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Case Report

Subgaleal hematoma in hereditary coagulopathy haemophilia B: a case report

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ABSTRACT

Subgaleal haemorrhage, characterized by blood accumulation in the subgaleal space, can lead to significant blood loss and shock, often exacerbated by hereditary coagulopathies like haemophilia. A full-term male neonate, born at 39 weeks via spontaneous vaginal delivery, presented 12 hours post-birth with temporal swelling, periorbital edema, and raccoon eyes. Examination showed increased head circumference, severe anaemia, and elevated aPTT. Imaging confirmed subgaleal hematoma. The infant received packed red blood cells, fresh frozen plasma, factor IX, IV fluids, antibiotics, and phototherapy.

Keywords: Subgaleal haemorrhage, Hereditary coagulopathies, Haemophilia, Racoon eyes, Phototherapy

INTRODUCTION

Subgaleal haemorrhage is a collection of blood in the loose connective tissue of the subgaleal space, located between the epicranial aponeurosis and the periosteum. There is often an association with vacuum-assisted delivery. The mechanism of injury is most likely secondary to rupture of emissary veins connecting the dural sinuses within the skull and the superficial veins of the scalp. Extensive subgaleal bleeding can result in sequestration of more than 40% of the newborn's blood volume, which can potentially result in haemorrhagic shock. Subgaleal haemorrhages can present clinically with the triad of tachycardia, decreased haematocrit, and increasing occipital frontal circumference.[1] The mortality can be up to 14% due to haemorrhagic shock and is occasionally secondary to a hereditary coagulopathy(hemophilia). Because neither factor VIII nor factor IX crosses the placenta, bleeding symptoms may be present at birth. Between 1% and 4% of neonates with haemophilia have intracranial haemorrhages, mostly from birth trauma.[1] So we are reporting a case of subgaleal hematoma and establishing its association with inherited coagulation disorders.

CASE REPORT

A full-term male neonate born at 39 weeks of gestation by spontaneous vaginal delivery with vertex presentation and birth weight of 3.6 kg. Apgar scores are 7 and 8 at 1 min and 5 min respectively. Baby cried immediately at birth and breast fed in 1st hour of birth,1st dose of vit k was given. Antenatal history comprised of non-consanguineous marriage, 2nd gravida (G2 P1 L1 A0). No relevant history of any illness or drug intake is present. No history of diabetes and hypertension was present. No history of any coagulopathy or bleeding disorder was present at admission.

At day 2 of life, he was admitted in out born NICU with a progressive swelling over the temporal region which was from the nape of the neck to the forehead and progressed bilaterally (Figure 1). It started 12 hours after birth along with periorbital oedema, bluish discolouration of skin

over right pinna and right eye (racoon eye). He had marked irregular scalp oedema with a retro auricular hematoma. There was a 4 cm increase on head circumference (39 cm). On examination baby was pale, dull and lethargic, had feeble cry with subcostal retractions with audible grunt and taken on HFNC (fio2-40%, flow 4l/min) (Figure 2). Vitals were taken with HR-188/min spo2-79%, RR-70/min and normal pulse volume with delayed CRT. CNS examination was done, AF could not be appreciated due to swelling, tone was normal. Neonatal reflexes were as follows: suck and rooting-absent, moro's-partial, plantar and palmar grasp- poor, stepping and placing- absent. Other systemic examination was found to be normal.

All routine investigations were done, including CBC-Hb (4.4 g/dl), HCT (15.2%), total WBC (28.4K), PLT count (195K), PT (normal), aPTT (144.70). PRBC and FFP was transfused accordingly. CRPQ was found to be negative. Other routine investigations like urea (61.2 mg/dl), creatinine (1.8 mg/dl), calcium (8.6 mg/dl), sodium (148.8 mmol/l), potassium (4.5 mmol/l), total SBR (4.7 mg/dl increased to 11.2 mg/dl on day 4 and later normalized). All viral markers like HIV, HBsAg and HCV were negative. Factor VIII was 203.00% (ref-50%-100%) and factor IX activity Stuart power was 12.00% (ref-70.00%-120.00%). Factor VIII and XI activity Stuart power assays were also sent in other sibling which was found to be 26% and 12% respectively and a diagnosis of combined haemophilia A and B was made in other sibling. Initially USG cranium was done suggestive of moderately echogenic fluid collection crossing the suture line. For confirmation NCCT head was done suggestive of subgaleal hematoma (Figure 3).

Baby was started on iv fluids and antibiotics along with ionotropic support. It was followed by transfusion of packed cells and FFP. Phototherapy was given for hyperbilirubinemia. Factor IX was also given post confirmation of the diagnosis.

Figure 1: Swelling over the temporal region.

Figure 2: On examination baby was pale, dull and lethargic, had feeble cry with subcostal retractions with audible grunt and taken on HFNC (fio2-40%, flow 4l/min).

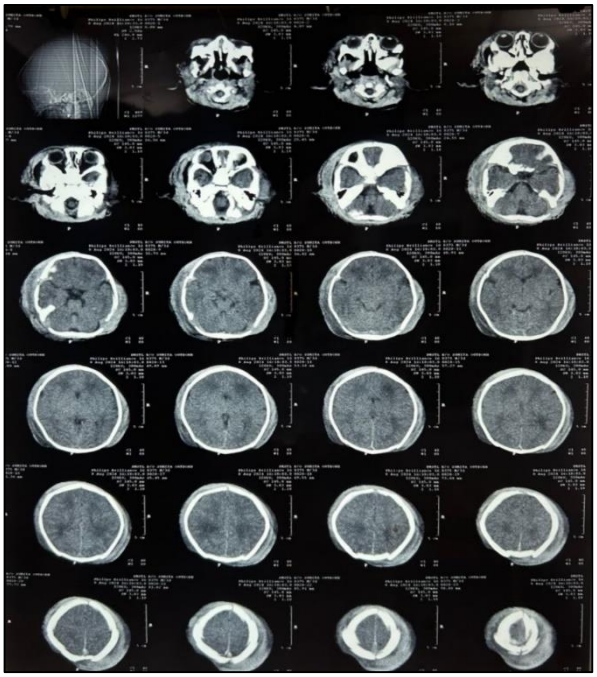

Figure 3: Subgaleal hematoma.

DISCUSSION

Subgaleal hematoma is haemorrhage under the aponeurosis of the scalp. It extends from the orbital ridges to the nape of the neck and laterally to the ears, the haemorrhage can spread across the entire calvarium. The initial presentation typically includes pallor, poor tone, and a fluctuant swelling on the scalp. The hematoma may grow slowly or increase rapidly and result in shock. The blood is resorbed slowly, and swelling gradually resolves.

The morbidity may be significant in infants with severe haemorrhage who require intensive care for this lesion. The mortality rate can be up to 14%. Death is attributed to significant volume loss, resulting in hypovolemic shock and coagulopathy.[2] When evaluating a neonate concerning for Haemophilia B, one must perform a thorough physical examination and obtain a detailed family history, CBC, coagulation profile, and imaging studies pertinent to examination findings.

Patients with haemophilia will present with an age-adjusted elevated aPTT and a normal prothrombin time, platelet count, and bleeding time. Factor 8 and 9 assays are done. Ideally, infants who are known to be at risk for haemophilia should have cord blood sent for diagnosis.[3] Prophylactic therapy for children with the severe form of the disease has improved prognosis over the years. Nevertheless, up to 25% of patients with severe haemophilia can suffer from below-average academic performance and behavioural issues, as protracted bleeds can cause developmental and cognitive abnormalities.[4]

In a case study Moreira A et al, who reported cases of 3 neonates presenting with acute, life-threatening haemorrhage who were subsequently diagnosed with severe haemophilia A (<1% factor VIII). The first infant was tachycardic, pale, and had a precipitous drop in his haemoglobin secondary to a subgaleal haemorrhage. The second patient sustained a splenic rupture, a sequela that has been reported in only 4 other neonatal cases. The last infant presented with tonic-clonic seizures and respiratory distress. Cranial imaging demonstrated extracranial and intracranial haemorrhage, complications that can result in 20% mortality. All 3 patients were successfully treated with clotting factor concentrate and blood products as was seen in this case.[5] But in our case, factor VIII was normal and factor XI was low.

Radovanović T et al reported that Subgaleal hemorrhage may be the first presentation of haemophilia A and reported a case of full-term male neonate was born at 40 weeks gestation and at the age of 23 hours, the baby became pale and lethargic. Large fluctuant swelling on his head was noted. He developed severe anaemia and hypovolemia as a result of massive subgaleal haemorrhage. After successful treatment, the baby fully recovered. They concluded that Infants without obvious risk factors for developing subgaleal haemorrhage should be evaluated for congenital bleeding disorder.[6]

A case report in Indian paediatrics by Dutta S et al of 3 kg baby was delivered by caesarean section after prolonged labour. He had massive subgaleal hematoma. He developed anaemia requiring packed cell transfusions and hyperbilirubinemia requiring a total of seven exchange transfusions and highly intensive phototherapy.[7]

CONCLUSION

Subgaleal haemorrhage in neonates, particularly with underlying coagulopathy, demands careful evaluation and management to address both the immediate haemorrhagic crisis and the underlying coagulation disorder.

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