CASE REPORT

Recurrent spontaneous massive hemothorax from intrathoracic extramedullary hematopoiesis resulting in respiratory failure

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Extramedullary hematopoiesis (EMH) is a compensatory response to many chronic anemic disorders. Intrathoracic EMH, usually presenting as paravertebral masses over the posterior mediastinum, is a rare entity and is usually asymptomatic. Hemothorax is a rare but possibly fatal complication. Local radiation for intrathoracic EMH is considered effective in preventing its recurrence. Here we describe a patient who had had α-thalassemia for many years and developed a spontaneous left-sided hemothorax from EMH. A chest film and a chest computed tomography (CT) scan had showed multiple paravertebral masses over the lower thoracic spine with left-sided pleural effusion. A pathological diagnosis of EMH was made by video-assisted thoracoscopic surgery. The patient had not received preventive local chest radiation. Ten years later, he suffered from a life-threatening hemothorax complicated by acute respiratory failure without traumatic history. A CT scan showed posterior mediastinal masses over the lower thoracic spine with right-sided pleural effusion. Thoracoscopy was performed to remove the blood clot in the pleural space for successful weaning from mechanical ventilation. This is the first case of intrathoracic EMH to have recurrent hemothorax associated with acute respiratory failure.

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Introduction

Extramedullary hematopoiesis (EMH) occurs as a compensatory phenomenon to several hematological diseases including thalassemia, myelofibrosis, and hereditary spherocytosis. Intrathoracic EMH is a rare clinical disorder that is often located in the lower thoracic paraspinal area and is usually asymptomatic. Non-traumatic spontaneous hemothorax is a rare complication of EMH. No cases of intrathoracic EMH with recurrent spontaneous hemothorax have been reported. We describe a thalassemic patient who developed spontaneous left-sided hemothorax and then 10 years later a contralateral hemothorax complicated by acute respiratory failure.

Case report

A 55-year-old man presented with a history of \(x\)-thalassemia for over 20 years. This disorder was diagnosed by positive hemoglobin H staining, and cytology of a bone marrow aspirate revealed erythroid hyperplasia. He had been admitted previously for dyspnea and left-sided chest pain without traumatic history. A complete blood count showed microcytic anemia without thrombocytopenia, and a physical examination showed hepatosplenomegaly. A chest computed tomography (CT) scan showed bilateral posterior mediastinal masses with left-sided pleural effusion (Fig. 1A). Thoracocentesis revealed bloody pleural effusion and negative cytology findings were obtained for the effusion. After admission, a chest tube was placed and video-assisted thoracoscopic surgery was then arranged for pathological diagnosis of the posterior mediastinal masses and correction of hemothorax. Pathologic analysis of the left mediastinal masses revealed a diagnosis of EMH with normal-maturation hematopoietic tissues of three lineage cells. A diagnosis of intrathoracic EMH was then made. No preventive local radiation therapy was given at that time at the patient’s request. He received blood transfusion therapy as needed at a local hospital every 2–3 months.

Ten years later, he suffered from sudden onset of right chest pain and dyspnea without any history of trauma. At the emergency department, a chest film showed right-sided pleural effusion, and a chest CT scan revealed right-sided massive pleural effusion and lobulated paravertebral masses over the thoracic spine with good contrast enhancement (Fig. 1B). A physical examination revealed normal vital signs, but pale conjunctiva, diminished right-side breathing sounds, and marked hepatosplenomegaly. A hematological examination revealed hemoglobin level of 10.3 g/dL and platelet count of \(160 \times 10^3\)/mm\(^3\). Thoracocentesis revealed bloody effusion with a red blood cell count of \(5,1 \times 10^6\)/mm\(^3\). Mechanical ventilation was applied because of progressively worsening dyspnea. Thoracoscopy was performed to remove an intrapleural blood clot for better lung expansion. Local low-dose radiation therapy of the paraspinal masses was then arranged after the patient had been successfully weaned from the ventilator.

Discussion

EMH occurs as a compensatory response in various chronic anemia conditions including thalassemia, sickle cell anemia, and myelofibrosis. It usually develops in blood-forming organs outside the bone marrow, such as the liver, spleen and lymph nodes, and also develops rarely as a mass-like lesion within the thorax. Pale conjunctiva and hepatosplenomegaly are common clinical findings. Intrathoracic EMH usually develops in the posterior mediastinum as multiple large lobulated tumors with good contrast enhancement. Although intrathoracic EMH is commonly asymptomatic, it can occasionally present with symptoms of spinal cord compression\(^1\) and traumatic hemothorax.\(^2\)

Very few patients with intrathoracic EMH complicated by spontaneous hemothorax have been reported. Smith et al reported the first patient with thalassemia who developed intrathoracic EMH and hemothorax and was treated with local radiation therapy to prevent recurrent bleeding.\(^3\) Chute et al reported the first fatal hemothorax from spontaneous rupture of intrathoracic EMH in 2004.\(^4\) Kupferschmid et al reported a case of myelofibrosis with intrathoracic EMH.\(^5\) This complicated massive hemothorax did not respond to tetracycline pleurodesis and was ultimately controlled.
with low-dose radiation therapy. Bartlett et al described a case of agnogenic myeloid metaplasia with hemothorax. The intrathoracic EMH, confirmed by a technetium-99 bone marrow scan, was treated successfully with low-dose radiation to prevent recurrent hemothorax. An eighth case of spontaneous left-sided hemorthorax due to EMH was reported by Pornsuriyasak et al in 2006. Our patient had previously had a spontaneous left-sided hemorthorax due to intrathoracic EMH, and he suffered from spontaneous recurrent hemorthorax due to EMH on the contralateral side 10 years later. A clinical diagnosis of intrathoracic EMH was made according to his previous history of \(\alpha\)-thalassemia and typical chest CT findings. Thoracoscopic decortication was performed for tissue diagnosis of the posterior mediastinal mass in the first episode, and for discontinuation of mechanical ventilation with better lung expansion during the second episode. To the best of our knowledge, this is the first reported case of recurrent massive hemothorax caused by EMH.

Various non-invasive diagnostic procedures, such as contrast-enhanced CT, magnetic resonance imaging of the thorax, and technetium-99 sulfur colloid radionuclide bone marrow scanning, have been used to diagnose EMH, while fine needle aspiration may carry a risk of hemorrhagic complications. In the presence of an underlying chronic anemic condition, the finding of lower paravertebral homogenous smooth masses on CT, a typical finding in intrathoracic EMH, is typically used to establish a definitive diagnosis of intrathoracic EMH.

It is usually unnecessary to treat asymptomatic intrathoracic EMH. Low-dose radiation has been used as an effective method to control symptomatic spinal cord compression and hemothorax in EMH. Our patient had refused local chest radiation 10 years previously, but agreed to receive thoracic radiation therapy after the second hemorthorax to prevent future life-threatening bleeding.

In conclusion, it is important to recognize the possibility of intrathoracic EMH in patients with chronic anemic disorders. Hemorthorax may develop spontaneously and may result in severe outcome. In our patient, no preventive local radiation may have been the most important reason for recurrence of hemorthorax. Radiation therapy, even including the contralateral side, may be one of the most important therapies to prevent recurrent life-threatening hemorthorax.

References