10,000-12,000 chil-

dren are born every year with β-thalassemia major. India has the estimated number of about 1 to 1.5 lakhs Thalassmia major cases and almost 42 million carriers of ß (beta) thalassemia trait. About 10,000 -15,000 babies with thalassemia major are born every year [1,4]. In India, the prevalence of α-thalassemia is estimated to be around 12.9%. The evaluated prevalence of β-thalassemia minor (carriers) in India is 3-8%, which transforms to 35-45 million carriers in 1.3 billion diverse population with culturally and linguistically and multi-ethnic (about 8% of tribal groups). The overall prevalence of α thalassemia carriers (single α gene deletion) is around 13% varying from 3% to 18% based on caste populations and is very reported to be high in some tribal groups reaching over 90% in some groups [3,4]. The current guidelines for Hemoglobinopathy carrier screening in adolescents is combined with screening and treatment of anemia. Antenatal screening of hemoglobinopathies are to be integrated with testing for HIV, hepatitis B, VDRL diabetes mellitus, hypothyroidism etc.

Though Thalassemia presents with a typical type of anaemia, a simple CBC test with MCH can be the first screening test in pregnancy for carrier status. However, the β-thalassemia trait is usually diagnosed by hemoglobin electrophoresis. Hemoglobin electrophoresis shows a preponderance of HbC and variable quantities of HbA depending on whether it is a β+ or β° thalassemia allele [4-6].

Case in Study

A female of 30 yrs, a housewife married 4 years back, reported with history of amenorrhea of 2.5 months in March 2020, after a pregnancy test using a home based rapid diagnostic kit. She had normal menstrual history since menarche. Initially she had complaints of excessive hunger and occasional vomiting since January 2020. LMP was not known.

On Physical examinations findings were: pulse rate 70/min, BP - 110/80 mmHg and Temperature was normal. Abdomen palpation confirmed a gestation of more than 16 wks of pregnancy. The urinary pregnancy (UPT) test confirmed the pregnancy.

Investigations done

- A GTT (Glucose Tolerance Test) excluded gestational diabetes (107 mg/dl).
- Hb was 8.2 gm% with MCV count 69fL
- CBC- WBC count - 7400/mm^3 and Platelet count -s 301000/mm^3, RBC count 3.7 million.
- HIV and HbsAg were also negative and the patient blood group was "O" positive.
- Cardiotocography (CTG) inidctaed Fetal Heart Sound from 150 - 160/min.
- Foetal Anomaly scan: A single live intrauterine fetus of gestational age 17wks 2 days +/- 8 days with no obvious anomalies.
- Initial Diagnosis: A normal pregnancy with EDD of 28 September 2020.

Treatment given

- T. Iron and Multivitamin -Bd for 30 days and
- T. Calcium carbonate 600mg + T. Vitamin D3 400 Iu - od for 30 days.

Follow-up

- After one month Haemoglobin level and USG scanning remained the same.
- Up to 3 months same treatment plan was followed and
- At gestation of 29 wks Hemoglobin remained 8.6gm% and the USG report was normal.

Further investigation was ordered as Hb% did not improve with oral Haematinics.

Hemoglobin electrophoresis (28-30 weeks) revealed elevated HbA2 and HbF levels.

To conclude to be a case of Beta thalassemia trait.

Revised management plan

- Inj. OROFER FCM 500 mg IV for one shot to improve Hemoglobin. After this IV infusion Hemoglobin level raised to 10 gm%.
- On 21st September 2020 elective LSCS was done.
- A baby girl with a 2900 gms birth weight was born
- No post-partum hameohhage or puerperal sepsis reported mother recouped well.
- After 5 days hospital stay with no complications she was discharged

- Another shot of Inj. OROFER FCM 500mg IV after LSCS and Hb% boosted to 10 gm%.
- Baby and mother both are fine now.

Discussion

The current national guidelines advocate following stepts for comprehensive management of Thalessemia cases [5].

Preconception care

Women identified as Thalssmics should be advised to use contraception despite the reduced fertility.At each visit, screening for end-organ damage and optimisation of complications prior to embarking on any pregnancy. Aggressive chelation in the preconception stage can reduce and optimise body iron burden and reduce end-organ damage. As Diabetes is common in women with thalassaemia, they should be referred to a diabetologist and be under their care thourhgout the pregnanacy. Women with established diabetes mellitus should ideally have serum fructosamine concentrations < 300 nmol/l for at least 3 months prior to conception. This is equivalent to an HbA1c of 43 mmol. All women should be assessed by a cardiologist with expertise in thalassaemia and/or iron overload prior to embarking on a pregnancy. An echocardiogram and an electrocardiogram (ECG) should be performed as well as T2* cardiac MRI. Thyroid function should be determined and woman should be euthyroid in prepregnancy. All women should undergo a bone density scan and Serum vitamin D concentrations should be optimised with supplements if necessary. Women should be assessed for liver iron concentration using a FerriScan® or liver T2*. Ideally the liver iron should be < 7 mg/g (dry weight). Since our case had no clue of the condition none ofthese were done

Antenatal care

Women with thalassaemia should be reviewed monthly until 28 weeks of gestation and fortnightly thereafter. The multidisciplinary team should provide routine as well as specialist antenatal care. Our case did undergo monthly ANC starting a bit late in 16 -17week, though it is recommended for an early scan at 7 - 9 weeks of gestation. As against the recommendation of an routine first trimester scan (11 - 14 weeks of gestation) and a detailed anomaly scan at 18-20+6 weeks of gestation, our case got had the later one only.

If there is worsening maternal anaemia or evidence of FGR, regular transfusions should be considered. FOGSI recommends IV infusion if the Hb% is less than 9.9 g/dl in the third trimester because of the high risk of severe anemia and the short time available for oral supplementation. Despite oral iron supplementation in the second trimester our case did not show improvement in Hb%, and therefore was given IV infusion around 28 - 30 weeks. Iron sucrose is the most commonly used IV infusion in India. There are two options 1) One-shot each before and after delivery of Inj. OROFER FCM 500 mg IV or 2) Injection Iron 200 mg/day 4 - 5 injections.The first optionwas opted in our case,that resulted in replenishment of iron stores with peak ferritin concentrations in a week'stime after infusion as expected.

Intrapartum care

Continuous intrapartum electronic fetal monitoring was done as per national guideliens. Active management of the third stage of labour is recommended to minimise blood loss. Though Thalassaemia minor in itself is not an indication for caesarean section, our Gyencologist decided for a elective cesarian.

Postpartum care

Women with thalassaemia should are at a high risk for venous thromboembolism, but our case did not have any postpartum haemorrhae or peurpeal sepsis. Breastfeeding is safe and was done after 4 hours.

In a recent study (August 220) out of 493, 108 (21.9%) individual haemoglobinopathies sufferers β-thalassemia trait was the commonest haemoglobinopathy (12.98%), followed by HbE trait (7.50%), and compound heterozygous HbS/β-Thalassemia trait (1.42%) in overall population. The HbF was significantly greater in HbS heterozygous (1.45 ± 1.41), whereas mean HbA2 was significantly greater in β-Thalassemia trait (5.17 ± 1.36) [3].

Intravenous iron sucrose therapy was effective in increasing Hb%, S.Ferritin and other hematological parameters in pregnant women with moderate anemia reported a study. The mean Hb raised from 7.82 ± 0.93 to 11.34 ± 0.82 ($p < 0.0005$) after 8 weeks of therapy. There was significant increase in serum ferritin levels from 12.1 ± 5.1 to 67.34 ± 20.5($p < 0.0005$). Other parameters like serum iron, TIBC, reticulocyte count and MCV also improved sig-

nificantly. No major anaphylactic reactions were observed during the study period [4].

Many studies in India erport that carriers of Beta Thalassemia minor are usually asymptomatic but sometimes may show a mild Anemia. When both parents are carriers there is a 25% risk at each pregnancy of having children with homozygous Thalassemia [1]. The total iron requirement during pregnancy is about 1000-1200mg. (for the growing fetus- 270 mg, Placenta - 90 mg, for expansion of RBC mass - 450 mg and blood loss during delivery - 150 mg).

Pregnancy in Thalassemia major is complicated with oxidative stress, cardiac, hepatic, endocrine and metabolic complications and demand to be managed by a multidisciplinary team including cardiologist, endocrinologist, obstetrician, andhaematologists specialized in the treatment of thalassemia.Howeer the pregnancies in Beta Thalassemia minor do not pose much problem, but regular pregnancy checkups are mandatory. With this condition, one faulty gene causes only mild anemia. Even if one partner does not have the Thalassemia trait, the child will still be at risk of getting one damaged gene [8]. All efforts are to be made to maintain Haemoglobin at 10mg/dl and above to facilitate normal fetal growth [7].

An exploratory study conducted in a PHC attached to the Department of Community Medicine of a Govt. Medical College Bangalore, India, in the year 2013 reported that out of the 210 pregnant women who were tested, 18 (8.5%) were thalassaemia carriers. 12 (66.6%) of them were between 20 - 25 years of age. 5 (27.7%) were born out of 2nd degree consanguineous marriages. 7 (38.8%) had a history of abortions, among which 6 (33.3%) were in the 1st trimesters of their pregnancies. Out of the 18 positive women, 9 (50%) had turned up with their husbands. All of the husbands were negative for the Thalassaemia carrier status. Thus, there was no pregnancy which was at a risk of delivering babies with thalassaemia major. None (100%) of the pregnant women were aware of the disease, thalassaemia [9].

Another study at Disha Fertility and Surgical center, Indore from June 2007 to May 2009 of 1,006 women who underwent screening 28 were found to be having β thalassemia trait and seven had other hemoglobin variants. Population comprised 2.78% of women with β thalassemia trait and 0.69% with other minor hemoglobin variants. Only one couple (both husband and wife) was found to have β thalassemia trait. The β thalassemia minor group showed prominent microcytosis (MCV 66.9 ± 2.6 fl) but only mild anemia (Hb 11.41 ± 12.9 g/l). Out of 1,006 pregnant women during the period of 1 year, 57.6% women's hemoglobin was > 11 gm%. 36.88% women had mild anemia (Hb 10 - 11 gm%), 4.17% moderate anemia (Hb 7-10 gm%), 1.79% severe anemia (Hb < 7 gm%). With selective screening out of 206 women with moderate and severe anemia, 7.76% women found to have had abnormal hemoglobin pattern. In universal screening of women with mild anemia and no anemia only 1.5% women were having hemoglobinopathy [10].

Conclusion

Beta Thalassemia is Anemia, caused because red blood cells that are abnormally small (microcytic), that are not produced in adeqaute number, and do not contain enough functional Hemoglobin.

Any Thalessemia woman with pregnancy has a risk of sufferring severe anemia and needs a continuous monitoring and mainteance of Hb% over 10 g/dl throughout antenatal, intra-natal and post natal epriod.

If the Hb% goes below 9.9 G/dl IV infusion of IRON is recommended in third trimester and after delivery.

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