Abstract

Hemophilia B is a rare and high-risk hemorrhagic disease. In emergency, maintaining a factor IX rate in a range to achieve the necessary action while ensuring the safety of the patient is the main challenge of the health care team. Post traumatic hemothorax in a hemophiliac B is not a common situation. We report the case of a great abundance hemothorax after a chest wound in a young hemophiliac B.

Keywords: Hemothorax; Thoracic Trauma; Hemophilia B; Hemostasis; Therapeutic Protocol

Abbreviations: AHF: Anti-Hemophilic Factor; APTT: Activated Partial Thromboplastin Time; FIX: Factor IX; IU: International Unit; Kg: Kilogram

Introduction

Haemophilia B is an X-linked constitutional haemorrhagic disease characterized by a quantitative or qualitative factor IX deficiency. It constitutes 15 to 20% of all haemophilia cases [1]. The haemorrhagic accidents can be spontaneous or post-traumatic, and concern the joints in 70% of cases [2]. Spontaneous or post-traumatic hemothorax is exceptionally reported. We describe the case of a young patient with hemophilia B, who suffered a penetrating chest traumatism causing a hemothorax of great abundance.

Observation

This is a 17 years old patient, followed since age 8 in pediatrics for severe haemophilia B with 2.3% factor IX (FIX), who was stabbed by a sharp object to the chest a day prior to admission, causing him chest pain and dyspnea that worsens in supine position. Upon admission, he was polyneic at 32 cycles / min, tachycardiac at 124 beats / min, normotensive, he saturated at 96% in the air, and his conjunctiva were slightly discolored, with the presence of a punctiform wound at the 7th intercostal space on the mid-axillary line associated with a hematoma and a fluid effusion syndrome of the entire left hemithorax. The abdomen was slightly sensitive, with no dullness. A Thoraco-abdomino-pelvic angio-CT scan was made upfront and showed a left hemothorax of great abundance, with extravasation of the contrast medium by the 7th left intercostal artery (Figure 1). The biological assessment showed anemia at 9.9 g / dl, APTT (activated partial thromboplastin time) at 50 and Prothrombin ratio at 56%. Blood grouping was O +. The dosage of factor IX is of no interest, because the hemophiliac keeps the same factor rate all his life.
The patient was hospitalized in intensive care unit, under monitoring of blood pressure, heart and respiratory rate, and oxygen saturation at the ambient air for better monitoring of his hemodynamic state and pain management. In consultation with the internal medicine department, factor IX supplementation was administered to the patient according to the following scheme: slow intravenous daily infusions of FIX, at the dose of 60 IU / kg / 24h which corresponds to 3500 IU / 24h in a single injection in order to reach a FIX rate ≥60%. The factor IX used was plasmatic of human origin with a half-life of 29 hours. During 4 days of hospitalization, and under monitoring of the vital functions, a dosage of the FIX rate was done daily after each infusion of the FIX. At Day + 4, the rate of the latter was 119.7%, which allowed us to predict the necessary hemostatic gesture.

To be ready to cope with any complication, the patient was admitted to the operating room, under monitoring, where he received a 5th dose of FIX, then 30 minutes later, an auxiliary thoracic drainage was performed to drain 2900 ml of blackish haematic fluid, with a marked immediate clinical improvement. Surgical exploration was therefore not necessary. Chest chest X-ray 6 hours after drainage was satisfactory, with a return of the lung to the wall, and a complete evacuation of the haemothorax (Figure 2). The patient remained under surveillance for 48 hours. He was supposed to receive regular supplementation of half the dose of FIX (30 IU / kg) to ensure haemostasis after drainage, but following the exhaustion of the hospital stock in FIX, he received only one half dose before drain removal to prevent hemorrhagic risk during and after ablation. During this whole process, the patient has not been transfused to avoid overload of the contralateral lung which will worsen his breathing difficulty. No antibiotics or other treatments were administered. After discharge, the patient presented to the control in a very good clinical condition, and in a perfect radiological evolution.

**Discussion**

Also called Christmas disease, hemophilia B is an X-linked recessive disorder that results from a congenital deficiency or absence of coagulation factor IX (FIX). It is a very rare pathological entity, its incidence is about 1/25 000 live births and represents 15% to 20% of cases of haemophilia [1,3]. Depending on the circulating anti-hemophilic factor (AHF), there are 3 forms of hemophilia: the severe form if the rate is less than 1%, moderate if the rate is between 2% and 5%, and the minor form if the rate is between 5% and 30%. Even in the minor form, the haemophiliac patient has a tendency to bleed spontaneously or following minimal trauma [1]. Generally bleeding is articular at the knee, ankle or elbow [1,4]. Externalized or life-threatening and / or functional haemorrhage is rare (Table1) [3-5]. Hemothorax of high abundance after penetrating trauma of the thorax, in a hemophiliac B patient endangers its vital and functional prognosis. This clinical situation is not a common emergency situation, which puts the health care team in front of two big challenges: On one hand, treating the hemothorax to relieve the patient, release the lung and allow it to re-spread normally, and on the other hand managing the situation of hemophilia [7,8].

<table>
<thead>
<tr>
<th>Site of Bleeding</th>
<th>Approximate Frequency</th>
</tr>
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<tbody>
<tr>
<td>Hemarthrosis:</td>
<td>70 % a 80 %</td>
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<tr>
<td>• More common in the joints : ankles, knees and elbows</td>
<td></td>
</tr>
<tr>
<td>• Less common in other joints: shoulders, wrists and hips</td>
<td></td>
</tr>
<tr>
<td>Muscle</td>
<td>10 % a 20 %</td>
</tr>
<tr>
<td>Other major bleeding</td>
<td>5 % a 10%</td>
</tr>
<tr>
<td>Central nervous system (CNS)</td>
<td>&lt;5 %</td>
</tr>
<tr>
<td>Newborn bleeding:</td>
<td>3.5%</td>
</tr>
<tr>
<td>Umbilical bleeding, cephalohematoma, post venipuncture, intramuscular hematoma</td>
<td></td>
</tr>
</tbody>
</table>

**Table 1: Approximate frequency of different bleeding sites [2,6]**

![Figure 2: Post-drainage chest X-ray showing the return of the lung to the wall](image)
Penetrating chest trauma is a very common cause of emergency room visits, but when the patient is hemophilic the situation becomes worrying. The care is made between emergency physician, thoracic surgeon, internist doctor and resuscitator, and it is based on the control of the hemorrhagic risk by the rapid administration of the substitution treatment.

Conclusion

In our situation, this attitude is not enough, because it is necessary to ensure hemostasis, before, during and after the haemostatic gestt by whether it is a drainage that is considered an invasive procedure for a hemophilic patient B, or a hemostasis thoracotomy. The target level of factor IX to be achieved before and after any action depends on its risk of bleeding [1,3,4,12,13] [Table 3]. In our case, considering that the proposed action is at high risk, it is necessary to ensure a preoperative rate of factor IX between 50 and 100%, and an immediate postoperative rate between 80% and 100%. To achieve this goal, a 24-hour infusion of factor IX will be administered in slow IV. The dose will be calculated according to the following formula [1,4]: Dose = [weight in kg x by expected increase].

A rare situation, that is necessary to keep in mind, is the hemophilic B carriers of inhibitory antibodies of factor IX. They represent only 3% of cases. Inhibitors are described as either a low titer (<5 Bethesda units) or a high titer (> 5 Bethesda units). Inhibitory antibodies of low titer can be overcome by the infusion of doses of factor substitutes, but high titer inhibitors generally require alternative factor products such as active recombinant Factor VII and active prothrombin complex concentrates. In unknown patients carrying the inhibitory Ab, anaphylactic shock may occur after supplementation with factor IX [1,3,14,15,16]. After controlling for bleeding and providing hemostasis, the management joins the usual care and monitoring protocols.

Conclusion

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References

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