

Extramedullary Hematopoiesis at the Posterior Mediastinum in Patient with Hereditary Spherocytosis: A Case Report

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Thoracic extramedullary hematopoiesis (EMH) is a rare disease entity that is usually associated with hematologic disorders, such as myelodysplastic or hemolytic disease. Because thoracic EMH is usually encountered as a mass during radiologic examinations, it should be differentiated from posterior mediastinal neurogenic tumors. Here, the authors report a case of EMH associated with hereditary spherocytosis. The patient underwent a complete excision by thoracoscopic surgery to differentiate it from other mediastinal tumors.

Key words: 1. Mediastinal neoplasms
2. Hematopoiesis, extramedullary
3. Hereditary spherocytosis

CASE REPORT

A 48-year-old male presented at our clinic with a thoracic mass, which had been incidentally detected on chest computed tomography (CT), and was not clear on routine chest radiography (Fig. 1A). Chest CT revealed a 3.2-cm flat posterior mediastinal mass at the T8 level (Fig. 1B), which led to suspicions of a neurogenic tumor, such as ganglioneuroma or neurilemmoma. However, magnetic resonance images showed slightly lower signal intensity in the bone marrow of all the thoracic vertebrae and a combined finding of myelofibrosis in the bone marrow, which suggested that the posterior mediastinal mass could have been extramedullary hematopoiesis (EMH). A technetium-99m phytate bone marrow scan was performed and asymmetric increased uptake was noted at the 8th thoracic vertebra (Fig. 1C). The patient had a history of hereditary spherocytosis, and had undergone splenectomy,

cholecystectomy, and distal pancreatectomy 4 years previously. His laboratory findings were as follows: white blood cells $6.06 \times 10^3/\mu\text{L}$, hemoglobin 14.7 g/dL, platelets 171,000 μL , segmental neutrophils 27%, lymphocytes 51.7%, monocytes 15%, and eosinophils 4.6%. Chemistry analysis findings were within the normal range. Although the patient had no specific symptoms and the possible diagnosis was EMH, we decided to confirm the pathologic diagnosis to differentiate the mass from neurogenic tumors and other posterior mediastinal diseases, and to remove the potential risk of spinal cord compression because the lesion was located close to the neural foramen.

Video thoracoscopic surgery was performed and a 2-cm dark-blue pleural mass was found (Fig. 2) and excised completely without difficulty. A frozen section examination showed that the mass was consistent with EMH. A permanent histopathologic examination revealed myeloid tissues

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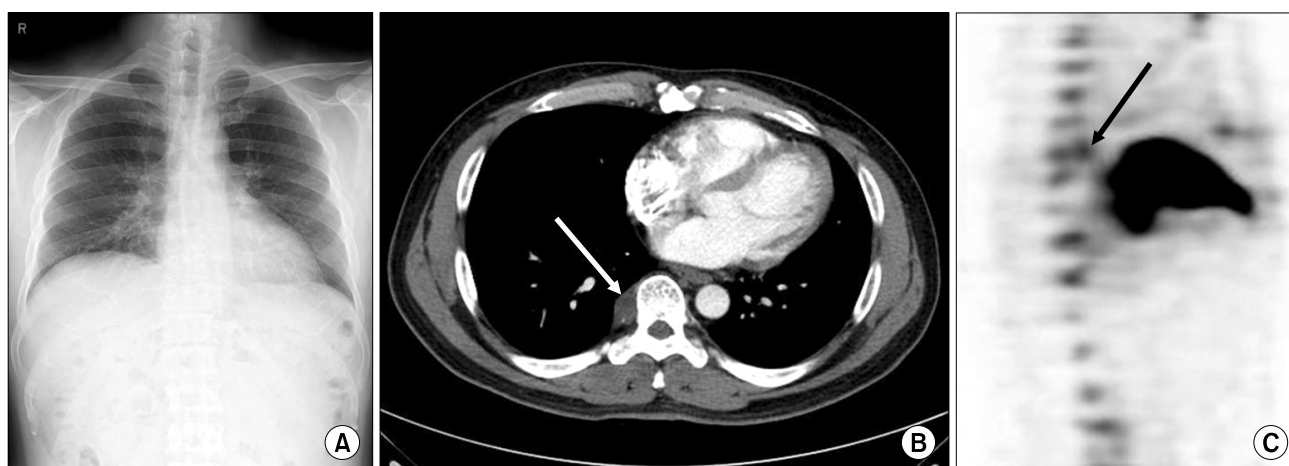


Fig. 1. (A) Chest radiography. (B) A mass-like lesion in the posterior mediastinum (arrow) and (C) strong uptake in a bone marrow scan (arrow).

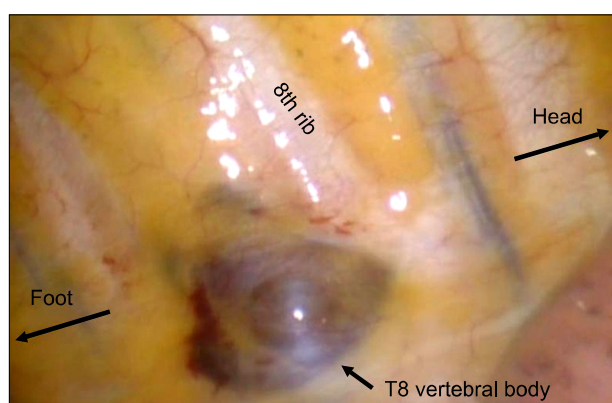


Fig. 2. Thoracoscopic findings of extramedullary hematopoiesis located on the T8 vertebra. Note an engorgement of the parietal pleura covering the extramedullary hematopoiesis.

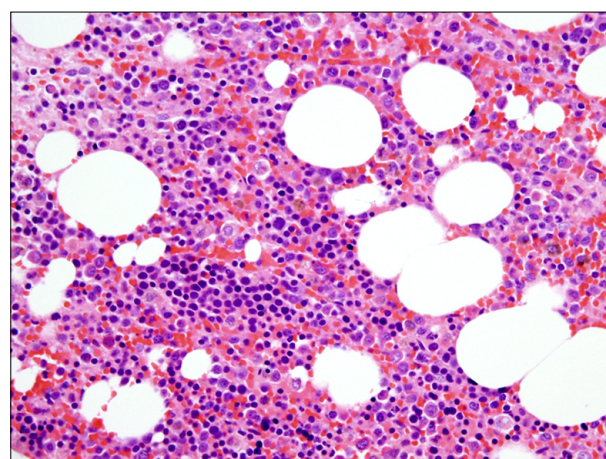


Fig. 3. The microscopic findings of the specimen (H&E, $\times 400$). Note the myeloid tissue composed of various hematopoietic cells suggesting extramedullary hematopoiesis.

composed of hematopoietic cells and confirmed a diagnosis of EMH (Fig. 3). The patient's postoperative course was uneventful and he was discharged without complications on the 5th postoperative day.

DISCUSSION

EMH is defined as production of blood components (hematopoiesis) outside the bone marrow. It has been hypothesized that EMH occurs because of 1) hematogenous spread of hematopoietic stem cells, 2) a compensatory response to a reduction in blood components during pathologic

conditions such as myelofibrosis, and 3) the activation of embryonic hematopoietic stem cells in other sites [1].

However, EMH is a rare disease and may accompany advanced myelofibrosis in patients with a myelodysplastic, hemolytic disease. In addition, chronic myelodysplastic and congenital hemolytic disorders are also risk factors of EMH [2].

During hereditary spherocytosis, an autosomal dominant genetic disorder of red blood cell membranes and cell membrane abnormalities can cause the loss of cellular architecture, and in particular, red blood cells tend to develop spherical features and may subsequently undergo lysis [3]. As a con-

sequence, patients with hereditary spherocytosis can present with symptoms of anemia, hepatosplenomegaly, jaundice, and venous thromboembolism, and may also present with EMH as a compensatory response [4].

In terms of its thoracic manifestations, EMH should be differentiated from neurogenic tumors arising in the posterior mediastinum. Its most common form is a paraspinal mass mimicking a neurogenic tumor [5]. In addition, it can present as spontaneous hemothorax, and rarely, as a lung nodule or mass, or as mediastinal lymphadenopathy or pleural seeding in the presence of a thoracic malignancy [2]. Furthermore, paraplegia can develop due to cord compression if EMH affects the spinal canal. Fine needle aspiration is an acceptable option for differential diagnosis, but it introduces the risk of post-procedural bleeding.

Surgical excision is recommended in patients with a symptomatic mass or spontaneous bleeding, or when a malignancy is suspected [2]. Surgical excision could be achieved using a minimally invasive thoroscopic procedure [6], and may be strongly recommended if a patient is symptomatic or when spinal cord compression is anticipated. After surgical excision, adjuvant radiotherapy may help prevent recurrence [7]. In our case, we decided on surgical excision because the mass was located in the proximity of the neural foramen and spinal canal invasion was anticipated. Low dose radiotherapy has also been advocated for the complete remission of EMH [8].

EMH has a relatively poor prognosis when it recurs or invades other organs. Causes of death in patients with EMH include the following: pulmonary embolism, conversion to acute leukemia, renal failure, and cerebral infarction [2]. We report a surgical case of EMH that presented as a paraspinal mass, which was treated by complete resection, in a patient

with hereditary spherocytosis.

CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

REFERENCES

1. O'Malley DP. *Benign extramedullary myeloid proliferations*. Mod Pathol 2007;20:405-15.
2. Koch CA, Li CY, Mesa RA, Tefferi A. *Nonhepatosplenic extramedullary hematopoiesis: associated diseases, pathology, clinical course, and treatment*. Mayo Clin Proc 2003; 78:1223-33.
3. Hassoun H, Palek J. *Hereditary spherocytosis: a review of the clinical and molecular aspects of the disease*. Blood Rev 1996;10:129-47.
4. Smith J, Rahilly M, Davidson K. *Extramedullary haematopoiesis secondary to hereditary spherocytosis*. Br J Haematol 2011;154:543.
5. Xiros N, Economopoulos T, Papageorgiou E, Mantzios G, Raptis S. *Massive hemothorax due to intrathoracic extramedullary hematopoiesis in a patient with hereditary spherocytosis*. Ann Hematol 2001;80:38-40.
6. Chang JW, Maeng YH. *VATS resection for a posterior mediastinal extramedullary hematopoietic mass: resection of extramedullary hematopoiesis*. Korean J Thorac Cardiovasc Surg 2010;43:542-5.
7. Dore F, Pardini S, Gaviano E, et al. *Recurrence of spinal cord compression from extramedullary hematopoiesis in thalassemia intermedia treated with low doses of radiotherapy*. Am J Hematol 1993;44:148.
8. Neben-Wittich MA, Brown PD, Tefferi A. *Successful treatment of severe extremity pain in myelofibrosis with low-dose single-fraction radiation therapy*. Am J Hematol 2010;85: 808-10.