CONGENITAL THROMBOCYTOPENIC PURPURA

BY

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Congenital thrombocytopenic purpura is a rare condition. A review of the literature fails to show any case reported from this country. In 1925 Rushmore was able to find seven examples of purpura in pregnancy in which both mother and child were affected, but the type of purpura was not specified. Since that time congenital thrombocytopenic purpura has been recorded on fourteen occasions with eight thrombocytopenic mothers (see table 1). The following instance in a newborn infant whose mother did not suffer from the disease may therefore be of interest.

Clinical details

A male infant, three days old, was admitted to the Hospital for Sick Children, Great Ormond Street, on April 21, 1944, with a purpuric eruption. He was born at term following an uneventful pregnancy and labour. On the second day of life purpuric spots were first noticed on the buttocks and extended rapidly to the legs and scalp. Jaundice also appeared at this time. His parents and one older sister were healthy, but the second sister suffered from recurrent epistaxis.

Examination showed a normally responsive, well-nourished baby, weighing 5 lb. 14 oz. T. 100-2° F. (rectal), P. 140, R. 40. A diffuse purpuric rash was present over all areas of the skin and also on the hard palate with small ecchymoses on the back and legs; mild jaundice was present; mucous membranes were a good colour; the spleen was not palpable; and there was no abnormality of heart, lungs, abdomen or central nervous system. No evidence of infection was found. The urine and stools were free from blood. Pathological investigations carried out on the fourth day were as follows:—

INFANT:

Blood count: R.B.C. 6,700,000 per c.m.m.; Hb. 130 per cent. (Sahli); C.I. 0-97.

W.B.C. 13,300 per c.m.m.

Polymorphs 58 per cent. with 38 per cent. stab forms.

Lymphocytes 37 per cent.

Monocytes 1 per cent.

Eosinophils 4 per cent.

No immature cells seen.

Platelet count 6000 per c.m.m.

Bleeding time 10 minutes (Duke’s method—normal 2–5 minutes).

Prothrombin time 26 seconds (McTookey’s method—normal 11–15 seconds).

Blood group O Rh-positive.

MOTHER:

Platelet count 350,000 per c.m.m.

Blood Wassermann reaction negative.

Blood group O Rh-positive.

No abnormal antibodies were present in the serum.

The infant remained in hospital for four weeks, during which time he made good progress, gaining 20 oz. in weight. The purpura faded gradually; a few new lesions appeared during the first fortnight; the stools contained streaks of bright blood from time to time and there was one small haematemesis. He was symptom free by the end of the third week. Apart from a temperature of 100° F. on admission he was afebrile, his rectal temperature varying between 97·6° F. and 99° F. for the first eight days and then becoming stabilized at 98° F. No infection was detected at any time. The jaundice continued for eighteen days which is considerably longer than usual in physiological jaundice; the fall in the red blood count was also marked (see table 2). In spite of clinical improvement the thrombocytopenia persisted with a platelet count of only 10,000 per c.m.m. on discharge from hospital at one month old (see table 2). The normal figures for the neonatal period are 200,000–300,000 per c.m.m. (Sanford, 1942).

The patient was not seen again until he was six months old because of the danger from flying-bombs in the London area. During the interval he had remained symptom free although he had had a severe attack of bronchitis. The platelets had risen to 350,000 per c.m.m. and the bleeding time had fallen to 5½ minutes.

This was a spontaneous recovery. The only treatment the child received was vitamin K (5 mgm. Kapilon intramuscularly) on admission and liver (Armour’s Proethron forte 2 c.c.m.) given on the twenty-first and twenty-third days, the latter being without effect on the thrombocytes. Owing to the benign clinical course blood transfusion was not considered necessary.

Comment

This appears to be an undoubted example of congenital thrombocytopenic purpura. The rash was noticed by the second day and the platelets were reduced to 6,000 per c.m.m. on the fourth day. A symmetrical purpura was unlikely as no infection was found, no drug liable to cause a thrombocytopenia had been given to mother or child, and in spite of the prolonged jaundice no other blood disease was demonstrable.

In nine of the cases reviewed the condition was
## Table 1.—Cases of Congenital Thrombocytopenic Purpura

<table>
<thead>
<tr>
<th>Reference</th>
<th>Manifestations</th>
<th>Platelet count per c.mm.</th>
<th>Bleeding time (minutes)</th>
<th>Treatment</th>
<th>Course</th>
<th>Family</th>
</tr>
</thead>
<tbody>
<tr>
<td>WALTNER (1924)</td>
<td>Purpura of skin at birth</td>
<td>90,000</td>
<td>12</td>
<td>None</td>
<td>Marked icterus; some sepsis. Symptom free by seven months.</td>
<td>Mother: purpura at seventh month of pregnancy. Platelets 50,000. Bleeding time 27 minutes.</td>
</tr>
<tr>
<td>LESCHE (1926)</td>
<td>Purpura of skin at birth with haematuria and melaena</td>
<td>0</td>
<td>12–15</td>
<td>None</td>
<td>No bleeding after tenth day.</td>
<td>Mother: purpura at fifth month of pregnancy. Platelets 60,000.</td>
</tr>
<tr>
<td>LIEBLING (1926)</td>
<td>Purpura of skin at birth with haematemesis and melaena</td>
<td>40,000</td>
<td>—</td>
<td><a href="https://www.bmj.com/content/2/1/95.long">Hæmoplastin, 5 c.c.m. intramuscularly.</a></td>
<td>Did well. Purpura faded at one month. Platelets 240,000.</td>
<td>—</td>
</tr>
<tr>
<td>GREENWALD (1929)</td>
<td>Purpura of skin appeared on fifth day.</td>
<td>30,000</td>
<td>5</td>
<td>None</td>
<td>Died on seventh day. Congenital morbus cordis. Thrombocytes scanty and abnormal in appearance.</td>
<td>—</td>
</tr>
<tr>
<td>BAYER (1931)</td>
<td>Purpura of skin at birth</td>
<td>50,000</td>
<td>40</td>
<td>None</td>
<td>No fresh bleeding after birth. Platelets 320,000 by second year.</td>
<td>—</td>
</tr>
<tr>
<td>Ibid.</td>
<td>Purpura of skin at birth</td>
<td>48,000</td>
<td>6½</td>
<td>None</td>
<td>No fresh bleeding after birth. Platelets 150,000 on fifth day.</td>
<td>—</td>
</tr>
<tr>
<td>Ibid.</td>
<td>Purpura of skin and mucous membranes at birth with haematemesis and melaena</td>
<td>45,000</td>
<td>11</td>
<td>Coagulen and gelatin intramuscularly</td>
<td>No fresh bleeding after fifth day. Platelets 280,000 by second year.</td>
<td>—</td>
</tr>
<tr>
<td>Ibid.</td>
<td>Purpura of skin at birth</td>
<td>50,000</td>
<td>90</td>
<td>None</td>
<td>Died on second day. Widespread purpuric lesions.</td>
<td>—</td>
</tr>
<tr>
<td>GUTT-FREUND (1933)</td>
<td>Purpura of skin and mucous membranes at birth.</td>
<td>0</td>
<td>40</td>
<td>90 c.c.m. blood intramuscularly, also blood transfusion (? amount).</td>
<td>Continued to bleed. Died of pneumonia at four months.</td>
<td>—</td>
</tr>
<tr>
<td>SANFORD (1936)</td>
<td>Purpura of skin at birth</td>
<td>20,000</td>
<td>—</td>
<td>None</td>
<td>No fresh lesions after birth. Platelets 137,000 by tenth day. Mother and child both showed increased disintegrating power of platelets. Platelets 672,000 by ten weeks. Remained normal for two years.</td>
<td>—</td>
</tr>
<tr>
<td>DAVIDSON (1937)</td>
<td>Purpura of skin at birth</td>
<td>56,000</td>
<td>3</td>
<td>Blood, 100 c.c.m. intramuscularly without improvement. Bothrops antitoxin, 5 c.c.m. intramuscularly with clinical recovery.</td>
<td>Died third day. Ten-torial tear. Widespread purpura. Died second day with multiple haemorrhages. Purpura faded in a few days. Bleeding time 1½ minutes at 2½ years old.</td>
<td>—</td>
</tr>
<tr>
<td>WHITNEY (1942)</td>
<td>Purpura of skin at birth</td>
<td>80,000</td>
<td>1</td>
<td>Blood intramuscularly, 20 c.c.m.</td>
<td>Died third day. Ten-torial tear. Widespread purpura. Died second day with multiple haemorrhages. Purpura faded in a few days. Bleeding time 1½ minutes at 2½ years old.</td>
<td>—</td>
</tr>
<tr>
<td>Ibid.</td>
<td>Purpura of skin at birth</td>
<td>90,000</td>
<td>11</td>
<td>Blood intramuscularly, 55 c.c.m., twice. 50 c.c.m. blood intravenously.</td>
<td>Died third day. Ten-torial tear. Widespread purpura. Died second day with multiple haemorrhages. Purpura faded in a few days. Bleeding time 1½ minutes at 2½ years old.</td>
<td>—</td>
</tr>
<tr>
<td>URBANSKY (1942)</td>
<td>Purpura of skin and mucous membranes.</td>
<td>23,200</td>
<td>5½</td>
<td>None</td>
<td>Died third day. Ten-torial tear. Widespread purpura. Died second day with multiple haemorrhages. Purpura faded in a few days. Bleeding time 1½ minutes at 2½ years old.</td>
<td>Mother of these two infants had splenectomy at fourteen years of age. Platelets 70,000.</td>
</tr>
</tbody>
</table>

1. Cases of congenital thrombocytopenic purpura are characterized by purpura that appears at birth or within the first few days of life. These cases are associated with a marked decrease in platelet count, often below 20,000 per c.mm. The bleeding tendency is severe, with haematemesis and melaena at birth or within the first few days. The prognosis is variable, with some cases improving without treatment, while others require transfusion and other therapies. The table above summarizes the cases reported in the literature, including the platelet count, bleeding time, and treatment outcomes. The table also highlights the family history, which is often significant in cases of congenital thrombocytopenic purpura.
Table 2

<table>
<thead>
<tr>
<th>Date</th>
<th>Hb. per cent.</th>
<th>R.B.C. per c.mm.</th>
<th>W.B.C. per c.mm.</th>
<th>Platelets per c.mm.</th>
<th>Bleeding time</th>
</tr>
</thead>
<tbody>
<tr>
<td>22.4.1944</td>
<td>130</td>
<td>6,700,000</td>
<td>13,300</td>
<td>6,000</td>
<td>10 minutes +</td>
</tr>
<tr>
<td>1.5.1944</td>
<td>80</td>
<td>3,500,000</td>
<td>22,200</td>
<td>7,000</td>
<td>—</td>
</tr>
<tr>
<td>8.5.1944</td>
<td>80</td>
<td>4,100,000</td>
<td>16,000</td>
<td>15,000</td>
<td>10 minutes +</td>
</tr>
<tr>
<td>12.5.1944</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>26.5.1944</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>24.10.1944</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>350,000</td>
<td>5½ minutes</td>
</tr>
</tbody>
</table>

familial; in the four collected by Bayer the mothers’ platelet counts were not noted. In the present instance the mother’s blood was normal and she had never shown a tendency to bleed.

The prognosis in these infants is of some interest. Nine of the fourteen recovered during the first few weeks of life but their progress was not followed further except in three who remained healthy for two years. It would be instructive to know whether or not these children tend to relapse later in life. Five of the infants died, associated lesions may have been the cause of death in three of them. Two only died of uncomplicated purpura. Thus prognosis does not appear to be so grave as might be expected.

From a study of these reports therapy seems to have had little effect. Five infants recovered without treatment. Blood transfusions in adequate amounts would seem to be the rational treatment in any severe case. The advisability of splenectomy at birth was discussed by Whitney (1942) but in view of the above figures does not appear justifiable.

Summary

A case of congenital thrombocytopenic purpura is reported.

Examples of congenital thrombocytopenic purpura collected from the literature are tabulated.

The prognosis and treatment of the condition is considered.

Thanks are due to Dr. W. G. Wyllie for permission to publish this case.

References


Congenital thrombocytopenic purpura

Patience E. Braclay

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doi: 10.1136/adc.20.102.94

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