Congenital haemophilia A presenting with subgaleal and intracranial haemorrhage following instrumental delivery

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DESCRIPTION

The case is of a male infant born by vacuum-assisted vaginal delivery at 38 weeks+3 days of gestation. The mother is a primigravida and was neither diabetic nor hypertensive. Her pregnancy course was uneventful. There was no family history of coagulopathy or bleeding disorder on the maternal or paternal side. She was presented to the delivery room with labour pain and rupture of membranes more than 18 hours before the delivery. She received one dose of penicillin G. At the delivery time, she was noted to have signs of fetal distress in the cardiotocography monitoring. On vaginal examination, the fetal head was located at +1 station. Kiwi ventouse was applied at the fetal flexion points, but it failed twice so it was removed. A silastic vacuum extractor was applied at the fetal flexion point. An episiotomy was done and the baby was delivered after 40 min from the fourth attempt. The baby was delivered in a good condition with a birth weight of 3030 g. Apgar score was 8 and 10 at 1 and 5 min, respectively. After delivery, the baby was examined by the neonatologist attending the delivery, who noted a scalp erosion and mild oozing controlled by pressure.

At the age of 7 hours, the baby was examined by the postnatal neonatologist who noted a soft tissue parietotemporal head swelling (figure 1). Skull X-ray was ordered and showed overlapped parietal bone over occipital bone with overlying soft tissue swelling. No bony fracture was noted. Complete blood count (CBC) was done and showed a haemoglobin (Hb) level of 125 g/L and platelet count of $234\,10^9$/L. At the age of 12 hours, the swelling was noted to be progressively increasing in size, so the baby was shifted to the neonatal intensive care unit (NICU) for further management. He remained haemodynamically stable during his postnatal and NICU stay. Head ultrasound was done in the NICU at the age of 14 hours and showed an extra-axial infratentorial haematoma at the left temporal region measuring 2.0 cm×1.4 cm with no midline shift. Subgaleal haematoma was also noted (figure 2). A follow-up CBC was done and showed a further drop in the Hb level to 97 g/L. Other CBC parameters including platelet count were normal. An urgent head CT scan was done at the age of 18 hours and showed bilateral subgaleal haematoma involving the temporal, parietal and occipital regions with evidence of left supratentorial bleed measuring 2.9 cm×1.4 cm. Another bleed was noted measuring 1.3 mm×0.6 mm in the left infratentorial region suggestive of tentorial subdural haematoma. No hydrocephalus, midline shift or mass effect was noted (figure 3).

Figure 1 Subgaleal haematoma involving the parietal (transverse arrow), temporal (vertical arrow) and occipital areas.

Given the obstetric history, sex of the baby and postnatal course, the neonatal team decided to investigate the baby for coagulopathy even with the absence of an index case in the family. Activated partial thromboplastin time was measured at that time and it was prolonged up to 102.8 s. Prothrombin time was 12.3 s, and the international normalised ratio was 1.2. Coagulation factor VIII was ordered, and it came back to be 0.4%, confirming the diagnosis of severe congenital haemophilia A.

The baby underwent packed red blood cells transfusion once and fresh frozen plasma three

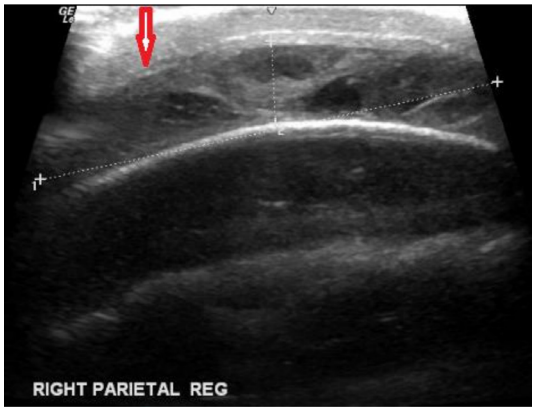

Figure 2 Skull ultrasound showing subgaleal haematoma in the right parietal region (vertical arrow).

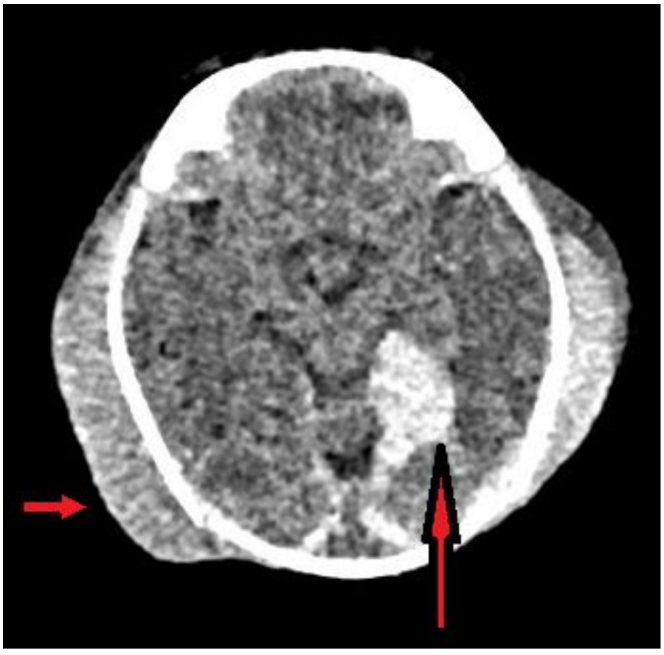

Figure 3 Head computed tomography (CT) CT scan showing bilateral subgaleal haematoma involving the temporal, parietal (transverse arrow) and occipital regions with evidence of left supratentorial bleed measuring 2.9 cm×1.4 cm (vertical arrow).

times. The paediatric haematologist was consulted and recombinant factor VIII was given to the baby for a total of 14 days; 50 IU/kg every 8 hours for 7 days, 50 IU/kg every 12 hours for 3 days and 30 IU/kg every 24 hours for 4 days. The baby was discharged at the age of 19 days when the factor VIII level reached 121%. Bethesda inhibitor assay antifactor VIII antibody is produced against functional epitopes of FVIII in about 30% of patients with severe haemophilia A, leading to inhibition of its procoagulant activity. It was done twice for the baby and it was 0.0 BU/mL.

Subgaleal haemorrhage is a serious neonatal condition that is caused by bleeding in the space between the occipitofrontalis muscle aponeurosis and periosteum of the skull. The most common cause of subgaleal haemorrhage is vacuum-assisted vaginal delivery. Based on the rate of increased head size, presence of jaundice and hypovolemia, it is classified into mild, moderate and severe. It might be lethal due to significant haemorrhagic anaemia and decompensated hypovolemic shock and should be cared for by the neonatal team very vigilantly. Independent risk factors associated with subgaleal haemorrhage include primiparity, vacuum duration, number of dislodgments, the prolonged second stage of labour (>120 min), fetal head station, presence of caput succedaneum, prolonged rupture of membranes (>12 hours) and meconium-stained amniotic fluid. It should be differentiated from other types of head swellings including caput succedaneum, cephalohaematoma and chignon.[1 2]

This is a rare presentation of congenital haemophilia A at birth without a family history. Congenital haemophilia is the most common congenital coagulation disorder affecting 1 per 5000 male infants.[1] Normal plasma levels of factor VIII range from 50% to 150% (0.5–1.5 U/mL). Haemophilia A is categorised according to the amount of factor VIII levels present: mild (5%–50%), moderate (1%–5%) and severe (<1%). Newborns with severe factor VIII deficiency might have many spontaneous bleeding episodes per month ranging from minor oral bleeds to extensive intracranial haemorrhage from minor head trauma. Treatment is based on the regular replacement of deficient coagulation factor VIII. Following the introduction of recombinant coagulation factor VIII, the treatment strategy of the disease has been shifted from episodic to prophylactic regular infusion of the factor concentrate that decreased mortality over time.[3–5]

Learning points

- Early recognition and close monitoring of subgaleal haematoma are crucial for survival.
- Coagulation studies are required to detect coagulopathy that might be the cause of the subgaleal haemorrhage. It should be considered standard practice in an enlarging subgaleal haemorrhage regardless of family history to early demonstrate any consumptive coagulopathy.
- In cases of family history of haemophilia, vaginal delivery and caesarean section carry similar risks of intracranial haemorrhages and major bleeds.

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