



## A Case of Hemophilia A Associated with Spontaneous Hemorrhagic Pleural Effusion and Intracranial Hematoma

### Spontan Hemorajik Plevral Mayi ve İntrakranial Hematom ile Seyreden Hemofili A Olgusu

Spontan Hemorajik Plevral Mayi / Spontaneous Hemorrhagic Pleural Effusion

Nuri Tutar<sup>1</sup>, Asiye Kanbay<sup>1</sup>, Hakan Buyukoglan<sup>1</sup>, Gokhan Buyukbayram<sup>1</sup>, Fatih Kurnaz<sup>2</sup>

<sup>1</sup>Department of Pulmonary Medicine, <sup>2</sup>Department of Hematology, Faculty of Medicine, Erciyes University, Kayseri, Türkiye

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#### Özet

Hemofili A cinsiyete bağlı, resesif, sıklıkla erkeklerde görülen ve faktör VIII eksikliğine bağlı oluşan bir koagülasyon bozukluğudur. Spontan hemorajiler ile karakterize nadir bir hastalıktır. Hem çocuk hem de erişkin hemofili hastalarında plevral aralığa kanama son derece nadir görülür. Biz burada 56 yaşında hemorajik plevral efüzyon ve intrakranial hematoma ile seyreden bir hemofili A olgusunu sunduk.

#### Anahtar Kelimeler

Hemofili A; Hemorajik Plevral Efüzyon; Kanama

#### Abstract

Hemophilia A is a sex-linked recessive coagulation disorder almost exclusively occurring in male subjects and caused by a deficiency of factor VIII. It is a rare disorder characterized by spontaneous hemorrhages. Spontaneous bleeding in the pleural space is very rare in hemophilia both in children and adults. Here in, we present the case of a 56-year-old hemophilia A patient with hemorrhagic pleural effusion and intracranial hematoma.

#### Keywords

Hemophilia A; Hemorrhagic Pleural Effusion; Bleeding

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Corresponding Author: Nuri Tutar, Department of Pulmonary Medicine, Erciyes University School of Medicine, Kayseri, Turkey.

T.:+903524374937 E-Mail: drnurututar@gmail.com

## Introduction

Hemophilia A is a sex-linked recessive coagulation disorder almost exclusively occurring in male subjects and caused by a deficiency of factor VIII. The common manifestations of hemophilia are prolonged and excessive bleeding after minor trauma, spontaneous hemorrhage into the tissues and joints, epistaxis, gastrointestinal hemorrhage, and hematuria [1]. Spontaneous bleeding in the pleural space is very rare in hemophilia both in children and adults [1,2]. Here in, we report a rare case of spontaneous hemorrhagic pleural effusion and intracranial hematoma in a patient with hemophilia A.

## Case Report

A 56-year-old male patient, who was diagnosed with hemophilia A nearly 3 years ago, was admitted to the emergency room with complaints of dyspnea. In his medical history he had twice experienced gastrointestinal tract and multiple intra-articular bleeding. Two months previously he had been hospitalized for gastrointestinal bleeding and admitted to a medical intensive care unit. The patient received factor 8 and erythrocyte suspension and was moved to the intensive care unit after 5 days. At emergency room admission his white blood cell count (WBC) was  $11.5 \times 10^3/\mu\text{L}$ , hemoglobin: 11.1 mmol/L, platelet:  $200 \times 10^3/\mu\text{L}$ , partial thromboplastin time (PTT): 83 seconds, prothrombin time (PT): 20 seconds and international normalized ratio (INR): 1.03. The patient had no history of trauma. A computerized tomographic (CT) scan of the chest showed bilateral pleural effusion which was more pronounced in the right lung (Figure 1). Abdominal ultrasound showed free fluid between



Figure 1. Computerized tomographic (CT) scan of the chest showing bilateral pleural effusion which was more pronounced on the right.

the intestinal loops. After arterial blood gas analysis showed respiratory acidosis (pH: 7.32, pCO<sub>2</sub>: 71 mmHg, pO<sub>2</sub>: 61 mmHg, HCO<sub>3</sub>: 39 mEq/L, SaO<sub>2</sub>: 91%) the patient was admitted to the intensive care unit. Noninvasive ventilation was started to use with bilevel positive airway pressure (BiPAP) with IPAP: 12 cmH<sub>2</sub>O, EPAP: 5 cmH<sub>2</sub>O, breath rate: 16/min. The patient was treated with Factor VIII and thoracentesis was performed. His pleural fluid hematocrit level was 7.4%. 1600 cc hemorrhagic effusion was drained from the patient. Respiratory acidosis was improved and the use of BiPAP was decreased. On the tenth day of treatment the patient became confused. Cranial CT showed

hyperdensity consistent with hematoma in the left frontal lobe (Figure 2). Surgery was risky for this patient. The inhibitor count was 11 BU and prothrombin complex concentrates (FEIBA) (75 IU/kg, twice a day) were administered. Although given the best care support, the patient died the same day.

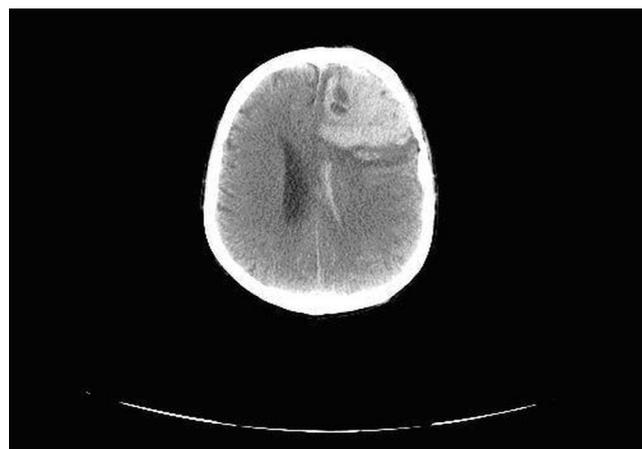


Figure 2. Cranial CT showing hyperdensity consistent with hematoma in the left frontal lobe, the left lateral ventricle is completely obliterated with hematoma.

## Discussion

The most common symptoms of congenital hemorrhagic diatheses, especially hemophilia, are bleeds into joints and intramuscular haemorrhages. Spontaneous hemorrhagic pleural effusion is an unusual clinical state, which is rare in hemophilia A patients. The present case illustrates the significance of complications for hemophilia A. There are two main problems in the management of these patients: first, the treatment of hemothorax requires the evacuation of blood from the pleural cavity to obtain complete expansion of the lung and to prevent the complications of fibrothorax, and, second, the management of hemophilia. Rasaretnam et al. drained pleural effusion in a mild hemophiliac patient [1]. On the other hand, Williams et al. used only factor concentrates to treat hemothorax and hemo-mediastinum in three hemophiliac patients [3]. We drained the pleural effusion because arterial blood gas analysis showed respiratory acidosis and better expansion of the lung might help in treatment.

Inhibitors that neutralize the hemostatic effect of factor VIII develop in 5-15% of patients on replacement therapy [4]. The incidence and prevalence of inhibitors are substantially higher in patients with severe hemophilia [4]. Furthermore, the current evidence of clinical effectiveness in the treatment of acute bleeding in hemophilia A with inhibitors is clinically and methodologically heterogeneous. FEIBA is an effective and safety agent in bleeding hemophilia patients with inhibitors. Negrier et al. performed a multicenter retrospective study to evaluate the use of FEIBA, and they presented data on 433 bleeding episodes, including surgical procedures, concerning 60 patients from 15 hemophilia centers [5]. The efficacy was judged as good or excellent in 352 episodes (81.3%), poor in 73 episodes (16.9%) and non-existent in 8 episodes (1.8%). Initially we treated our patient with factor VIII, drained the pleural effusion and used noninvasive ventilation. After intracranial hematoma occurred, we started FEIBA treatment but this was not enough to save the patient.

In conclusion, if unusual bleeding develops in patients with hemophilia A, these cases should be followed closely for life-threatening bleeding. At the same time, in order to prevent life-threatening bleeding, fast and intensive supportive therapy should be given.

#### **Competing interests**

The authors declare that they have no competing interests.

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