Case Report

Massive Hemothorax Due to Intrathoracic Extramedullary Hematopoiesis in a Patient with Beta Thalassemia Hemoglobin E Disease

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Case report of a 28-year-old man with beta thalassemia/hemoglobin E disease who developed a massive hemothorax due to bleeding from multiple large intrathoracic, paraspinal hematopoietic masses. Initially, a thoracotomy was required for the control of bleeding. Postoperatively, the patient received 2,000 ml of packed red cells but he continued bleeding after the blood chemistries and coagulopathy specimen were corrected. The decision was made to return to the operating room for a thoracotomy to control the bleeding. However, before re-operating, the patient underwent a collapse and failed resuscitation. This complication has never been previously reported in Thailand.

Keywords: *Massive hemothorax, Extramedullary hematopoiesis, Thalassemia*

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Massive hemothorax due to intrathoracic extramedullary hematopoiesis is an unusual phenomenon, first reported in 1988 by Smith et al[1]. This condition is most frequently observed as a response to the intense intramedullary hemolysis associated with the homoxygous beta-thalassemias and related conditions such as hemoglobin E-thalassemia. In general, extramedullary hematopoietic masses are usually asymptomatic, although spinal cord compression has been reported[2]. Spontaneous bleeding from extramedullary hematopoiesis is very rare. This paper describes a patient with beta-thalassemia with a large erythroid mass resulting in a massive hemothorax.

Case Report

The patient was a 28-year-old man with beta-thalassemia (hemoglobin E) with a history of a spleenectomy at eight years of age. His hemoglobin baseline was 7.5-8 g/dl and received a transfusion of two units of packed red cells monthly. His history of spinal cord compression at the lower thoracic level occurred on June 1, 2011 with symptoms of paraparesis, motor power grade three. After treatment with dexamethasone intravenously and radiotherapy 1,200 cGy, his paraplegia improved with a return to normal motor power one month after treatment. The patient had long-standing hepatomegaly with mild abnormal liver function test but remained actively employed and clinically stable. The intrathoracic extramedullary hematopoiesis was not identified prior to this admission.

The evening of the day before admission, upon his return home, the patient felt chest discomfort and developed right pleuritic chest pain, and increasing dyspnea. His parents brought him to a local hospital. At the emergency room of the local hospital the blood pressure (BP) was 100/70 mmHg; pulse rate, 100 beat/min and regular; respiratory rate, 28/min; and temperature 37.2°C orally. Physical examination results showed signs of a massive right pleural effusion, which a chest roentgenogram showed to extend almost to the lung apex as shown in Fig. 1A. Thoracocentesis yielded gross blood with a hematocrit (Hct) equal to the venous hematocrit. After 100 ml of hemothorax was relieved he developed cardiac arrest and cardiopulmonary resuscitation (CPR) was initiated. About five minutes after starting CPR his heart rate was 120 beat/min and regular, BP was 90/60 mmHg, oxygen saturation was 95% under endotracheal tube and positive pressure ventilation. He was transferred to the provincial hospital.

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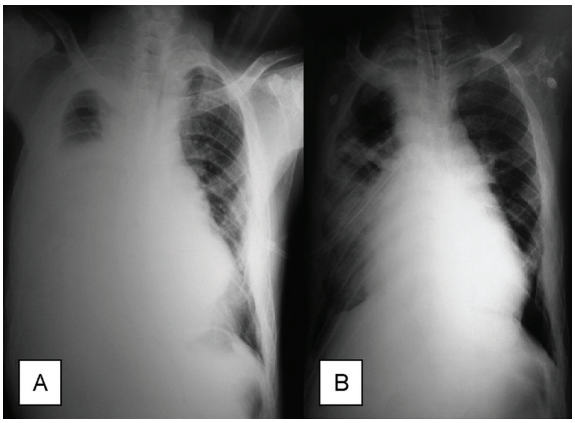

Fig. 1 (A) Chest roentgenogram showing massive right pleural effusion. (B) Chest roentgenogram post insertion of an intercostal drainage tube at the provincial hospital.

At the provincial hospital, he remained unconscious with stable vital signs. The emergency department physician inserted an intercostal drain tube obtaining 3,000 ml of bloody fluid within 30 minutes. The chest roentgenogram after releasing the hemothorax was shown in Fig. 1B. The patient received four units of packed red cell then he was transferred to Chiang Mai University Hospital, a tertiary care center.

At the emergency room, the systolic BP was 80 mmHg with a PR 140 beat/min. He had severe metabolic acidosis (pH 6.45) and coagulopathy (PTT more than 200 seconds), with a hematocrit of 17 mg/dl and platelet count of 40,000 per milliliter (Laboratory results were received during the operation). He was sent to the operating room immediately. Before starting a skin incision, cardiac arrest occurred and CPR for five minutes was successful. A right posterolateral thoracotomy was performed and multiple intrathoracic extramedullary hematopoiesis along the paravertebral area was found. The capsule of the largest lesions had a five mm transverse tear with active bleeding. The edges of the laceration were apposed with prolene 4/0 with pledgetted mattress sutures (Fig. 2) and packed with four swab pieces to control the bleeding. The intra-operative blood loss was estimated to be one liter and the operative time was 30 minutes. The thoracotomy was temporarily closed and the patient was transferred to the intensive care unit (ICU). Four hours after operation, the BP was 120/75 mmHg (with dopamine administration 10 microgram/kilogram/minutes); PR 120 beat/min and temperature 37.8°C. The bleeding continued for approximately 1,500 ml per four hours. After correcting the coagulopathy achieving to normal PT, PTT, and platelet, the decision was made to operate again due to ongoing bleeding. Before induction, cardiac arrest occurred. CPR was performed for one hour with asystole and finally the patient was pronounced deceased. A re-thoracotomy was performed to remove the swab packing. The intrathoracic extramedullary hematopoiesis is shown in Fig. 2.

Discussion

Extramedullary hematopoiesis is seen in various hematologic disorders including severe hemolytic anemias such as thalassemia or conditions such as myelophthistic anemia or myeolofibrosis, where there is extensive replacement of normal bone marrow. The most common sites of extramedullary hematopoiesis are the liver, spleen, and lymph nodes. Less common sites are thymus, kidney, retorperitoneum, and paravertebral areas of the thorax[3].

Intrathoracic extramedulary hematopoiesis is an uncommon and usually asymptomatic condition. Tumor-like masses are formed that have a characteristic appearance[4]. They are usually located in the lower thoracic paravertebral areas and are usually multiple, bilateral, and vary in size. No previous report showed the destruction of adjacent ribs and vertebrae, however Luyendijk et al[2] in 1975 reported an intrathoracic mass of erythropoietic tissue with spinal cord compression like the present case. Pathologically, the masses

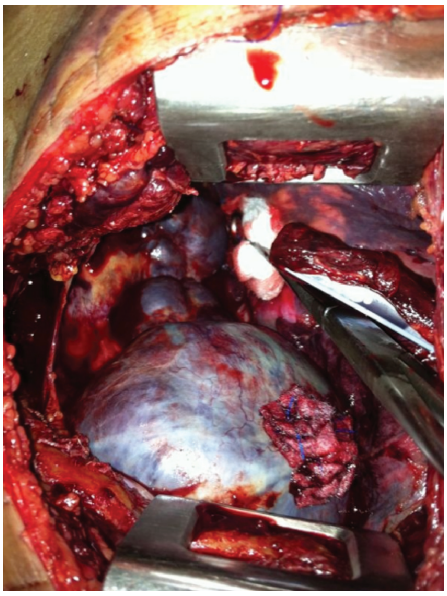

Fig. 2 Multiple intrathoracic extramedullary hematopoietic lesions with multiple prolene 4/0 with pledgetted mattress sutures for controlling bleeding point.

were soft, deep red, and resemble splenic tissue on the cut surface. Histologically, they are formed of hematopoietic elements mixed with adipose tissue[1]. This is the first reported case of hemothorax in Thailand with this condition. There are no conclusions in terms of the origin of these lesions. They may arise from the extrusion of hyperplastic marrow through the thinned cortex of ribs or vertebrae, with the periosteum forming the capsule of mass or embryonic rests may transform into hematopoietic tissue under situations of bone marrow stress[1].

The diagnosis of intrathoracic extramedulary hematopoiesis can be established with the basis of the characteristic radiologic findings in a patient who has a predisposing hematologic condition. Paravertibral masses in the posterior mediastinum especially neurogenic tumors are the main considerations in differential diagnosis[5]. Features that help to discriminate these two diseases are finding of lobulation (nearly 95 percent of cases) and the absence of bony destructions in intrathoracic extramedulary hematopoiesis. Almost fifty percent of patients with neurogenic tumors have bone erosion and sclerosis and lobulation was not seen[4]. Furthermore, enhanced visualization on post contrast computerized tomography of the masses assists the diagnosis of intrathoracic extramedulary hematopoiesis[1]. Invasive diagnostic procedures, including thoracotomy or video-assisted thoracoscopic surgery (VATS) and fine-needle aspiration (FNA)[6] are usually necessary. Moreover, they are potentially dangerous because these lesions are highly vascular in nature. In-111 chloride and Tc-99m Sn colloid bone marrow scintigraphies may be used to confirm the diagnosis[7]. The blood supply derives from intercostal arteries and the esophageal plexus. Venous drainage is into the azygos system.

Treatment is required only in the presence of complications such as spinal cord compression or hemothorax. Extramedullary hematopoietic tissue is highly radiosensitive, and relatively small doses of radiation have been effective after surgical decompression in cases of spinal cord compression[2] or after surgical control of the acute bleeding causing hemothorax[1]. The lacerations in the capsule of the largest lesion seen during the thoracotomy may have resulted from spontaneous rupture. Giblin et al[8] report that using VATS is an amendable and less-invasive means of tumor removal. In an emergency situation like this using prolene 4/0 with pledgetted mattress sutures and packing with swabs may be not enough to control the bleeding. Some experts suggest that using a pericardial patch to entirely cover the laceration may be useful, however this procedure requires time experience so is not suitable for an unstable patient.

Treatment of this potential complication of intrathoracic extramedullary hematopoiesis is difficult to handle. The best way is to utilize a short and effective procedure (either direct closure with pledgetted mattress sutures or pericardial patch). Awareness of this condition may make it possible to successfully treat future cases with pericardial patch and radiation post-operatively.

Potential conflicts of interest

None.

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ภาวะเลือดออกในช่องเยื่อหุ้มปอดรุนแรงเนื่องจากเนื้อเยื่อไขกระดูกที่เจริญมากผิดปกติบริเวณ ทรวงอกในผู้ป่วยโรคบีตาธาลัสซีเมียร่วมกับฮีโมโกลบินอี

อภิชาติ ตันตระวรศิลป์, สมเจริญ แซ่เต็ง

ผู้ป่วยชายอายุ 28 ปี ได้รับการวินิจฉัยว่าเป็นบีตาธาลัสซีเมียร่วมกับฮีโมโกลบินอี มาพบแพทย์ด้วยมีภาวะเลือดออกมากในช่องเยื่อหุ้มปอดเนื่องจากมีการแตกออกของไขกระดูกที่เจริญเติบโตผิดปกติที่กระดูกซี่โครง ผู้ป่วยได้รับการรักษาโดยการผ่าตัดฉุกเฉินเปิดทรวงอกด้านขวาในห้องผ่าตัดเพื่อหยุดเลือดที่ออก หลังผ่าตัดได้แก้ไขภาวะความเป็นกรดของเลือดและความผิดปกติของการแข็งตัวของเลือด แต่พบว่ายังคงมีเลือดออกทางท่อระบายทรวงอกอยู่ จึงได้ทำการผ่าตัดเปิดทรวงอกอีกครั้ง แต่ก่อนที่จะทำผ่าตัดเปิดทรวงอกครั้งที่สองหัวใจผู้ป่วยหยุดเต้น ได้ทำการนวดหัวใจและใช้ยากระตุ้นหัวใจ แต่สุดท้ายผู้ป่วยเสียชีวิต ภาวะเลือดออกในทรวงอกอันเนื่องมาจากการแตกออกของไขกระดูกที่เจริญเติบโตผิดปกติที่กระดูกซี่โครงพบได้น้อยมาก และยังไม่เคยมีรายงานในประเทศไทย