SICKLE CELL ANAEMIA IN PREGNANCY: A CASE REPORT

S.P. RACHAGAN
S. RAMAN
R. CHERIAN

SUMMARY

To our knowledge, sickle cell anaemia in pregnancy has not been reported in Malaysia. We describe here such a case with emphasis on modern methods of management.

INTRODUCTION

Sickle cell anaemia (i.e., HbSS) represents the homozygous state of the HbS gene inherited from each parent who has the sickle cell trait. In this disorder there is substitution of valine for glutamic acid in the $\nu$ chain of the haemoglobin molecule. It is inherited as a Mendelian dominant and it occurs mainly among the Negros, although the trait has been reported among the tribal population of South India. Kobak first described how this condition affects pregnancy and since then there have been extensive reviews on this subject. We describe below a case of sickle cell anaemia in pregnancy in an Indian patient and to our knowledge, this is the first case of this condition in pregnancy described locally.

CASE REPORT

The patient was a 29-year-old Indian para 0 with one previous abortion. She was first seen at the University Hospital at 31 weeks of gestation with a history of bone pains for three months and jaundice one month prior to admission.

She was a known case of sickle cell anaemia diagnosed about five years ago at another hospital. For the past four years, she had been having recurrent attacks of jaundice with tea-coloured urine. She was also treated for anaemia. There was no history of any associated infections or legs ulcers. She gave no history suggestive of sickling crisis.

In 1979, she underwent a cholecystectomy for gallstones. There were no complications during or after surgery.

Subsequently in the same year she had a spontaneous abortion at 12 weeks gestation. Since it was a complete abortion, no curettage was done.

There was no relevant family history of similar problems. She was admitted to the antenatal ward of the University Hospital where she stayed until
Table I
ANTENATAL COURSES AND MANAGEMENT

<table>
<thead>
<tr>
<th>Gestation in weeks</th>
<th>Fundal Height</th>
<th>BP</th>
<th>Treatment</th>
<th>Sickle cell %</th>
<th>Hb (g/dl)</th>
</tr>
</thead>
<tbody>
<tr>
<td>31/52</td>
<td>32/52</td>
<td>110/70</td>
<td>Transfusion – Packed Cells 250ml</td>
<td>86</td>
<td>7.0</td>
</tr>
<tr>
<td>32/52</td>
<td>32/52</td>
<td>110/70</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>34/52</td>
<td>34/52</td>
<td>120/80</td>
<td>Exchange transfusion – packed cells 500ml</td>
<td>69</td>
<td>8.2</td>
</tr>
<tr>
<td>35/52</td>
<td>110/70</td>
<td></td>
<td>Exchange transfusion – packed cells 500ml</td>
<td>43.5</td>
<td>9.2</td>
</tr>
<tr>
<td>36/52</td>
<td>36/52</td>
<td>120/80</td>
<td>Exchange transfusion – packed cells 500ml</td>
<td>43.0</td>
<td>10.6</td>
</tr>
<tr>
<td>37/52</td>
<td>110/70</td>
<td></td>
<td></td>
<td>19.5</td>
<td>12.3</td>
</tr>
<tr>
<td>38/52</td>
<td>38/52</td>
<td>120/80</td>
<td></td>
<td>11.7</td>
<td>11.5</td>
</tr>
<tr>
<td>39/52</td>
<td>120/80</td>
<td></td>
<td></td>
<td>9.5</td>
<td>10.1</td>
</tr>
<tr>
<td>40/52</td>
<td>130/90</td>
<td></td>
<td>Spontaneous labour</td>
<td></td>
<td></td>
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<tr>
<td></td>
<td></td>
<td></td>
<td>Foetal distress</td>
<td></td>
<td></td>
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<tr>
<td></td>
<td></td>
<td></td>
<td>Lower segment caesarean section</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

delivery. Examination on admission revealed that there was moderate pallor, mild jaundice and minimal ankle oedema. The blood pressure was 110/70mm Hg and the pulse was 80/minute and regular. There were no abnormalities detected in the cardiovascular and respiratory systems.

Abdominal examination revealed a healed surgical scar in the right hypochondrium. There was no hepatosplenomegaly. The fundal height corresponded to dates with a single foetus in longitudinal lie and cephalic presentation. The foetal heart was clearly heard. Her antenatal course and management is summarised in Table I.

Relevant investigations revealed the following: Haemoglobin 7.0 g/decilitre; Reticulocyte count 44.5%; Total bilirubin 27 µmol/l (Normal 3.4 – 24); Serum iron 95% transferrin saturation; Haemoglobin electrophoresis – HbS 88.2%, HbA absent, HbF 10.5%, HbA₂ 1.3%.

The serum iron and haemoglobin electrophoresis of the husband were normal.

Exchange transfusions were done at regular intervals. At 32 weeks the patient was transfused with 250mls of packed cells. This was followed by three partial exchange transfusions at 34, 35 and 36 weeks gestation. Each time, 500mls of whole blood was removed from the patient and 500mls of packed cells were transfused immediately after. The blood used was as fresh as was available. After the third exchange transfusion, the patient’s haemoglobin was 12.3 g % and the level of HbS was 19.5%. The level of HbS was monitored weekly thereafter and it fell progressively to 11.7% at 38 weeks and 9.5% at 39 weeks. This is a well known occurrence due to the correction of the anaemia and thereby removal of the stimulus to the patient’s bone marrow to continue producing HbS at the original rate.

The foetus was closely monitored clinically, biochemically and bioelectrically using the cardiotocograph throughout her stay in the ward. These investigations revealed no abnormalities. The maternal condition was also monitored carefully, particularly her haemoglobin and sickle cell percentage. Her blood pressure was also monitored twice daily and her urine checked at regular intervals for any evidence of urinary tract infection. All these were normal throughout her stay in the ward. She was put on thrice daily doses of folic acid during her antenatal stay. No oral iron was given.

At 40 weeks gestation she went into spontaneous labour. On vaginal examination the cervix was 3 cm dilated and the station was –1. Forewater
amniotomy revealed thick meconium-stained liquor. Cardiotocographic monitoring showed late deceleration pattern suggestive of foetal distress.

Emergency lower segment caesarean section was done under general anaesthesia. A live male baby weighing 2,620 g with an Apgar score of 9/10 was delivered. Blood loss at operation was estimated to be 600mls. The baby was sent to the special care nursery for subsequent management.

Her post-operative recovery was uneventful. Her haemoglobin was 8.6g/dl and she was transfused with packed cells and discharged home well with her baby. At post-natal follow-up six weeks later, both mother and baby were well.

DISCUSSION

Sickle cell anaemia is common among the American Negroes in whom the incidence varies from 0.1 to 1.3%. Other races like the Greeks, Turks and Indians rarely inherit this disorder. This patient is prone to all the maternal complications like sickling crisis (both haemolytic and thrombotic) as well as infections, particularly of the urinary tract. Our patient did not give any history suggestive of thrombotic problems but she had repeated attacks of jaundice and gallstones which suggests repeated haemolytic episodes in the past. There is a higher maternal and foetal morbidity and mortality in this condition. The patient’s foetus was thus monitored closely for any evidence of growth retardation and foetal distress.

The mother’s sickle cell percentage level and haemoglobin levels were carefully monitored. To reduce haemolytic and thrombotic episodes, the modern management is to do exchange transfusions to keep the sickle cell percentage level below 20% and the haemoglobin level between 10-11g/dl. This was done in this patient.

Although this patient did not develop atypical erythrocytic antibodies due to repeated blood transfusions, it has been known to occur in Negro patients after repeated transfusions from Caucasian donors. This can lead to serious problems.

Since folic acid deficiency is common in these patients, our patient was put on high doses of folic acid. Preterm labour is another common complication but our patient went to term. Fetal distress in labour is also more likely as in this case.

The mean birth weight is usually lower for the gestational age in this condition but in this patient the baby’s weight was above the mean birthweight for the Indian female of her social class.

In cases where both husband and wife are traits, foetoscopy with foetal blood sampling for antenatal diagnosis should be advised after genetic counselling. We do not have such facilities yet in this country.

General anaesthesia is an added hazard for these patients. Therefore, care was taken during caesarean section to avoid hypoxia, hypoventilation, hypotension, hypothermia, dehydration and acidosis, as all these could precipitate sickling crisis.

REFERENCES