A Case of Acute Upper Airway Obstruction in a Pediatric Hemophilia A Patient Because of Spontaneous Retropharyngeal Hemorrhage

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We report a rare case of acute upper airway obstruction caused by spontaneous retropharyngeal hemorrhage as a result of hemophilia A in a 16-year-old pediatric patient who routinely received factor VIII replacement. Initial diagnosis was delayed because the patient presented with symptoms, such as throat pain and odynophagia, similar to those of common benign upper airway infections. Within 2 days of the initial presentation of symptoms, the patient went into respiratory failure as a result of retropharyngeal hemorrhage. The possibility of spontaneous retropharyngeal or epiglottic hemorrhage or hematoma should be considered as a cause of rapidly progressing odynophagia and dyspnea by hemophilia patients. [Ann Emerg Med. 2016;67:616-619.]

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INTRODUCTION

Acute upper airway obstruction is an urgent life-threatening event requiring immediate diagnosis and appropriate intervention. Hemophilia causes coagulopathy because of insufficient coagulation factors (factor VIII or IX), and patients present with increased bleeding times, as well as spontaneous hemarthrosis in large joints such as the knee.[1-3]

We describe a case involving a 16-year-old pediatric patient with hemophilia A who initially presented with throat pain. Within 2 days, the patient showed odynophagia and dyspnea, which progressed to acute upper airway obstruction as a result of spontaneous retropharyngeal hemorrhage.

CASE REPORT

The patient was a 16-year-old 70-kg adolescent boy who routinely received 4,000 units of factor VIII replacement therapy 3 times a week at another hospital to manage hereditary hemophilia A. He initially presented to his pediatrician within 1 day of the onset of throat pain. The pediatrician prescribed a course of oral amoxicillin to treat suspected lymphadenitis or other bacterial upper airway infection. The patient sought treatment at our pediatric hospital at approximately midnight of the same day because of increased throat pain, new odynophagia, and dyspnea. The lung fields were clear to auscultation bilaterally and his vital signs were stable and within normal limits. The patient was unable to completely open his mouth because of increased neck swelling, pain, and pressure. Inhospital observation was recommended by the pediatrician. However, the patient and his mother declined to follow this recommendation and left the hospital against medical advice.

Unfortunately, the patient was transported back to our pediatric hospital emergency department a few hours later by ambulance because of increased throat pain and labored breathing. Again, the patient was lucid, with clear bilateral lung sounds and normal vital signs, including an SpO_2 of 99% on room air. The patient was admitted to the general pediatric ward for observation and testing to determine the cause of worsening symptoms. The blood tests showed the following results: hemoglobin 14.9 g/dL, WBC count 10.2 × 10^3 / μL, thrombocytes 221 × 10^3 / μL. Studies of C-reactive protein, factor VIII level, and other coagulation characteristics such as prothrombin time, prothrombin time:international normalized ratio, and activated partial thromboplastin time were not conducted at this time. Examination showed that the oropharynx was not injured. Intravenous administration of cefuroxime was initiated to treat suspected lymphadenitis or other upper airway infection. Testing for infectious diseases such as Epstein-Barr virus, adenovirus, and coxsackievirus was conducted later, with negative results.

An epinephrine nebulizer treatment was ordered to treat symptoms of dyspnea. Supplemental oxygen by nasal cannula was ordered, but the patient refused treatment. SpO_2 remained 100% on room air.

Ultrasonography of the patient's neck and a chest radiograph were requested; however, the patient developed

severe inspiratory stridor and agitation in the radiology department. Therefore, he was sent back to the ward and began receiving treatment with a rebreathing mask, with oxygen flow at 3 L/minute. This treatment helped the patient maintain SpO_2 at 100% and his agitation resolved. The patient's lungs remained clear to auscultation bilaterally. He was again sent to the radiology department for an ultrasonographic study of the neck and a chest radiograph. After the chest radiograph was performed (Figure 1), before ultrasonographic study of the neck, the patient again became dyspneic and rapidly progressed to respiratory failure. Seizures and urinary incontinence were observed.

A hospital emergency code was called. Support staff, including the on-call anesthesiologists, arrived and supported the patient's spontaneous respiration with a bag-valve-mask device. An attempt was made to intubate the patient; however, the anesthesiologists could not obtain a view of the vocal cords because of severe oropharyngeal swelling. Bag-valve-mask–assisted ventilation became increasingly difficult; therefore, a laryngeal mask airway was inserted as an emergency measure and successfully used for ventilation. The patient was then transported to the operating room to attempt to secure the airway by orotracheal intubation in a more controlled environment with other anesthesiologists, anesthesia nurses, several instruments for difficult airways such as an emergency percutaneous cricothyroidotomy set, and so on. In addition, surgeons operating in other operating rooms were also available to assist with a surgical airway if necessary.

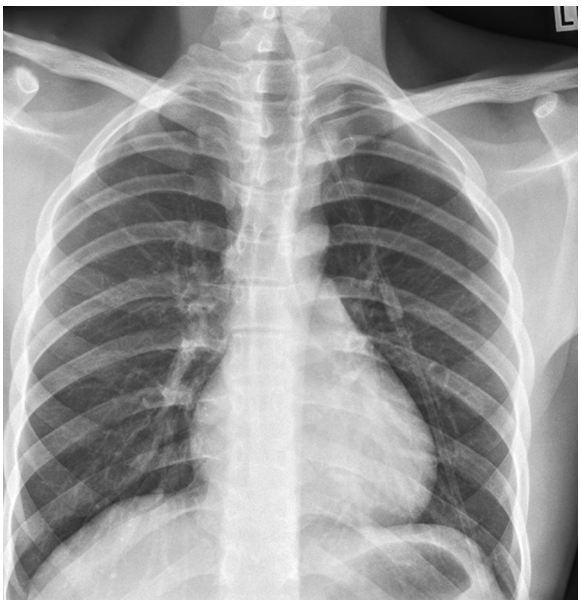

Figure 1. The chest radiograph performed directly before the onset of respiratory failure and concomitant seizures. The upper part of the trachea has significant stenosis, with only a 3-mm-diameter opening, whereas both lung fields show no abnormality.

The laryngeal mask airway was removed and bronchoscopic oral intubation with insufflation of 100% oxygen was attempted. However, this failed because of excessive oropharyngeal swelling. Bag-valve-mask ventilation was again attempted but was unsuccessful. A laryngeal mask airway was inserted again, but this attempt was also unsuccessful. The patient was now considered to have "cannot intubate, cannot ventilate" status and rapidly progressed to bradycardia and eventual cardiac arrest as a result of frank respiratory failure.

Cardiopulmonary resuscitation (CPR) was initiated immediately and the patient received intravenous administration of 1 mg epinephrine and a second intravenous 1-mg epinephrine bolus after approximately 3 minutes. CPR was continued while an emergency percutaneous cricothyroidotomy was performed with the QuickTrach II cannula for adult patients (4.0 mm inside diameter) (VBM Medizintechnik GmbH, Sulz am Neckar, Germany), and this resulted in return of spontaneous circulation. The total duration of CPR was approximately 5 minutes. Ventilation was continued through the QuickTrach II cannula until the airway was secured through transnasal bronchoscopic intubation with a 7.0-mm normal endotracheal tube. The cricothyroidotomy cannula was removed and the patient was transported to the ICU. In all, approximately 40 minutes passed from the onset of respiratory failure and concomitant seizures until a secure airway with an endotracheal tube was established. The chest radiograph could not be checked until this time because the hospital emergency code was called directly after the chest radiograph was performed, and therefore the anesthesiologists were engaged in emergency measures for the patient.

In the ICU, the patient continued to receive positive-pressure ventilation and was sedated with continuous intravenous midazolam and fentanyl. Bilateral rales were noted and assumed to be associated with pulmonary hemorrhage as a result of vigorous CPR or edema as a sequela of negative inspiratory pressure.[4,5] To maintain adequate ventilation, high positive end-expiratory pressure of 12 mbar was used.[6,7]

On admission to the ICU, the blood tests and coagulation studies from samples obtained while CPR was performed showed the following results: hemoglobin 10.7 g/dL, WBC count 20.5 × 10^3 / μL, thrombocytes 228 × 10^3 / μL, C-reactive protein 18.0 mg/L, factor VIII 62%, prothrombin time 60%, prothrombin time:international normalized ratio 1.30, and activated partial thromboplastin time 43.1 seconds. The significant decrease of hemoglobin level from 14.9 g/dL to 10.7 g/dL in such a short time