PREGNANCY IN THALASSAEMIA

(Two Case Reports)

by

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Thalassaemia with pregnancy is not very common. Previously most patients did not live upto the child bearing age and few of those who happened to have pregnancy were treated as cases of simple anaemia. Earlier the disease was thought to be prevalent only in Mediterranean region but lately it has been observed that the disease is not so rare in the south-eastern hemisphere.

Lahiri and Konar (1977) reported 6 cases of pregnancies in thalassaemia patients (2 intermediate type, 1 trait and 3 having HbE thalassaemia) from Eden Hospital, Calcutta amongst 50,000 obstetric admissions covering 5 years. Here 2 more cases of Thalassaemia E disease with pregnancy are presented amongst 8209 obstetric admissions covering mid. 1977 to mid. 1978.

Case 1 Mrs. S. R. 26 years, $P_2 + O$, was admitted in Eden Hospital with severe anaemia, while carrying 36th week of pregnancy.

Menstrual history: Menarche at 13; cycles 4-5 days L.M.P. 12-6-77, E.D.D., 19-3-78.

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Obstetric history: She had 2 previous pregnancies; during first one, from 12th week, she suffered from severe anaemia along with palpitations and dyspnoea, which compelled her to be hospitalised in N.R.S.M.C., when blood transfusion was attempted but abandoned due to precipitation of severe reactions. She was discharged after 8 weeks. At 32 weeks, she delivered a still born baby, 2 kg in weight at home. During her second pregnancy, she was again hospitalised at R.G.K.M.C. at 20th week, and stayed there upto 36th weeks when she delivered a stillborn female baby again, weighing 2 kg. She received 1 bottle of blood; other attempts for transfusions precipitated severe reactions.

Past History: She was aware of enlarged spleen since her childhood and suffered from occasional bouts of high fever, but Thalassaemia was diagnosed in hospital during her first pregnancy. She availed no further treatment for this during all these years excepting when she was hospitalised during her second conception. She had no antenatal check up during this current pregnancy also.

Family History: Nothing suggestive except that one of her younger brothers was also suffering from splenomegaly since childhood, but no investigations were carried out.

Examination: Severe anaemia, mongoloid facies, engorged neck veins and axillary lymphadenopathy were detected. Jaundice or oedema of legs were not present B.P. 120 mm of Hg.

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pulse 90 p.m. reg. Liver palpable, 5 cm. below costal margin, firm, non-tender, Spleen, palpable 15 cm. below left costal margin, firm, non-tender. Soft systolic murmur was present in mitral area and there was also congestion in both lung bases. Uterus corresponded to 32 weeks pregnancy, vertex presentation. Foetal heart sounds, regular.

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Investigations

12-2-78: Hb, 5 gm%, hypochromia Anisocytosis Poikilocytosis, Target Cells, Normoblasts, 8/100. W.B.C., Normocytes, 16/100 W.B.C., T.C.—8500, poly, 58, lympho 26, mono, 10, eosino 6%. Platelets, adequate, Total serum bilirubin—0.7 mg %.

24-2-78 Hb–4.4%, hypochromia, +, Anisocytosis, Poikilocytosis, Macro, micro, target cells, Polychromasia, basophilic stippling +, Reticulocytes 12%, Plasma cells-occasional, W.T.C.—10,000; poly 68, lympho 15, eosino 16, mono-1%, normocytes 25/100 WBC, Normoblasts, 10/100 WBC, Nucleated myelocytes 1%, Nucleated metamyelocyte 3%, Platelets adequate, E.S.R. 52 mm. (1st hour, wintrode method).

Electrophoresis done: HbF-67.6%, Electrophoresis pattern-E.F.

Diagnosed thalassaemia syndrome complicated with infection in pregnancy.

Management

The patient was treated by Folic acid, fresh blood transfusion twice a week, one unit at a time (received altogether 3 such). Haemogolobin on 10-3-78 showed improvement (6.8 gm%).

The patient went into labour on 16-3-78 at full term and after 6 hours of labour pain assisted breech delivery was undertaken when a living premature baby weighing 2 kg was born. The baby died on the following day due to pulmonary haemorrhage. The patient went home on 23-3-78 with advice.

Case 2

Mrs, S. F., 21 years, Para 0, 0 was admitted on 16-2-78 with 28th weeks of pregnancy, severe anemia, and exertional dyspnoea. She was married for 1½ years.

Menstrual History: Menarche, 15 years, Cycles 4-5 days L.M.P. 17-8-77 E.D.D. 24-5-78.
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Past History: Anaemia was detected when she was 5 years old and she was treated in haematology Dept, School of tropical Medicine for weakness, lassitude and lethargy. She was diagnosed there as a case of Thalassaemia. At 13 years, she was again admitted to Railway Hospital, Sealdah, Calcutta with similar complaints and also for pain and aches all over the body, where blood transfusions were given and splenectomy was advised, which the patient refused. She was further advised not to marry, which she did not comply with. She was also having splenomegaly since her childhood. She had past histories of epistaxis, haematemesis, recurrent janudice, leg ulcers and skin infections. Dysponea developed since last 3 months.

Family History. Nothing suggestive.

Examination on admission showed poor nutrition, poor build with mongoloid facies, severe anaemia and engorged neckveins but no jaundice or oedema legs. B.P. — 116/60 mm of Hg, pulse—112 P.M. with pulse deficit. Liver, enlarged 5 cm. below right costal margin, firm, non-tender; spleen 15 cm. below costal margin, firm, not tender. Sternal tenderness was present and there was a soft systolic murmur over both pulmonary and mitral area of heart. Height of the fundus of uterus corresponded to 24 weeks of pregnancy. Foetal heart sounds, regular.

Investigations

Previously on 27-5-71 she was diagnosed as HbE thalassaemia by haematological investigations in S.T.M. After admission now, haematological investigations were carried out on 17.2.78, 24.2.78, 11.3.78, 19.3.78, 28.3.78, 4.4.78, 18.4.78 & 12.5.78 in collaboration with Haematology Dept., Calcutta Medical College and School of tropical medicine, Calcutta.

17.2.78, Hb-4.5 gm%, W.B.C. 6400, poly-78, lympho-18, eosino-3, and mono–1%.

24.2.78 at S.T.M., Electrophoresis done, HbF, 19.4%, Electrophoresis pattern—EF., Hb-5 gm% 11.3.78 at S.T.M.,

Gross hypochromic microcytic anaemia, Gross aniso-poikilocytosis, target cells and oval cells +, polychromasia — normocytes 5/100 W.B.C., Normoblasts plenty, Total bilirubin-1 mg.%.

Chest X-Ray—no abnormality, X-Ray skull and metacarpophalangeal joints (taken after confinement), no abnormality.

She received folic acid 1 tab daily, packed cells transfusion and blood, 8 bottles.

Subsequently her haemoglobin level was increased to 6 gm% on 18-4-78. Her peripheral blood was examined on 12-5-78 which confirmed the earlier findings. Platelets were adequate. On 5-6-78, foetal maturity was assessed by X-ray which showed single foetus, presentation, vertex, maturity more than 36 weeks. Labour started on 8-6-78 but did not progress satisfactorily within next 12

hours L.U.C.S. under epidural anaesthesia was done after medical induction failed. A living female baby weighing 2.4 Kg. was born. She received another 5 bottles of blood during the post operative period and was discharged on the 16th day along with healthy baby. Postnatal check up showed that the haemoglobin level had again came down to 5.5 gm%, otherwise she was well.

Discussion

Two cases of pregnancy in Thalassaemia are presented. Though in both of them living baby was born, yet in the first case, the baby died on the following day.

Thalassaemia represents a group of hereditary disorder of haemoglobin synthesis, believed to be transmitted as Mendelian dominant through either or both parent.

Depending on the globin-peptide chain, the disease is divided into α and ß, the latter is the classical variety. When the abnormal gene is homozygos, it may give rise to Thalassaemia major and intermedia. If it heterogygos, thalassaemia minor or trait may occur. There may also be heterozygos state of Hbc thalassaemia, HbD thalassaemia and HbE thalassaemia. Thalassaemia either in homozygos form or in combination with HbE appears to be the commonest form of haemoglobonopathies in Bengal and their incidences are approximately 3.7 or 3.9% in Bengalees (Chatterjee, 1968).

Thalassaemia major is almost always fatal before puberty is reached and hypogonadism appears in them between 8 to 10 years (Willoughly, 1973). Hence chances of conceptions are very unlikely in them. The intermedia, minor and HbE thalassaemia are the usual types where pregnancy at times may occur. The 2 cases of the present series also belonged to HbE thalassaemia.

The management of pregnancy and labour in these patients requires constant supervision. Haemoglobin level is to be maintained as high as possible, better as close as to 8 gm% (Lahiri and Konar, 1977). Labour is to be conducted taking all precautions necessary for a severely anaemic expectant mother. Since haemolytic crisis may occur, large quantity of blood may be transfused during labour. Though thalassaemia per sé is not an indication for L.U.C.S. yet these severely anaemic patients should not be made to undergo prolonged and difficult labour. In the 2nd case also L.U.C.S. was undertaken not for thalassaemia but for prolonged labour with uterine inertia and the result was satisfactory.

These cases are very prone to infections. The patient should be hospitalised if successful pregnancy is to be expected. Repeated blood transfusions are the sheet anchor of treatment.

It is needless to mention that pregnancy should be avoided in thalassaemic patients, whenever possible or practicable and repeated pregnancies may prove fatal.

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