

Severe Thoracic Trauma in Hemophilia-A Patient with an Inhibitor

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Abstract

Hemophilia is a congenital bleeding disorder caused by the absence or deficiency of a coagulation factor. Treatment consists of intravenous administration of the deficient factor VIII in the case of hemophilia A or factor IX if it is hemophilia B. The presence of alloantibodies (inhibitors) can make these factors less effective. The management of these patients becomes difficult especially in the context of the emergency. Our clinical case illustrates the difficulty of making therapeutic decisions in the face of this kind of patient with a focus on therapeutic alternatives.

Keywords: Haemophilia; Inhibitor; Thoracic Trauma

Introduction

Hemophilia A is a congenital bleeding disorder characterized by a deficiency of factor VIII, and in whom infusion of the deficient factor has considerably reduced the risk of hemorrhage linked to this pathology. Nevertheless, the presence of an inhibitor may limit the effectiveness of factor infusion. Inhibitors are a major complication in the treatment of haemophilia. The cumulative incidence in cases of severe haemophilia A is reported to be 20 to 30% [1]. The management of these patients in the context of trauma requires a different strategy from other cases of hemophilia because of the high risk of bleeding and the potential for factor VIII ineffectiveness. We report the case of a young patient with major hemophilia A with an inhibitor, who was a victim of severe thoracic trauma and for whom conservative medical treatment was successfully employed.

Observation

We report the case of a 19 years old patient with severe hemophilia A; with frequent on-demand use of factor VIII following minor trauma. The patient fell from a height estimated at one meter landing on his right side. He presented to the emergency department with bruising of the right flank with loss of consciousness, and was hemodynamically and respiratory stable. He was immediately infused with factor VIII, and then an injury assessment was performed. The brain scan was normal, and the abdominal CT scan revealed a 15-mm right peritoneal hematoma with psoas hematoma and moderate peritoneal effusion. The patient was hospitalized in the hematology department where he continued to receive factor VIII, the hemoglobin was 5.2 g / dl

transfused with two red blood cells (CG) with a control at 7.2 g / dl and an activated partial thromboplastin time (TCA) elongated to 100 sec after receiving factor VIII. A search for inhibitor was positive. The day after his trauma, the patient presented respiratory distress for which he was admitted to intensive care. At admission, the patient was agitated, tachypneic, cyanotic, had signs of Acute respiratory failure, tachycardia at 135 beats / min, and pulse oxygen saturation (SPO2) was 60% under FIO2 at 100%. A chest X-ray performed on the patient's bed showed opacity of the right lung suggesting a pleural effusion (Figure 1), the pulmonary ultrasound showed a moderate right effusion with bilateral condensation. After stabilization with non invasive ventilation (NIV) and injection of recombinant factor VII (rFVIIa), a thoraco-abdominal CT scan was performed showing bilateral pulmonary condensation with moderate effusion and stationary images at the abdominal level (Figures 2 and 3). After discussion we decided not to drain this patient and opt for non invasive treatment. The progress under NIV, rFVIIa factor, corticotherapy, diuretics was satisfactory with regression of the effusion, radiography (Figure 4) and control pulmonary ultrasound, which became minimal at the fourth day with aeration of the two lung fields. Patient transferred after 7 days of hospitalization.

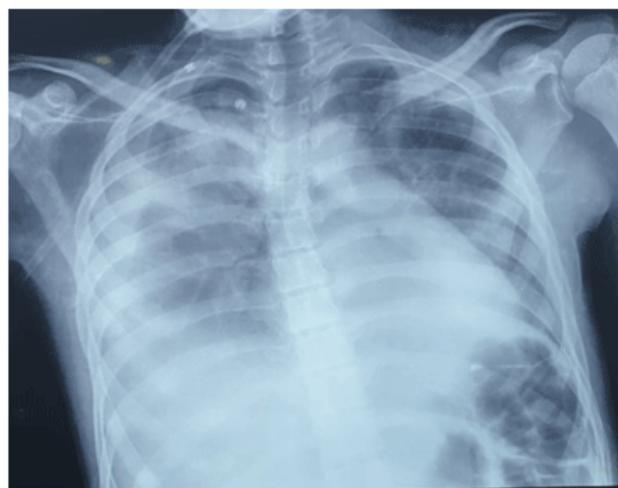


Figure 1: Chest x-ray shows right pleural effusion

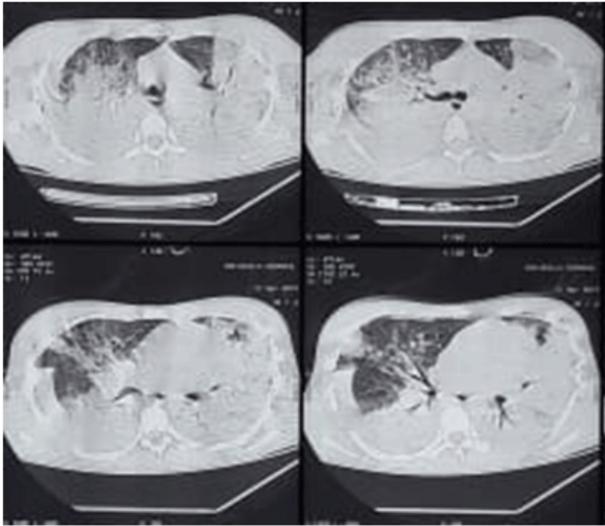


Figure 2: Thoracic CT scan showing bilateral pleural effusion with pulmonary condensation

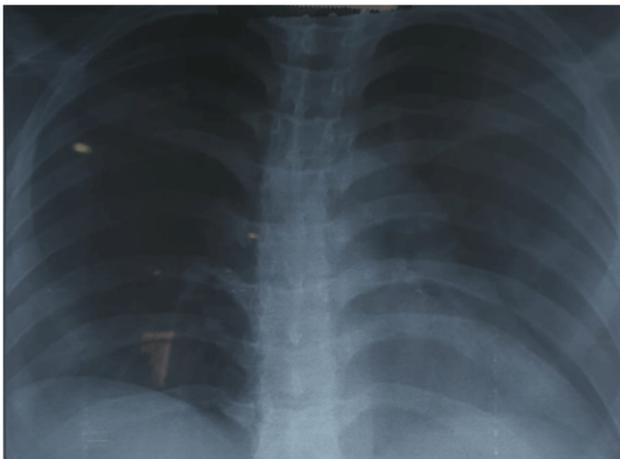


Figure 4: Chest x-ray showing good outcome without drainage

Discussion

In thoracic traumas, thoracic drainage is indicated when faced with any fluid or air effusion responsible for a respiratory and / or hemodynamic (G1 +) failure, as well as in the face of any haemothorax evaluated at more than 500 ml on the ultrasound and / or the CT Scan (G2 +) [2]. However, no recommendation is made when dealing with patients with haemophilia whereas this area is considered as a criterion for severity of thoracic trauma [2]. For any hemophilic patient, the principle is to administer factor VIII immediately after admission to the emergency department in order to restore normal hemostasis before considering other procedures. However, frequent infusion of factor risks the development an inhibitor which makes the substitution factor ineffective and requires the use of another treatment [3]. At first we should ensure an effective treatment of bleeding syndrome [4], and then ensure the suppression of the immune response.

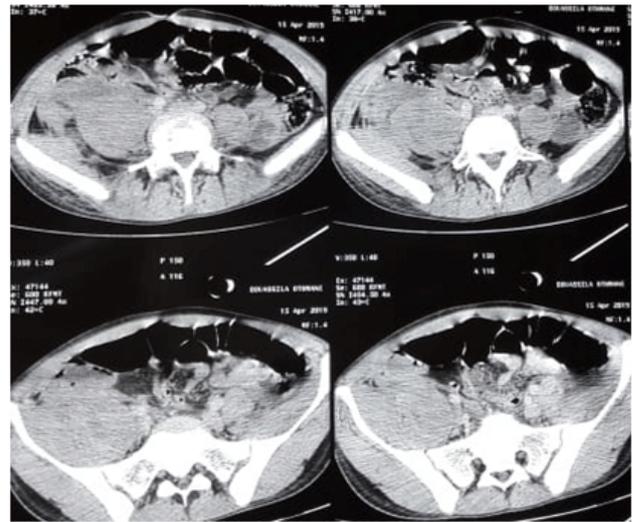


Figure 3: Abdominal CT scan showing right peritoneal hematoma with psoas hematoma

Screening for anti-VIII antibody should be done in any hemophilic patient who has been taking on-demand factor therapy at times and at frequencies that depend on the severity of the hemophilia.

For treatment of hemorrhagic syndrome [3-5], it depends on the inhibitor titer: Low responders, defined by an antibody titer of less than 5 Bethesda units, can be treated with large amounts of factor VIII at a single dose. whereas strong responders: a titer of antibodies greater than 5 Bethesda units are treated by agents bypassing the inhibitor, two molecules can be used which are: the recombinant activated factor VII (rFVIIa at a dose of 90-120 µg / kg every 2-4 hours) and plasma prothrombin-activated factor concentrate (FEIBA® dose of 50-100 U / kg every 8-12 hours). Regarding the immunomodulatory treatment, it must be as early as possible in order to reduce the time of presence of the inhibitor and therefore the duration of exposure to bleeding risk. It combines oral corticosteroids (1 mg / kg / day) with oral cyclophosphamide (1 to 2 mg / kg / day) [1].

In our case, the patient had received factor VIII several times and has a major deficiency, which implies regular screening of inhibitor in order to avoid unjustified intake of factor VIII, especially in the context of the emergency. In our patient, although thoracic drainage was indicated according to the recommendations, we opted for a wait-and-see attitude with close ultrasound monitoring in view of the high risk of bleeding and especially to the clinical improvement under symptomatic and haemostatic treatment with rFVIIa.

The favorable outcome we describe gives food for thought to the indications of drainage in this kind of patients.

Conclusion

For successful outcome, it seems necessary to make joint recommendations between hematologists and resuscitators as well as thoracic surgeons taking into account its particular situations.

Thanks

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