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COMPLICATIONS AND MANAGEMENT OF SICKLE BETA THALASSEMIA IN PREGNANCY: A CASE REPORT

Gynecology

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ABSTRACT

Background: Complications of Sickle beta thalassemia in pregnancy are underreported. Due to paucity of well controlled studies there are no clear guidelines for management especially with respect to blood transfusion. Here we describe a successful management of sickle beta thalassemia and discuss competing opinions and relevant recommendations.

Case summary: A 28 years old primigravida with previously undiagnosed sickle beta thalassemia had severe anemia and suffered from a crisis episode. She was given blood transfusions after worsening of anaemia and during painful crisis. Her pregnancy was completed successfully till term.

Conclusion: Underutilisation of antenatal services is a problem in India due to which such cases remain undiagnosed. In absence of clear guidelines, obstetricians are faced with the dilemma of selective vs. prophylactic transfusion.

KEYWORDS

Sickle beta thalassemia, sickle cell anemia, blood transfusion, pregnancy complications.

INTRODUCTION

Sickle beta thalassemia, a type of sickle cell disease, is an autosomal recessive inherited condition. The combination of the sickle cell mutation and beta-thalassemia (β-Thal) mutation gives rise to a compound heterozygous condition known as Hb S/β thalassemia (Hb S/β-Thal), which was first described in 1944 by Silvestroni and Bianco [1]. Depending on the type of mutation, patients may have completely abnormal haemoglobin which is defined as "sickle beta zero thalassemia" (Hb S/β0-Thal) or a reduced amount of normal hemoglobin called "sickle beta plus thalassemia" (Hb S/β+-Thal) [2,3]. Different amounts of Hb A arise due to quantitatively different β-globin synthesis caused by the heterogeneity of the β-Thal mutations. As a result, variable clinical manifestations present that range from nearly asymptomatic to a severe condition similar to homozygous Hb S state [1,4]. It is now known that sickle cell gene is fairly common in India and with frequency of sickle cell trait in some communities reaching as high as 35 percent [5].

Higher frequencies of pregnancy complications have been reported among women with sickle cell disease (SCD) compared with those without SCD. Further, pregnancy with sickle beta thalassemia is problematic due to lack of clear consensus on management as a result of few cases reported in literature. Although there is paucity of data, it has been suggested that patients suffering from sickle beta thalassemia can continue pregnancy till term with proper vigilance and care [6].

CASE REPORT

A 28 years old primigravida was diagnosed with sickle beta plus thalassemia in her second trimester when she started her antenatal visits. Her husband had no hemoglobinopathies. Her Hemoglobin (Hb) at the time of first antenatal visit was 8 gm/dl. However at 30 weeks of gestation her Hb fell to 6.5 gm/dl, for which she was given 2 units packed red blood cell (PRBC) transfusion. Patient was admitted in labour room at 39.1 weeks of gestation with pain abdomen. At the time of admission she was pale, icteric with vitals in normal range, per abdominally uterus was 36 weeks size, relaxed and she was not in labour. Admission NST was reactive. On admission, investigations revealed moderate anaemia of haemolytic type, hyperbilirubinemia and deranged coagulation profile (Hb-7gm/dl, direct bilirubin-1.67mg/dl, total bilirubin-3.94mg/dl, LDH-695,corrected reticulocyte count-7.4%, PT-15.2seconds). Her whole abdomen ultrasound showed hepatosplenomegaly. Her Serum ferritin in second trimester was 502ug/l and at the time of admission it was 1220ug/l. She complained of extreme pain in both lower limbs the next day. She was shifted to intensive care unit and managed with analgesics, hydration and oxygen. She was given 2 units PRBC the same day. Post transfusion Hb was 8gm/dl. The next day she went into labour and had fetal distress with meconium stained liquor for which emergency caesarean section was performed and a live female baby of 3.1kg was delivered. On her third postoperative day her Hb dropped to 7gm/dl, for which she was given 1 unit PRBC again. She remained in intensive care unit and recovered well.

DISCUSSION

Our patient, living in a first tier city like Delhi, had no counselling prior to conception and reported to antenatal clinic only in her second trimester. As she had no previous history of crisis episodes, blood transfusions, abortions or any other medical or surgical complication, her hemoglobinopathy had remained undiagnosed. Antenatal services have improved in India due to government policies and National Health Mission [7] but the awareness and utilisation of antenatal services even in urban setting remains unsatisfactory [8]. In such cases where women don't visit health facilities, pregnancy provides an opportunity to avail health services wherein Obstetrician is generally the first point of contact. This signifies the role of Obstetrician in diagnosing such cases that can go a long way in contributing to the general health.

In cases of sickle cell anaemia, pre-conceptional and antenatal visits are of utmost importance. Pre-conceptional visit enable clinician to stop current medication like hydroxyurea three months prior to conception [9]. These visits give clinician opportunity to take prior history of sickle cell crisis, abortion and consider the patient for low dose aspirin, blood transfusions considering the risk of alloimmunisation in current pregnancy or later conceptions, check for carrier status in the partner and offer genetic screening accordingly, screen for end organ damage and assess serum ferritin levels to determine iron overload [22].

Patient needs to be counselled about obstetrics complications like pregnancy induced hypertension, intrauterine growth retardation, preterm labour, venous thromboembolism, bone pain, end organ damage and transfusion related complications. Patient should also be educated about signs and symptoms of sickle beta thalassemia like anemia, repeated infections, frequent episodes of pain, acute chest syndrome, stroke, enlarged spleen and/or liver, jaundice, urinary tract infection and recurring perinatal mortality.

There are differences in literature on giving prophylactic transfusion in SCD. In a Randomized Cooperative study, Koshy et al 1988 [10] found that there was significant decrease in Sickle Cell Disease related complications including incidence of painful crises, in the group receiving prophylactic transfusions. However, there were no differences in fetal outcome or obstetric complications irrespective of the pregnant women receiving prophylactic transfusions. Authors concluded that there was no justification for routine use of prophylactic

transfusions in pregnancies for patients with Sickle Cell Disease. El-Shafei, et al 1995 [13], Tuck, et al 1987 [14], Ngo, et al 2010 [15] also did not found evidence of benefits to recommend prophylactic transfusions. A Cochrane review also concluded that other than a marginal reduction in the rate of painful crises, prophylactic transfusion conferred no clear advantage over selective transfusion with regards to pregnancy outcome [11].

However, other studies by Cunningham, et al 1983 [16], Morrison, et al 1991 [17], Howard, et al 1995 [18], Gilli, et al 2007 [19], Asma, et al 2015 [20], Benites, et al 2016 [21] have given evidence regarding benefits of transfusion.

Despite limited and conflicting data, there is a consensus that transfusion should be given in cases of pregnant women with a history of severe SCD-related complications, such as recurrent ACS and stroke, individuals on chronic transfusion prior to pregnancy, and those with repeated sickle cell crises or exacerbation of anaemia during pregnancy. Prophylactic transfusion should also be considered for women with other serious medical or obstetric conditions or with a multiple pregnancy [9].

In our case, patient received blood transfusion for worsening of anemia at 30 weeks of gestation. After admission, she had painful crisis for which she was managed with hydration, analgesics and oxygen along with blood transfusion. In post-operative period due to fall in Hb she further received blood transfusion and she recovered well.

CONCLUSION

It is to be noted that blood transfusion has potential complications including risk of alloimmunisation [12]. Hence, in the light of trade-off, the situation poses a challenge for the treating obstetrician to decide on the number of blood transfusions to be given. Updated guidelines by British Society for Haematology [9] can help in taking a better decision. Timely pre-conception counselling and ante natal care are important but often ignored due to lack of awareness. However, timely intervention sickle beta thalassemia can lead to lesser complications and better maternal and perinatal outcomes.

REFERENCES

[1] Dacie J. (1988). The hereditary haemolytic anaemias, 3rd ed. Churchill Livingstone; New York

[2] Stuart M.J., Nagel R.L. (2004) Sickle-cell disease. The Lancet, 364(9442):1343-60.

[3]. Thein, S. L. (2011). Genetic modifiers of sickle cell disease. Hemoglobin, 35(5-6), 589-606.

[4]. Kinney T.R., Ware R.E. Compound heterozygous states. In: Embury S.H., Hebbel R.P., Mohandas N., Steinberg M.H., editors (1994). Sickle cell disease: basic principles and clinical practice. 1st ed. Raven Press Ltd.; New York:437–451.

[5]. Colah, R. B., Mukherjee, M. B., Martin, S., & Ghosh, K. (2015). Sickle cell disease in tribal populations in India. The Indian journal of medical research, 141(5), 509.

[6]. Koshy, M. (1995). Sickle cell disease and pregnancy. Blood reviews, 9(3), 157-164.

[7]. National Health Mission - Governnment of India. (n.d.). Retrieved December 5, 2018, from http://nhm.gov.in/

[8]. Patel, B., Gurmeet, P., Sinalkar, D., Pandya, K., Mahen, A., & Singh, N. (2016). A study on knowledge and practices of antenatal care among pregnant women attending antenatal clinic at a Tertiary Care Hospital of Pune, Maharashtra. Medical Journal of Dr. DY Patil University, 9(3), 354-354.

[9]. Davis, B. A., Allard, S., Qureshi, A., Porter, J. B., Pancham, S., Win, N., ... & British Society for Haematology. (2017). Guidelines on red cell transfusion in sickle cell disease Part II: indications for transfusion. British journal of haematology, 176(2), 192-209.

[10]. Koshy, M., Burd, L., Wallace, D., Moawad, A., & Baron, J. (1989). Prophylactic red-cell transfusions in pregnant patients with sickle cell disease. A randomized cooperative study. International Journal of Gynecology & Obstetrics, 29(3), 277-278.

[11]. Okusanya, B. O., & Oladapo, O. T. (2016). Prophylactic versus selective blood transfusion for sickle cell disease in pregnancy. The Cochrane Library.

[12]. Vichinsky, E. P., Earles, A., Johnson, R. A., Hoag, M. S., Williams, A., & Lubin, B. (1990). Alloimmunization in sickle cell anemia and transfusion of racially unmatched blood. New England journal of medicine, 322(23), 1617-1621.

[13] El-Shafei, A. M., Dhaliwal, J. K., Sandhu, A. K., & Al-Sharqi, M. R. (1995). Indications for blood transfusion in pregnancy with sickle cell disease. Australian and New Zealand journal of obstetrics and gynaecology, 35(4), 405-408.

[14] TUCK, S. M., Brewster, E. M., Studd, J. W. W., JAMES, C. E., & Pearson, T. C. (1987). Prophylactic blood transfusion in maternal sickle cell syndromes. BJOG: An International Journal of Obstetrics & Gynaecology, 94(2), 121-125.

[15] Ngo, C., Kayem, G., Habibi, A., Benachi, A., Goffinet, F., Galacteros, F., & Haddad, B. (2010). Pregnancy in sickle cell disease: maternal and fetal outcomes in a population receiving prophylactic partial exchange transfusions. European Journal of Obstetrics & Gynecology and Reproductive Biology, 152(2), 138-142.

[16] Cunningham, F. G., Pritchard, J. A., & Mason, R. (1983). Pregnancy and sickle cell hemoglobinopathies: results with and without prophylactic transfusions. Obstetrics and gynecology, 62(4), 419-424.

[17] Morrison, J. C., Morrison, F. S., Floyd, R. C., Roberts, W. E., Hess, L. W., & Wiser, W. L. (1991). Use of continuous flow erythrocytapheresis in pregnant patients with sickle cell disease. Journal of clinical apheresis, 6(4), 224-229.

[18] Howard, R. J., Tuck, S. M., & Pearson, T. C. (1995). Pregnancy in sickle cell disease in the UK: results of a multicentre survey of the effect of prophylactic blood transfusion on maternal and fetal outcome. BJOG: An International Journal of Obstetrics & Gynaecology, 102(12), 947-951.

[19] Gilli, S. C., De Paula, E. V., Biscaro, F. P., Marques, J. F., Costa, F. F., & Saad, S. T. (2007). Third-trimester erythrocytapheresis in pregnant patients with sickle cell disease. International Journal of Gynecology & Obstetrics, 96(1), 8-11.

[20] Asma, S., Kozanoglu, I., Tarım, E., Sarıturk, C., Gereklioglu, C., Akdeniz, A. ... & Boga, C. (2015). Prophylactic red blood cell exchange may be beneficial in the management of sickle cell disease in pregnancy. Transfusion, 55(1), 36-44.

[21] Benites, B. D., Benevides, T. C. L., Valente, I. S., Marques Jr, J. F., Gilli, S. C. O., & Saad, S. T. O. (2016). The effects of exchange transfusion for prevention of complications during pregnancy of sickle hemoglobin C disease patients. Transfusion, 56(1), 119-124.

[22] No 61, G. T. G. (2011). Management of sickle cell disease in pregnancy. London: Royal College of Obstetricians and Gynaecologists.