ABSTRACT

The intracranial haemorrhage (ICH) of the term new-born is a rare pathology; the prevalence is estimated at 2% of live births. The clinical manifestations are variable and not specific. The causes of ICH are multiple and often intricate, the main pathophysiological mechanisms are deregulations of cerebral flow, obstruction of the vessels or intravascular coagulation or a direct injury from trauma. We report the case of a new-born at 30 hours of life admitted to our service (Neonatal resuscitation service and intensive care at Children's Hospital Rabat) for the management of respiratory distress on hypovolemic shock following which the clinical and biological examination was in favor of severe hemophilia A with factor VIII at 0.80%.

Keywords: Hemophilia; hypovolemic; haemorrhage; neonatal resuscitation.

1. INTRODUCTION

Bleeding is a common neonatal emergency, which can result from several causes, either from a primary haemostasis disorder or from a coagulation disorder. Hemophilia is one of these causes; it is an inherited bleeding disorder most often caused by deficiencies in clotting factors;
VIII (FVIII) for hemophilia A and IX (FIX) for hemophilia B. They most often affect men; females are carriers. Depending on the concentration of FVIII or FIX, these coagulation disorders are divided into: severe, when the plasma levels are less than 1%; moderate, when the rates are 1 to 4%; or light when the rates are 5 to 40% iii. Spontaneous bleeding in muscles and joints are typical symptoms of hemophilia, but intracranial haemorrhage is the most serious event that can occur in hemophilia, resulting in high rates of death and disability iiiiv. Mortality caused by ICH is still around 20% v.

We report the observation of a new-born baby admitted on D2 of life in a state of hypovolemic shock with respiratory distress, arterial hypotension and severe mucocutaneous pallor associated with an enormous serosanguineous lump whose clinical and paraclinical diagnosis was in favor of a severe hemophilia A. The birth process is the first hemostatic challenge for a new-born baby with severe hemophilia A because it carries a risk of one of the most devastating types of bleeding which is intracranial haemorrhage (ICB). The frequency of HIC in new-borns depends on several factors, such as the health status of the study population, knowledge of the mother's carrier status, obstetric guidelines in the management of female carriers, mode of delivery and finally, monitoring and routine management of new-borns with known or suspected hemophilia vi.

The aim of our article is to study the degree of severity caused by severe factor A deficiency (severe hemophilia A).

2. PRESENTATION OF CASE

This is a new-born baby admitted to our service at the d2 of life of a male from an out-of-consanguineous marriage with no particular family history, in particular no hemophilia. The pregnancy was brought to term without incident; a medicalized vacuum-assisted vaginal delivery was conducted. The new-born presented a good adaptation to extra uterine life and he was therefore returned to the mother. He was under exclusive breastfeeding. The new-born received his prophylactic dose of vitamin K in the delivery room. He was admitted in a state of hypovolemic shock at h30 of life with: tachycardia, arterial hypotension, polyneea, respiratory distress, mucocutaneous pallor, the patient was bloodless on an icteric complexion. (Fig. 1).

Clinical examination noted a temperature of 37.6, jaundice on a pale background and bruising on the face, especially on the forehead. (Fig. 2) Respiratory distress rated 2/10 according to the Silvermanvii score made up of an intercostal in drawing and flapping of the wings of the nose. The neurological examination found axial hypotonia with a serosanguineous lump and macrocrania with a head circumference of 37.5 cm. (Fig. 3) The diagnosis of traumatic brain haemorrhage associated with a haemostatic disorder was suspected. Brain CT was performed revealing a punctiform vernal hematoma and large bilateral subgaleal hematomas extending to the nape of the neck with slight cerebral edema. (Fig. 4).

The biological workup revealed severe anaemia with a haemoglobin level of 4.9 g / 100ml, and a normal platelet count of 150,000. The haemostasis assessment was disturbed with an PTT (Partial thromboplastin time) prolonged at 102 secs and a normal PT (Prothrombin time) at 64%. Pending the assessment, the new-born was put on injectable vitamin K at a rate of 10 mg and transfused with a red blood cell and put on anticonvulsants, antibiotics based on amoxicillin, meningeal dose as well as an analgesic treatment based on paracetamol.

During his hospitalization, the evolution was marked by the improvement of the respiratory distress; the patient became eupneic after 36
hours of his hospitalization. We also noted an improvement in the neurological state, a good reflex of sucking, the new-born became more reactive, the cranial perimeter went from 37.5 to 36 cm. The haemostasis assessment carried out initially with assay of coagulation factors revealed a factor VIII level of 0.80% (VN 70 to 180). All other factors were at normal rates. The diagnosis of haemophilia A in its severe form with neonatal revelation was retained.

After discussing our case in the haematology staff, we decided not to treat the patient for the moment except for complications, to vaccinate him according to the national immunization program, however all the vaccines should be administered by strict subcutaneous route. With respect for good practices, namely compressing immediately after the vaccine, putting on ice cubes and monitoring the injection site.

The evolution has been favorable. The new-born was seen in consultation at the age of 1 month and then at 3 months, his neurological examination was normal.

It was therefore a severe haemophilia A revealed in the neonatal period by a state of hypovolemic shock following anaemia related to cranial damage.

3. DISCUSSION

Coagulation disorders represent 32.1% of the aetiologies of cerebral haemorrhages, and are of several types giving different clinical presentations and lesions. Haemophilia is the most common condition that causes brain haemorrhages. Most of the time, it causes subdural hematomas but also parenchymal or cerebellar haemorrhages, often complicated by sequelae. These brain haemorrhages occur in 1 to 5% of births to children with haemophilia.

The frequency of neonatal intracranial haemorrhages (HIC) caused by haemophilia has been underestimated for a long time, no doubt because of the poor consideration of the numbers of deaths and the lack of a diagnosis of haemophilia made in these circumstances. The haemorrhagic risk (HIC and/or serosanguine lump) is increased by instrumental extraction manoeuvres (forceps, suction cup). In our case it was the use of the suction cup responsible for the serosanguine lump revealing the disease. New-born diagnosis of haemophilia A is rare, reported in less than 10% of haemophilia cases. The clinical signs are early occurring in the first days of life, as described in our patient from birth. It is therefore necessary to know how to detect family risk factors in order to adapt the mode of delivery in case of doubt. In addition, the cerebral haemorrhage being able to be a mode of revelation of haemophilia, it is thus necessary to know how to look for it in front of any unexplained HIC, even not to hesitate to treat it as such while awaiting the diagnostic proof. The severity of the haemorrhagic manifestations correlates with that of the clotting factor.
deficiency, it should be noted that the constitutional deficiency in factor VIII: Constitutional haemophilia A gives the most severe manifestations, namely a state of hypovolemic shock as described in our patient. It is an inherited disease of coagulation, transmission linked to sex. It affects around 1/5000 male births and is characterized by the occurrence of so-called internalized haemorrhagic manifestations, mainly muscular and articular but also of the central nervous system. Genetic abnormalities are carried by the F8 gene on the X chromosome (Xq28). These molecular abnormalities are deletions, point mutations, inversions (inversions in intron 22 = 50% of severe haemophilia A). The biological deficit is variable, defining several types of haemophilia:

- Severe haemophilia A: factor VIII level <1;
- Moderate haemophilia A: factor VIII level and between 1 and 5%;
- Haemophilia A minimal: factor VIII rate between 5 and 30%.

In our case, the severity of the clinical picture along with the notion of using instrumental manoeuvres during childbirth explains and reflects the severity of the disease. In fact, the severity of the haemorrhagic manifestations is directly linked to the level of circulating factor VIII.

4. CONCLUSION

Intracranial haemorrhage is reported in 1 to 5% of new-borns with haemophilia. They are the most serious of the problems that can occur in people with hemophilia. The lack of consensus on this topic demonstrates the need for further research on the care of new-borns with haemophilia and for sharing neonatal-revealing cases to learn useful lessons for practice.

CONSENT

As per international standard, parental written consent has been collected and preserved by the authors.
ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the authors.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES