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Surgical resection of extramedullary haematopoiesis in the posterior mediastinum

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Abstract

Extramedullary haematopoiesis is a rare disease that is usually associated with haematologic disorders such as thalassemia, myelodysplastic syndrome, and hereditary spherocytosis. It frequently occurs in the liver, spleen, and lymph nodes. Rarely, it occurs in the posterior mediastinum. We report the case of a 59-year-old man with lateral posterior mediastinal masses that were incidentally detected during treatment for hereditary spherocytosis. We performed video-assisted thoracic surgery to confirm the diagnosis and differentiate the masses from neurogenic tumours and other posterior mediastinal diseases. The pathological findings were consistent with intrathoracic extramedullary haematopoiesis. Although extramedullary haematopoiesis can be managed without interventions, surgery may be required in some cases. In such cases, video-assisted thoracoscopic surgery is advised because it is a useful and less invasive procedure.

Introduction

Extramedullary haematopoiesis is a rare disease usually associated with haematologic disorders such as myelodysplastic syndrome. The liver and spleen are common sites for extramedullary haematopoiesis. However, it has been reported in other locations such as the posterior mediastinum [1–3]. It is difficult to differentiate extramedullary haematopoiesis from other tumours, such as neurogenic tumours, in the posterior mediastinum because radiological studies and other less invasive investigations can be limited. We believe that video-assisted thoracoscopic surgery can be useful for extramedullary haematopoiesis.

Case Report

A 59-year-old man with hereditary spherocytosis presented to our hospital for follow-up of his condition. On performing a chest radiographic examination, an abnormal shadow was detected. Physical examination revealed jaundice; however, no other significant findings were observed. Laboratory tests revealed a white blood cell count of 8.04 × 103/μL, haemoglobin level of 12.2 g/dL, platelet count of 9.90 × 105/μL, aspartate aminotransferase level of 122 U/L, alanine aminotransferase level of 127 U/L, and total bilirubin level of 6.83 mg/dL. Other laboratory findings were within the normal ranges. Computed tomography and magnetic resonance imaging of the chest revealed a mass with a maximum diameter of 68 mm in the right posterior mediastinum. The magnetic resonance imaging showed that the tumour was mainly fat component, and was difficult to be distinguished from a malignant tumour such as liposarcoma. Despite the absence of any specific symptoms and the suspicion of extramedullary haematopoiesis, we performed video-assisted thoracoscopic surgery for confirmation, to eliminate other differential diagnoses, including neurogenic tumours, liposarcoma, and other posterior mediastinal diseases, and to avoid the potential risk of bleeding of the mass [4] and spinal cord compression by the lesion because it was located close to the neural foramen.

Surgery was performed under general anaesthesia with isolated lung ventilation in the left lateral decubitus position. Three port site incisions were made in the fourth intercostal space of the anterior axillary line (30 mm), sixth intercostal space of the posterior axillary line (20 mm), and seventh intercostal space of the middle

axillary line (camera port). Thoracoscopy showed tightness and a highly vascularized haemorrhagic mass along the paravertebral region (Fig. 1). Although the mass was hypervascular, we were able to safely remove it using electrically and ultrasonically activated scalpels. There were no signs of air leakage or significant bleeding. At the end of the surgery, a chest tube was inserted into the thorax. The total operation time was 225 min, and total blood loss was 340 mL. The patient's postoperative course was uneventful, and the chest tubes were removed two days after surgery. On postoperative day 9, he was discharged without complications.

A histological examination revealed that the masses comprised mainly mature adipose tissue and a polymorphic population of mature haematopoietic cells (myeloid, lymphoid, erythroid, and megakaryocytic lines) with no heterocysts (Fig. 2). The final pathological diagnosis was extramedullary haematopoiesis.

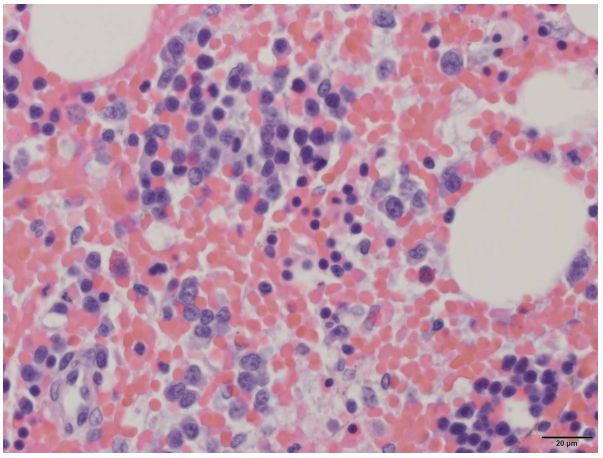

Figure 2. Microscopic findings of the specimen. The histological analysis shows islands of myelopoietic cells surrounded by adipose tissue. Haematoxylin and eosin staining (magnification, ×400).

Discussion

Extramedullary haematopoiesis is defined by the production of blood components (myeloid and erythroid elements) outside of the bone marrow. It was hypothesized that extramedullary haematopoiesis occurs to compensate for haemolytic anaemia caused by spherocytosis, thalassemia, and hereditary spherocytosis, or as a reaction to abnormal bone marrow function seen in disorders such as myelofibrosis and leukaemia or due to the activation of embryonic haematopoietic stem cells at other sites [5]. Although extramedullary haematopoiesis frequently occurs in the liver, spleen, and lymph nodes, it can rarely occur in the posterior mediastinum [1–3]. Asymptomatic patients with posterior mediastinum extramedullary haematopoiesis usually require no treatment, and management usually depends on the patient's symptoms [5]. For symptomatic patients, such as those with symptoms of tumour compression and bleeding, or for those with suspected malignancy, surgical resection may be recommended.

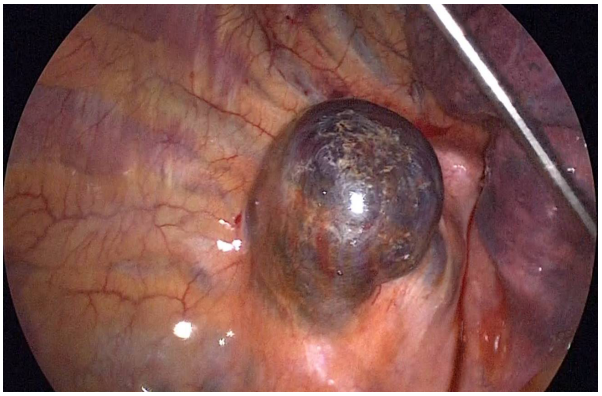

Figure 1. Thoracoscopic findings. Tightness and a highly vascularized mass are seen in the paravertebral region.

Various non-invasive, radiological diagnostic tools such as computed tomography, magnetic resonance imaging, and technetium-99 sulphur colloid radionuclide bone marrow scanning are used to diagnose extramedullary haematopoiesis. However, they can be limited. In addition, less invasive examinations such as fine-needle biopsy can cause organ injury and haemorrhagic complications. In surgical cases, posterior mediastinal extramedullary haematopoiesis removal can be complicated by bleeding of the mass and lung and spinal nerve injury. Therefore, video-assisted thoracoscopic surgery can be beneficial in such cases. In our case, expanding our field of view with thoracoscopy was useful for controlling the haemorrhage and avoiding other organ injuries while completely resecting the masses.

In summary, it is important to identify intrathoracic extramedullary haematopoiesis in patients with chronic anaemic disorders. Video-assisted thoracoscopic surgery can be a useful and less invasive approach for the diagnosis and surgical removal of extramedullary haematopoiesis.

Disclosure Statement

Appropriate written informed consent was obtained for publication of this case report and accompanying images.

References

1. Park JB, Lee SA, Kim YH, et al. 2017. Extramedullary hematopoiesis mimicking mediastinal tumor in a patient with hereditary spherocytosis: case report. Int. J. Surg. Case Rep. 41:223–225.

2. Giblin E, Frankel K, and Mortman K. 2007. Video-assisted thoracoscopic surgery for intrathoracic extramedullary hematopoiesis. J. Minim. Access. Surg. 3:32–34.
3. De Montpréville VT, Dulmet EM, Chapelier AR, et al. 1993. Extramedullary hematopoietic tumors of the posterior mediastinum related to asymptomatic refractory anemia. Chest 104:1623–1624.
4. Xiros N, Economopoulos T, Papageorgiou E, et al. 2001. Massive hemothorax due to intrathoracic extramedullary hematopoiesis in a patient with hereditary spherocytosis. Ann. Hematol. 80:38–40.
5. O'Malley DP. 2007. Benign extramedullary myeloid proliferations. Mod. Pathol. 20:405–415.