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Post-Traumatic Hemothorax of Great Abundance in a Hemophiliac B: What Management?

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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: Activted 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 polypneic 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 (activted 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.

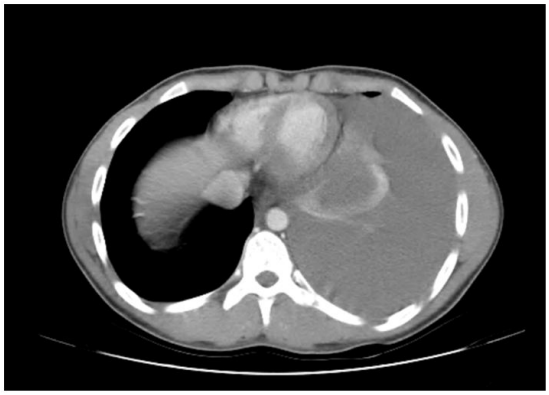

Figure 1: Thoracic CT scan with contrast injection, showing the hemothorax of great abundance with extravasation of the contrast medium through an intercostal artery

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 axillary 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.

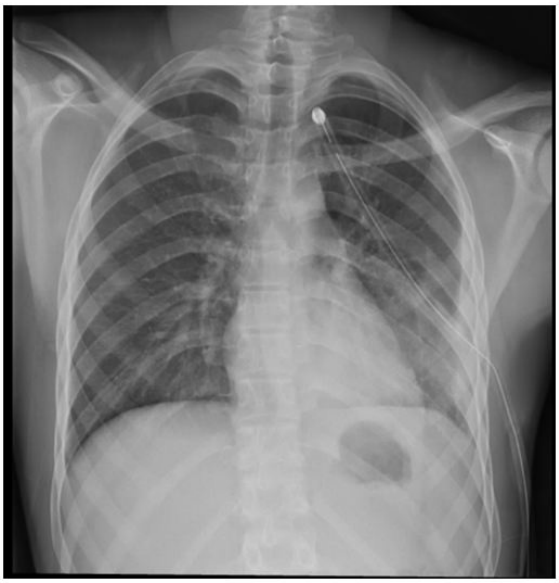

Figure 2: Post-drainage chest X-ray showing the return of the lung to the wall

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].

Site of Bleeding	Approximate Frequency
Hemarthrosis: •More common in the joints : ankles., knees and elbows •less common in other joints: shoulders, wrists and hips	70 % a 80 %
Muscle	10 % a 20 %
Other major bleeding	5 % a10%
Central nervous system (CNS)	<5 %
Newborn bleeding: Umbilical bleeding, cephalohematoma, post venipuncture, intramuscular hematoma	3,5%

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

In all published studies, which have treated the subject of bleeding in patients with haemophilia in general and type B in particular, hemothorax whether spontaneous or post-traumatic is reported in less than a dozen cases [7-11]. Management of this situation is not standardized [4,9]. Maintaining a factor IX rate within a range to achieve the necessary gesture while ensuring the safety of the patient is a simple equation, but difficult to ensure. In all the cases reported in the literature, no invasive procedure has been carried out probably for fear of not being able to control the bleeding, especially in the absence of the necessary anti-hemophilic products. Nowadays, the better knowledge of the pathology (Table 2) and the availability of the different anti-hemophilic agents, helped to ensure the equation of care. The biggest problem in a hemophiliac who bleeds in emergencies, is to restore haemostasis by providing as soon as possible the missing anti-haemophilia factor, which will subsequently treat the patient according to the usual protocol of care [4].

- A haemophiliac has the same level of factor IX all his life.
- Hemophiliac B does not bleed a lot, but a long time.
-The problem of product choice does not arise because a hemophiliac B always uses the same product.
-Never make an invasive gesture in a hemophiliac B before supplying it with an antihemophilic factor.
-A hemophiliac B who has received antihemophilic factor has normal hemostasis.
-There is no immediate risk of overdose with the antihemophilic factor.

Table 2: Basic knowledge of how to manage a hemophiliac B in emergencies [4]

In our situation, this attitude is not enough, because it is necessary to ensure hemostasis, before, during and after the haemostatic gesture whether it is a drainage that is considered an invasive procedure for a hemophiliac 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.

	Low risk surgery	High risk surgery
Preoperative desired rate	30%-80%	50%- 100%
Postoperative desired rate	30%-80% for 1 to 5 days depending on the type of surgery	80%-100% from day l to day 3
		60%-80% from day 4 to day 6
		40%-60% starting from day 7

Table 3: Desired factor IX rate in a patient with hemophilia B according to the bleeding risk of surgery [3,12,13].

Conclusion

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.

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