A case of recurrent postpartum haemorrhage in three consecutive pregnancies due to isolated factor V deficiency.

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BACKGROUND

- Isolated factor V deficiency is a rare autosomal recessive disorder with its incidence of 1 in 100000.
- Its manifestations vary from easy bruising to fatal bleeding imposing a great risk on pregnancy in susceptible women.

THE CASE

- A 34 year old mother was transferred to a tertiary care unit in Sri Lanka on postpartum day 01 of her third pregnancy.
- It was following a vaginal delivery complicated by primary postpartum haemorrhage requiring multiple blood transfusions. There was no uterine hypotonia or retained products and delivery was uneventful and spontaneous without significant trauma.
- It was revealed that her previous two vaginal deliveries were also complicated by significant PPH.
- Her past medical, surgical, drug and family history were otherwise unremarkable.
- Examination revealed a 5x6 cm sized vulval haematoma evolved around episiotomy.
- Her haemoglobin and platelet count were normal.
- However both prothrombin time (PT=18.4 seconds) and activated partial thromboplastin time (APTT=47.1 seconds) were prolonged.
- Other coagulation tests, liver function tests and liver ultrasound scan were normal.
- Possible factor V deficiency was suggested by rotational thromboelastometry (ROTEM) which was confirmed by a subsequent factor V assay.
- She was managed with evacuation of haematoma, antibiotics, patient education and counselling. Interval sterilisation was performed later at her request.

DISCUSSION

- Postpartum haemorrhage particularly if recurrent may be due to a primary coagulation disorder.
- Rare, yet clinically significant coagulation disorders include deficiency of fibrinogen, prothrombin, factor V, factor VII, factor X, factor XI and factor XIII either as isolated or combined deficiency disorders.
- Factor V deficiency is inherited in an autosomal recessive manner mostly in a context of consanguinity.
- Patients with severe deficiency are either homozygous or compound heterozygous and present early within the first six months of life.
- .Heterozygous deficiency is usually asymptomatic and unrecognized despite prolonged PT and APTT value.
- In the absence of commercially available factor V preparations, fresh frozen plasma is useful for therapeutic correction during a bleeding episode. A factor V level between 10%-20% of normal is sufficient to prevent bleeding.
- Fibrinolytic inhibitors like - tranexamic acid may also be useful.
- The mode of delivery is determined by the obstetric condition and vaginal delivery is not contraindicated. The delivery should be managed in a tertiary care centre with availability of 24 hour transfusion services and haematologist inputs.
- Prognosis of these patients with timely interventions is generally good. Therefore actively seeking for an underlying cause instead of attributing it to commoner causes of obstetric haemorrhage is prudent to prevent unanticipated maternal morbidity and mortality.
- Genetic counselling should be offered to all patients, their families and offspring.

REFERENCES

1. Kadir R, Chi C, and Bolton-Mags P. Pregnancy and rare bleeding disorders. *Hemophilia* 2009; **15:** 990–1005.

2. Girolami A, Scanderalli R, Lombardi AM, et al. Pregnancy and oral contraceptives in factor V deficiency: a study of 22 patients and review of literature. *Hemophilia* 2005; **11**: 26–30.

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