



## LETTER TO EDITOR

# Massive Spontaneous Hemothorax in a Child with Hemophilia

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## Dear Editor

The most common symptoms of congenital hemorrhagic disorders, mainly hemophilia and von Willebrand's disease are bleeding into the joints and muscles (1). However, epistaxis and bleeding following dental extractions, hematuria, gastrointestinal and subcutaneous hemorrhages could also be observed in hemophilia. Bleeding into the lungs have rarely been reported, but spontaneous bleeding into the pleural space in hemophilia has been quite rare and an unusual complication (1, 2).

Haemothorax is a clinical condition that can be caused by trauma, coagulopathy, or iatrogenic causes. Spontaneous hemothorax is a subcategory of hemothorax that involves the accumulation of blood within the pleural space in the absence of trauma or other causes. Hemothorax is a very rare event in hemophilia occurring in less than 1% of cases (3).

We present a 12-year-old boy with severe hemophilia A and high titer inhibitor who developed spontaneous hemothorax without history of any trauma. He was referred to our department due to an episode of sudden-onset severe chest pain. On physical examination, respiratory distress was noticeable. CT scan of the chest revealed massive right-sided pleural effusion which on aspiration; after receiving FEIBA and rFVIIa, yielded large amounts of the blood. The patient was diagnosed with hemothorax. Due to lack of improvement in general condition of the patient, replacement therapy with sequential administration of FEIBA and rFVIIa along with blood transfusion was continued for the patient.

The patient underwent tube thoracostomy which did not alleviate the symptoms of the patient, so open thoracotomy was performed to remove extravasated blood and decompress the right lung. Unfortunately, all attempts were unsuccessful and the patient died.

Rasaretnam and colleagues report a 27-year-old man in whom the diagnosis of hemophilia was established when he developed spontaneous hemothorax (4). He reported five previous cases of hemothorax developing in patients with hemophilia (2, 5-8). A 56-year-old man diagnosed with hemophilia A was also reported who was admitted to the emergency room with complaints of dyspnea. CT scan of the chest showed bilateral pleural effusion which was more prominent in the right lung (9). There was another report of a 39-year-old man with hemophilia who presented to the emergency department with acute spontaneous left-sided hemothorax. He underwent a successful thoracotomy and decortication (10). In the review of the literature, hemothorax was also reported in a 64-year-old man in whom bloody parapneumonic effusion was the first symptom of hemophilia B. In this patient, hemothorax was a complication of pneumonia and pleural inflammation (11).

Besides the rarity of this condition, there is no guideline in the pediatric literature regarding the optimal management of hemothorax in hemophiliac children. The management strategies for thoracic surgeries are not well addressed in the World Federation of Hemophilia guidelines, making the management of these patients more problematic. Conservative management versus

treatment with recombinant factor VIIa and/or activated prothrombin complex concentrate (FEIBA) are the recommended options for management of hemophiliac bleedings (12). A 4-year-old boy with hemothorax is reported who was successfully managed with multiple doses of rFVIIa to induce hemostasis, without any evidence of further systemic activation of hemostasis (13). However, the price of such a treatment should be seriously taken into account.

We report this case due to its rarity and emphasize that early recognition and immediate appropriate management whether conservative or with specific factor replacement therapy should be considered for such patients. In addition, spontaneous hemothorax in hemophilia should be kept in mind in patients who present with respiratory symptoms.

**Conflict of Interest:** None declared.

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