Spontaneous central nervous system bleeding in a child with cyanotic congenital heart disease

A 9-year-old Indian boy was admitted with complaints of bifrontal headache since 2 days and one episode of left-sided tonic-clonic convulsion lasting for 10 minutes followed by weakness of left upper and lower limb. There was no history of fever, drug intake, unconsciousness, trauma or bleeding manifestations. Patient was a known case of cyanotic congenital heart disease (CCHD) diagnosed at the age of 3 years.

Echocardiography showed double outlet right ventricle with ventricular septal defect (5 mm) and pulmonary stenosis. In view of cyanotic spells, he was started on propranolol. On admission, child was afebrile with a heart rate 104/min, respiratory rate 24/min and blood pressure 96/60 mmHg. Central cyanosis and grade 3 clubbing was present. There were no petechiae or purpura. An ejection systolic murmur of grade 3/6 was audible in the pulmonary area.

The child was conscious with normal cranial nerve and sensory system examination. Power was normal in the right upper and lower limb. It was grade 3/5 in the left upper limb and grade 2/5 in the left lower limb. Bilateral plantar reflexes were extensor with well-sustained ankle clonus. There were no meningeal or cerebellar signs. Other systemic examination was normal. Investigations done on the day of admission are shown in Table 1.

Table 1. Summary of haematologic and biochemical investigations

<table>
<thead>
<tr>
<th>Variables</th>
<th>1st day of admission</th>
<th>8th day of admission</th>
<th>Discharge</th>
</tr>
</thead>
<tbody>
<tr>
<td>Haemoglobin (gm/dl)</td>
<td>24.1</td>
<td>19.6</td>
<td>18.1</td>
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<tr>
<td>Haematocrit</td>
<td>75</td>
<td>59</td>
<td>54</td>
</tr>
<tr>
<td>Platelet count(per cumm)</td>
<td>53,000</td>
<td>95,000</td>
<td>70,000</td>
</tr>
<tr>
<td>Prothrombin time</td>
<td>42 sec/12 sec</td>
<td>20 sec/12 sec</td>
<td>22 sec/12 sec</td>
</tr>
<tr>
<td>Partial thromboplastin</td>
<td>56 sec/32 sec</td>
<td>38 sec/32 sec</td>
<td>42 sec/32 sec</td>
</tr>
<tr>
<td>Bleeding time (N: 1–6 min)</td>
<td>8 min</td>
<td>4 min</td>
<td>4 min</td>
</tr>
<tr>
<td>Serum bilirubin (mg/dl)</td>
<td>2.7</td>
<td>1.6</td>
<td>1.1</td>
</tr>
<tr>
<td>Serum aspartate aminotransferase (IU/L)</td>
<td>112</td>
<td>67</td>
<td>54</td>
</tr>
<tr>
<td>Serum alanine aminotransferase (IU/L)</td>
<td>120</td>
<td>78</td>
<td>57</td>
</tr>
</tbody>
</table>

The fibrinogen levels, D-dimer level, factor VII, factor VIII, and antithrombin III were normal. Renal function tests, blood sugar and serum electrolytes were normal. Arterial blood gas analysis revealed hypoxemia and saturation of 65%. Computed tomography (CT) scan of brain showed an isodense lesion (Hounsfield unit – 45) of 3.7×2.5×4.8 cm-sized peripherally-enhancing lesion in the right centrum semiovale reaching up to the right high parietal region with tiny adjacent satellite lesion with surrounding perilesional oedema suggestive of resolving hematoma with superadded infection. The lesion was without enhancing wall or definite borders, usually seen with abscess (Figure 1).
Figure 1. Computed tomography scan showing a 3.7×2.5×4.8 cm-sized peripherally-enhancing lesion (Hounsfield unit – 45) in the right high parietal region with surrounding perilesional oedema suggestive of a haematoma with superadded infection.

Echocardiography confirmed earlier findings. The child was started on intravenous vancomycin and metronidazole. In view of high haematocrit and the child being posted for surgery, phlebotomy was done five times over a period of 7 days. Fresh frozen plasma and platelet transfusion were given preoperatively. The child was taken for surgery on the eight admission day. A right parietal burr hole was done and 30 ml of blood admixed with pus was drained. Blood culture sent on the day of admission was negative.

Postoperatively he was started on physiotherapy and a repeat CT scan was done after 1 month which showed decrease in the size of the hematoma. He was discharged after 6 weeks of intravenous antibiotics. He was well on follow-up at 2 months and is currently awaiting corrective cardiac surgery.

Thromboembolism is generally considered as a cause when a patient of CCHD presents with a focal neurological deficit. Our case highlights the haemorrhagic tendencies associated with CCHD and the importance of considering them in the
differential diagnosis. Abnormal haemostasis was the cause of spontaneous CNS bleed in our patient.

Polycythemia due to chronic hypoxia is mainly responsible for these abnormalities which include thrombocytopenia, decreased production of coagulation factors and accelerated fibrinolysis. Both qualitative and quantitative defects in platelets occur in CCHD. Qualitative defects include abnormal aggregation of platelets in response to adenosine diphosphates, epinephrine and collagen, which is directly related to the degree of polycythemia. Quantitative defects appear when the haematocrit is more than 65% leading to the margination of platelets. High shear stress caused by hyperviscosity leads to formation of markedly smaller platelet aggregates.

Chronic hypoxia shortens platelet survival, affects megakaryocyte differentiation, maturation, and apoptosis via oxygen tension and alteration of the physiological regulator (thrombopoietin). Hypoxic damage to the liver and sluggishness of microcirculation caused by hyperviscosity causes deficient synthesis of the coagulation factors—i.e. II, V, VII, IX and X. Abnormal haemostasis resulting from a state of consumptive coagulopathy or disseminated intravascular coagulation has also been observed in some patients with elevated D-dimer levels.

Bleeding tendency is usually mild to moderate and is characterized by easy bruising, petechial haemorrhage, gingival bleeding, epistaxis and haemoptysis. However, serious and sometimes fatal bleeding can occur during haemostatic stress following trauma or surgery. In the usual clinical setting, phlebotomy is recommended only in patients with symptomatic hyperviscosity when haematocrit levels exceed 65%.

Symptoms related to hyperviscosity include headache, fatigue, faintness, dizziness, visual disturbance, paresthesia, irritability, myalgia, reduced mentation, and anorexia. Also, preoperatively, phlebotomy is recommended to reduce the haematocrit below 65%, which improves haemostasis and decreases the risk of postsurgical haemorrhage.

Volume replacement with an equal volume of 0.9% saline or colloid should be done whenever venesection is performed for preventing the acute fall in systemic blood flow, oxygen delivery, and cerebral perfusion. Platelet and fresh frozen plasma transfusion should be given if thrombocytopenia and coagulation abnormalities are present.

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Acknowledgements: We thank Dr Sandhya Kamath (Dean of our institution) for permitting us to publish this manuscript and Dr Mamta Manglani, Head of Department of Paediatrics for her guidance in the management of the case.

References: