A Case Report: Thoracic Extramedullary Hematopoiesis Found by Occurring Spontaneous Pneumothorax

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Extramedullary hematopoiesis (EMH) is a rare disease associated with hematologic disorders. This report describes a case of posterior mediastinal mass found by occurring spontaneous pneumothorax in a 48-year-old male. The intrathoracic mass resected using video-assisted thoracic surgery (VATS) was diagnosed thoracic EMH (TEMH). No disorders were found by hematologic exams before or after surgery. This report suggests that such lesions must therefore be considered in the differential diagnosis of posterior mediastinal tumors presenting with no hematologic disorders. (Ann Thorac Cardiovasc Surg 2008; 14: 382–385)

Key words: mediastinal tumor, extramedullary hematopoiesis, video-assisted thoracic surgery

Introduction

Extramedullary hematopoiesis (EMH) is a rare disease associated with hematologic disorders such as myelofibrosis, thalassemia, spherocytosis, and sickleemia. This report describes a patient with no hematologic disorders who was diagnosed with thoracic EMH (TEMH) after a surgical resection by video-assisted thoracic surgery (VATS).

Case

A 48-year-old male was admitted to the Nippon Medical School Main Hospital because of chest pain in August 2006. Chest roentgenogram revealed a spontaneous pneumothorax on the left side. The pneumothorax was only followed up on, not treated, because there was only a minor lung collapse. One month later, chest computed tomography (CT) was used to detect bullous change in the lung. At that time a 1 cm mass lesion was identified on the right posterior mediastinum. A neurogenic tumor was suspected after chest magnetic resonance imaging (MRI). He was informed and thereafter underwent a surgical resection.

A chest roentgenogram showed a very slight lung collapse on the left (Fig. 1). The chest CT revealed a small bullous lesion on the apex of the left lung and a well-demarcated mass measuring 15 × 12 mm in diameter with soft tissue density on the right side of the tenth thoracic vertebra (Fig. 2). On chest MRI, the mass showed a homogenous and slightly high intensity in both T1- and T2-weighted images. There was no permeation to the vertebral hole (Fig. 3). From these images, the mass was diagnosed to be a neurogenic tumor of the posterior mediastinum.

Blood cell count exams revealed a hemoglobin level of 15.2 g/dl, a hematocrit value of 43.6%, a mean corpuscular volume (MCV) of 99.8 fl, a mean corpuscu-
Fig. 1. A chest roentgenogram showed a very slight lung collapse (arrow) on the left.

Fig. 2. The chest CT revealed a well-demarcated mass (arrow), measuring 15 × 12 mm in diameter with soft tissue density on the right side of the tenth thoracic vertebra.

Fig. 3. On chest MRI, the mass showed a homogeneous and slightly high intensity in both T1- and T2-weighted images, (A) and (B) respectively. The vertebral hole had no permeation.

Fig. 4. Pathology revealed a foci of EMH characterized by the presence of myeloid tissue such as megakaryocytes in the resected specimen (Fig. 4). After diagnosis, further hematologic inspections, such as the population and abnormality of red blood cells, were done, but no disorder could be identified.

Discussion

EMH is regarded as a physiologic compensatory mechanism that occurs when the bone marrow is unable to maintain sufficient red cell production to supply body
demand, usually in association with a hematologic disorder such as myelofibrosis, thalassemia, spherocytosis, and sickleemia, and it usually occurs in the spleen, liver, or adrenal gland. TEMH, especially posterior mediastinal EMH, is very rare.

Two hypotheses are considered for the ethiopathogenesis of TEMH.

(1) Heterotopic hematogenesis tissue is captured and allowed to develop into TEMH in the intercostal vein.

(2) Hyperplasia of the marrow grew out of the bone through the lack of a cortex section of the rib.

In the present case, the resected tumor was not associated with the bone based on the CT, MRI, and intraoperative appearance, thus indicating that the TEMH was formed by the mechanism of the first hypothesis.

Most occurrences of TEMH are asymptomatic and found by chance, as in this case, but in rare cases TEMH may cause neurogenic symptoms resulting from epidural localization, thus leading to spinal cord compression\(^1,2\) and thus causing pleural effusion,\(^3\)\(^-\)\(^5\) hemothorax,\(^6\) and chylothorax.\(^7\)

Regarding diagnostic imaging, an \(^{111}\)indium transferring scan as a bone marrow tracer is reported to be useful for the detection of EMH,\(^8\) in addition to CT and an MRI scan. If TEMH is suspected from these images, a needle biopsy should not be recommended because it may induce the hemothorax.\(^1\)

Although EMH is diagnosed or highly suspected from the clinical findings (including the existence of hematological disorders) and images, and if the patient has no symptoms because of the tumor, such as pain or neurothlipsis, basically simple follow-up would be chosen instead of a resection of the EMH. This is because hematologic symptoms such as chronic anemia may worsen after a resection of EMH, thus supporting the inadequate hematogenous functions as a physiological compensatory mechanism that we previously described.

However, if the patient has any symptoms, a surgical procedure is then indicated. In some cases, low-dose radiation or pleurodesis by minocycline instead of a surgical procedure has proven to be very effective for patients with massive pleural effusion or chylothorax resulting from TEMH.\(^3,7\)

Because TEMH is very rare as a mediastinal tumor, it tends often to be overlooked in the differential diagnosis, but with a patient who has hematologic disorder, such as chronic anemia, the possibility of EMH should therefore always be considered. Moreover, an \(^{111}\)indium transferring scan should be performed to confirm or deny TEMH.

However, in regard to a patient who has no hematologic disorder preoperatively, as in the present case, an accurate diagnosis would be very difficult based on only CT or MRI images. Therefore in such cases, a diagnosis can be confirmed only after a surgical resection has been performed.

**Conclusion**

This report documents a case of posterior mediastinal EMH in a 48-year-old male associating with spontaneous pneumothorax on the opposite side. The patient presented with no clinical evidence of anemia, and the diagnosis was successfully made by means of VATS.

This report suggests that such lesions must therefore be considered in the differential diagnosis of posterior mediastinal tumors presenting with no hematologic disorders.

**References**


