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Case Report

Surgical Act in a Child with Congenital Afibrinogenemia and History of Post Transfusion Urticaria

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Abstract

Congenital afibrinogenemia is a very rare bleeding disorder, results from mutation that affects plasma fibrinogen concentration. Bleeding diathesis of varying severity frequently associated needs factor replacement as well as surgery act does. We describe the case of a 11-year-old female child diagnosed of congenital afibrinogenemia who presented an inguinal hernia and was posted for surgery repairment. Fresh frozen plasma was infused with antibrinolytic agent and antihistamine due to urticaria transfusion history in pre and post-operatively to the child. Patients with congenital afibrinogenemia should be managed by a comprehensive bleeding disorder care team. Anaesthesiologist, surgeons and haematologist worked together to manage this surgical emergency. Outcome was favourable, no bleeding nor urticaria occurred.

Keywords: *Congenital Afibrinogenemia; FFP; Fibrinogen; Antihistamine*

Introduction

Congenital afibrinogenemia is characterized by the complete absence of fibrinogen [1]. Fibrinogen is the precursor of the major protein constituent of the blood clot, fibrin. The absence of fibrinogen allows no clotting in front of any wound. Any surgical procedure constitutes a significant risk of bleeding that must be controlled and must be covered by a correct haemostatic line [2].

The purpose of this paper is to report how haemostasis was achieved during the repair of an inguinal hernia in a child with congenital afibrinogenemia within a history of post transfusion urticaria.

Case Report

H.E.R., a female child, 11 years old, Malagasy origin, has congenital afibrinogenemia. The disease was diagnosed at the age of 3 years. Prothrombin time (PT) and activated partial prothrombin time (aPTT) as well as the bleeding time were prolonged, the rate of fibrinogen was less than 0.1g /l. In her history, repetitive episodes of bleeding were reported such as epistaxis, spontaneous hematoma, secondary trauma, and sometimes mild bone pain.

The management of the disease was based on a transfusion of fresh frozen plasma, due to non availability of fibrinogen concentrate, with antifibrinolytic treatment based on tranexamic acid.

The child showed at an early age an inguinal hernia. In order to avoid potential complications, surgical repair has been prescribed. The surgical team and the anaesthesia resuscitation team were informed of the patient's condition and the need for replacement therapy to provide the missing fibrinogen by plasma and urticaria transfusion history.

Haemostatic management for the surgical procedure was the following: 24 hours before surgery, transfusion of two frozen fresh plasma bags, just before surgery one FFP bag, continued with another FFP bag during the day of surgery, transfusion of one FFP bag the next day and every three days a bag of FFP per day.

Antifibrinolytic adjuvant treatment was instituted 24 hours before the intervention based on tranexamic acid 40 mg/kg/ day continued for a week.

Since the patient has a history of post-transfusion urticaria, an antihistamine treatment has been instituted as well 24 hours before the procedure: dexchlorpheniramine 1 vial in IV twice a day then a tablet 2 mg three times a day as long as FFP was infused.

Ultrasound monitoring of the operated area ensured the absence of blood collection postoperatively according to the surgical opinion.

Outcome was favourable and complete healing achieved in tend days.

Comment

Replacement therapy was required to provide the fibrinogen needed to form a fibrin clot as surgical act was necessary in the context of afibrinogenemia [3]. Basically, plasma contains 2 to 4 g/l. FFP is the adequat source of fibrinogen. Nevertheless, plasma contain other proteins which can lead to allergic condition such as urticaria.

An inguinal hernia is found in both boys and girls. But the reported case is rare because it was found in a child with bleeding disorder and a history of transfusion urticaria. The fibrinogen deficiency had to be replaced within the scope of surgery from plasma. Two molecules had to be made in the management of this surgical case: the antihistamine to be able to continue to transfuse the plasma despite the allergy and antifibrinolytic to keep the clot in place longer [4].

All this procedure avoided the bleeding complications of a risky surgery since the patient was afibrinogenemic and allergic. The favourable evolution confirms a good cooperation between surgeon, anaesthetist and haematologist.

Conclusion

A patient with congenital afibrinogenemia should be followed up by a comprehensive bleeding disorder care team. Replacement therapy with plasma derived fibrinogen concentrate is the treatment of choice but fresh frozen plasma is alternative treatment that should be used only when fibrinogen concentrate is not available. Antifibrinolytic and antihistamine drugs were used in addition for this female child and the outcome was favourable because the anaesthesiologist, surgeons and haematologists worked in unison.

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