Congenital factor X deficiency of coagulation revealed by epistaxis

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The congenital factor X (FX) is a plasmatic glycoprotein, which plays a crucial role in hemorrhage in child. It could be acquired or congenital. The main hemorrhagic manifestations are epistaxis, hematuria, and menorrhagia in females, sometimes hemarthrosis even craniocerebral hemorrhage. The measurement of the FX rate in the blood allows the diagnosis. The healthcare consists of perfusion using prothrombin complex concentrate (PCC). In North Africa, no FX case was reported. Therefore, our goal is to report a new case of congenital FX of haemorrhage in our Moroccan female patient.

She is from Amazigh ethno-linguistic group; her parents personal or familial antecedents of bleeding or hepatic injury were recorded. She was presented with epistaxis, which was installed one week earlier, the bleeding occurred in a context of apyrexia and maintenance of the general state. No medicamentous treatment carried out with the patient according to the parents. The examination shows pallor without icteric or cutaneous eruption neither other signs of bleedings, nor adenopathy. A stable hemodynamic state without hepatosplenomegaly was noticed. The remaining examination is completely normal. The initial assessment carried out shows a normochromia normocytic anemia at 6.8 g/L, a low prothrombin rate time. The platelets rate is normal. The remaining hepatic assessment is strictly normal. The rate of the plasma in FX. The urgent action to be considered consisted of transfusing the child by red blood cells and frozen fresh plasma, this treatment allowed stopping the hemorrhage and the control of PR was as the factor of Stuart. It is a very rare coagulation abnormality characterized by an autosomal recessive hereditary attribute. Few authors reported the pathology in terms of isolated case or in terms series of patients.1 So far, the clinical expressions are not well established due to the rarity of this pathology. The prevalence in the general population is one per one million.2

malabsorption syndrome, a medicamentous intake,3 a poison ingestion1 and amylose.

is characterized by a great heterogeneity in clinical, phenotypic and genotypic aspects. The prevalence homozygote or heterozygote is approximately one per 2000. The responsible gene would be localized at heterozygotes (FX San Antonio) or of homozygotes

The coagulation factor could be an isolated one such as the case of our observation, or associated to other factors in particular coagulation factor VII or more complex factors FII, VII, IX and X.1

the symptoms result from the site of change in the gene. The heterozygotes are generally asymptomatic; hemorrhages are complicating the aggressive surgical acts without hemostasis or serious traumasisms. At the composite heterozygotes and homozygotes, the hemorrhagic manifestations are related to the residual of the hemorrhagic syndrome are correlated with the 1,2 The epistaxis is the most frequent symptom, followed by menorrhagia, extierized hemorrhages are principally represented by the hemarthrosis and the hematoma.1 (Table I in the severe forms.2 In addition, a hemorrhage observed at the fall of the umbilical cord might direct

1 The diagnosis is evoked when a quick time is lengthened (or rate of prothrombin lowered) associated to lengthened activated cephalin time. It can be also evoked when Stypven time is lengthened (activation of FX by the venom of Russel viper). The thrombin time is normal. Currently, with progress of molecular biology, the FX of the FX: C and the FX: Ag using endogenous and
exogenous methods. The rate of FX: C allows the
classification of the deficit, whatever the character
of the molecular anomaly. The deficit is severe for
rates of FX: C lower than 1%, moderated for rates
between 1 and 5% and minor for rates higher than
5%. The congenital deficit has to be distinguished
from the isolated deficit in FX observed in primitive
amylose, and more rarely the secondary amylose with
of FX coagulation, which is qualitatively normal, but
in smaller quantity (such as in our observation). The
normalization is acquired by the third month. The
indication of a substitutive treatment depends on the
hemorrhagic risk, which can be estimated by the type
of intervention, the severity of hemorrhages and the
FX concentration. There are 2 sources of FX, frozen
the human complex concentrates, which contains the
deficit.

The posology and the duration of the treatment
of the bleeding and the type of treatment to be started
(preventive or curative) as well as clinical and
biological state of the patient. For the constitutional

coagulation is a rare affection but of real gravity in the
without other obvious causes of hemorrhage in patients
with family antecedents of bleeding would evoke the
diagnosis, and dosing the rate of FX. The treatment
frozen fresh plasma remains very useful. The surgical
acts must be covered by higher dose than those used
usually used, this is useful to limit the blood losses.

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References

of bleeding symptoms in 32 Iranian patients. Br J Haematol

Dev

Brain

Thromb Haemost

Blood

combine en facteur VII et X revele par un allongement du
Arch Pediatr

<table>
<thead>
<tr>
<th>Symptom</th>
<th>All patients (n = 32)</th>
<th>Factor X: c &lt; 1% (n = 18)</th>
<th>Factor X: c 1-5% (n = 9)</th>
<th>Factor X: c 6-10% (n = 5)</th>
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<tbody>
<tr>
<td>Hematuria</td>
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<td>Hematoma</td>
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Table 1 - Prevalence of bleeding symptoms in congenital factor X deficiency (1).