

A case of congenital factor X deficiency in Côte d'Ivoire, West Africa

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SUMMARY

Congenital factor X deficiency is a bleeding disorder never reported in sub-Saharan Africa. It is seen most often in communities where consanguineous marriages are accepted. The clinical picture is mostly due to hematoma and mucosal bleeding well controlled by derivatives of progesterone. In our context, the management of bleeding is done with fresh frozen plasma, available in most African countries.

Keywords: factor X deficiency, Africa, Cote d'Ivoire, Bleeding, Fresh Frozen Plasma.

INTRODUCTION

Congenital factor X deficiency is an inherited bleeding disorder reported for the first time in mid-1950 in subjects with a hemorrhagic syndrome resembling factor VII deficiency. ¹ Factor X or Stuart-Prower factor (names of the first two patients in whom the disease was identified) ¹ is a vitamin-K-dependent factor whose constitutional deficiency is rare. ² It is a condition insufficiently reported in Sub-Saharan Africa. We report a case from the clinical hematology department of Yopougon University Hospital (Abidjan, Côte d'Ivoire).

CLINICAL CASE

A seven year-old girl of Malinke ethnic group was seen at the haematology clinic February 13, 2015 for persistent gingival bleeding after minimal dental trauma. She had a history of repeated gingival bleeding after mild traumatic bruising which usually regressed after whole blood transfusions.

The father and mother are cousins. The father frequently had bruises and mucosal bleeding until the age of 10. Two of the older brothers of the patient died after heavy bleeding. Physical examination revealed pallor of the conjunctivae, with no features of symptomatic

anaemia. A small gingival bleeding wound was noted.

Full blood counts showed anaemia with hemoglobin level of 6.9 g/dl. Bleeding time, thrombin time and fibrinogen level were normal. Prothrombin time was 10%. The activated partial thromboplastin time was extended. These tests were corrected when normal control plasma was added to our patient's plasma (mixing study). The immunoassays of factors II and V were normal unlike that of factor X, which was very low, lower than 1% (Pasteur laboratory Cerba, Paris). Assays of factor X from the father and mother respectively showed 2% and 4%. The samples were collected in the tube containing trisodium citrate. The samples were centrifuged at 3000 rpm/min for 15 min at a constant temperature maintained between 18 and 22 ° C. Plasmas obtained were decanted and frozen immediately, then transported to France in a suitable device containing dry ice provided by the laboratory Cerba Pasteur in Paris. The assays of the activity of the factors were performed by chronometer by this laboratory.

Therapeutically, our patient received two units of fresh frozen plasma, an antibiotic and iron treatment (iron: 10 mg/kg/day). The bleeding stopped.

DISCUSSION

Congenital factor X deficiency is a very rare bleeding disorder affecting approximately one person per two million.² As far as we know, only 50 cases have been described in the literature.³ In Ivory Coast, the disease exists but has never been reported before.

Factor X is a vitamin-k dependent factor whose synthesis is controlled by a gene on chromosome 13. The disease is transmitted in a recessive mode and is prevalent in communities where consanguineous marriages are practiced,³ as shown in our case.⁴ The age of first symptoms is between 0 and 4 years.² This is most often by way of spontaneous mucosal bleeding. Haemarthrosis is rare and when it exists, it is often secondary to trauma.⁵ In 1960, Brody and Stuart reported the case of a 27 year-old woman who often had epistaxis, gingival bleeding or haematuria. In the third trimester of her first pregnancy, all symptoms had disappeared and the prothrombin time that was extended before pregnancy (170 seconds) was significantly reduced (22 seconds) until delivery. They then suggested a correlation between hormones and certain clotting factors.² The same woman was

treated four years later without any pregnancy by Haber with a progesterone derivative (norethynodrel 10mg/day) without the occurrence of any bleeding event. This derivative of progesterone had had the same effects on the occurrence of bleeding as well as on the pregnancy.⁵ It is also known that hypercoagulability during pregnancy is due to an increase in fibrinogen and Factors II, VII, IX, and X.⁶⁻⁸

Factor X is present in fresh frozen plasma and prothrombin complex (proconvertin, Proacelerin, Stuart factor, antihaemophilic factor B).⁹⁻¹¹ In our context, fresh frozen plasma is available. In addition, the use of anti fibrinolytic substances is recommended especially during gingival bleeding or during tooth extraction because saliva naturally contains fibrinolytic enzymes.¹

CONCLUSION

These observations show that congenital factor X deficiency, although uncommon, exists in Africa and especially in communities that accept consanguinity. Its management is possible with the therapeutic methods available.

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