Anaesthetic approach to epidural hematoma, after minor head trauma, in an infant with previously unknown fibrinogen deficiency

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Introduction

Congenital fibrinogen deficiency is a rare autosomal disorder of coagulation with unknown prevalence and diverse range of bleeding phenotypes. Only few cases of CNS bleeding in paediatric patients with fibrinogen deficiency are reported and are related with increased morbidity and mortality. The perioperative management of an infant with an acute epidural hematoma after minor head injury, with previous unknown congenital fibrinogen deficiency, is presented.

Case Report

A previously healthy 10months old infant (11.7 kg) was admitted to the emergency department after minor head trauma followed by vomiting episodes. The infant became lethargic and anisocoria was observed. Emergency brain CT revealed an epidural hematoma and the infant was transferred to the operating theatre for emergency craniotomy. The vital signs were HR 200/min, BP 85/38 mmHg, SpO₂ 100% and he was reacting only to painful stimulus. General anesthesia was induced and arterial and central venous catheter were inserted.

Preoperative Hb=7.8g/dl, PLT=271.000 and coagulation tests were normal (INR=1.33, APTT=26,8 sec). Major blood loss (>250ml) was recorded and resuscitation with 500ml crystalloids, 250ml of RBCs, 200ml of FFP and vasopressors was performed. Perioperatively new blood tests were ordered and arterial blood gases were repetitively assessed. At the end of operation another 50ml of blood were removed by the drainage resulting in severe hypotension. Despite the infusion of noradrenaline, bolus of adrenaline (100 mcg), crystalloids and 120ml RBCs were needed for haemodynamic stabilization.

Transfer of the patient to the PICU was decided. During discussion with the family about the major blood loss, the mother reported for the first time positive family history of fibrinogen deficiency. The results of previously obtained blood tests revealed fibrinogen=13mg/dl, Hct=18.2%, Hgb=6,2g/dl, PLT=110.000 and INR 2.36, APTT>120 sec. Therapy with fibrinogen concentrates was promptly initiated. During the stay in the PICU the patient suffered from epileptic seizures requiring treatment with anti-epileptic medication. The rest of the stay remained uncomplicated, and the infant was discharged without neurological injury.

Discussion

Fibrinogen has a key role in primary and secondary haemostasis and it is the first coagulation agent that is affected during haemorrhage¹. Normal plasma fibrinogen level is approximately 200-450 mg/dl.

Congenital fibrinogen deficiencies are either quantitative (afibrinogenemia–hypofibrinogenemia) or qualitative (dysfibrinogenemia). The afibrinogenemia incidence is estimated at 1:1.000.000, while the incidence of hypofibrinogenemia and dysfibrinogenemia is unknown and probably underdiagnosed. There is no consistent clinical presentation. The patient can experience bleeding episodes of varying severity or might be asymptomatic^{2,3}.

The treatment depends on the severity of bleeding tendency. Antifibrinolytic amino acids (e.g. tranexamic acid) may be used for less severe forms of haemorrhages. In major blood loss episode, administration of fibrinogen concentrates is the treatment of choice. Suggested dose(g) = desired increment in g L⁻¹ x plasma volume. Fresh frozen plasma and cryoprecipitate may be an alternative treatment. For major surgery, a fibrinogen level >100mg/dl is recommended until healing is completed².

Base excess in arterial blood gases was <-6mEq/L, which is related with hypofibrinogenemia during haemorrhage. However, in the reported case decision was made not to administer fibrinogen concentrates before measurement of fibrinogen plasma level due to lack of experience in fibrinogen concentrates use in infants. Therefore we opted for FFPs instead⁴.

Conclusion

Congenital fibrinogen deficiency:

- May result in life threatening hemorrhage
- Base excess < -6mEq/L indicates hypofibrinogenemia during hemorrhage
- Fibrinogen concentrate is the treatment of choice
- Fibrinogen dose(g) = desired increment in g L⁻¹ x plasma volume

References

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