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Case Report

Unilateral Intrathoracic Extramedullary Hematopoiesis: A Rare Case Report

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Abstract

Intrathoracic extramedullary hematopoiesis (ITEMH) is an extremely rare condition characterized by the abnormal proliferation of hematopoietic tissue outside the bone marrow. Here, we present a case of unilateral ITEMH in a 56-year-old male without a history of hematological disorders, which to the best of our knowledge, represents the first reported occurrence of this rare tumor condition. The patient presented with respiratory symptoms and was initially misdiagnosed with a mediastinal mass. Subsequent imaging studies revealed a localized lesion in the left thoracic cavity, which was surgically excised. Histopathological examination confirmed the diagnosis of intrathoracic extramedullary hematopoiesis. This case highlights the importance of considering ITEMH in the differential diagnosis of intrathoracic masses, even in the absence of underlying haematological disorders.

Categories: Family/General Practice, Internal Medicine, GEN SURGERY.

Keywords: Unilateral Intrathoracic Extramedullary Hematopoiesis; Rare Tumor Condition; Hematological Disorders; Differential Diagnosis; Surgical Resection; Adjuvant Therapies; Recurrence; Prognosis

Introduction

Intrathoracic extramedullary hematopoiesis is an uncommon phenomenon in which hematopoietic tissue develops outside the bone marrow, typically occurring in patients with underlying haematological disorders such as myelofibrosis or thalassemia. Isolated unilateral ITEMH is an exceedingly rare occurrence, with only a few cases reported in the medical literature. We present a unique case of unilateral ITEMH in a patient without a history of haematological disorders.

Case Presentation

A 56-year-old male, a lifelong non-smoker, presented with a four-month history of gradually worsening dyspnea and left-sided chest pain. Physical examination revealed decreased breath sounds on the left side of the chest. Initial imaging studies, including computed tomography (CT) of the chest, suggested the presence of a mediastinal mass compressing the left lung. The patient underwent a diagnostic video-assisted thoracoscopic surgery (VATS), during which a localized lesion was identified in the left thoracic cavity, adherent to the mediastinal structures. The lesion appeared well-defined and was carefully dissected and excised.

Discussion

Unilateral intrathoracic extramedullary hematopoiesis (ITEMH) is an extremely rare condition characterized by the abnormal proliferation of hematopoietic tissue outside the bone marrow. Although most reported cases of ITEMH are associated with underlying hematological disorders such as myelofibrosis or thalassemia, our case is unique as it occurred in the absence of any known hematological abnormalities. This suggests that there may be other

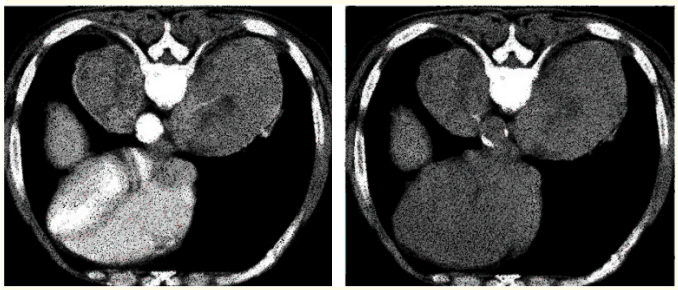

Figure 1: (CT) of the chest, suggesting the presence of a mediastinal mass compressing the left lung.

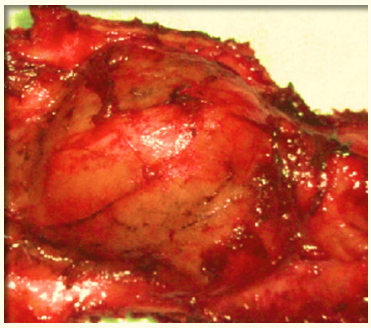

Figure 2: Specimen: well-circumscribed mass measuring 5 cm in greatest dimension.

underlying factors or mechanisms contributing to the development of ITEMH, which require further investigation.

The clinical presentation of ITEMH can mimic other intrathoracic masses, leading to misdiagnosis and delays in appropriate management. In our case, the patient initially presented with respiratory symptoms and imaging studies suggested a mediastinal mass compressing the left lung. This highlights the importance of thorough evaluation and considering ITEMH in the differential diagnosis, even in patients without a history of hematological disorders.

Surgical resection, as performed in our case, can be curative and alleviate symptoms, resulting in a favorable prognosis. The excised mass in our case showed well-defined borders and was composed of hematopoietic tissue with evidence of maturation, confirming the diagnosis of ITEMH. Immunohistochemical staining further supported the presence of CD34-positive hematopoietic cells. The absence of malignancy in the histopathological examination indicates the benign nature of ITEMH in our case.

Histopathological examination

Microscopic examination of the excised specimen revealed a well-circumscribed mass measuring 5 cm in greatest dimension. The mass was composed of hematopoietic tissue, including mature and immature myeloid and erythroid elements, with evidence of hematopoietic cell maturation. No evidence of malignancy was identified. Immunohistochemical staining demonstrated the presence of CD34-positive hematopoietic cells, further supporting the diagnosis of intrathoracic extramedullary hematopoiesis.

The role of adjuvant therapies, such as radiotherapy or chemotherapy, in the management of ITEMH remains uncertain due to the limited number of reported cases and lack of consensus guidelines. The rarity of ITEMH poses challenges in conducting large-scale studies to establish optimal treatment strategies and long-term outcomes. Further research is needed to investigate the underlying mechanisms of ITEMH development, identify potential risk factors, and determine the efficacy of adjuvant therapies.

Follow-up and outcome

Postoperatively, the patient experienced a significant improvement in respiratory symptoms, with resolution of dyspnea and chest pain. Chest radiography and CT scans performed during follow-up visits at 3 months, 6 months, and 1 year postoperatively showed no evidence of recurrent or residual masses. Laboratory investigations, including complete blood count, peripheral blood smear, and bone marrow aspiration, did not reveal any underlying hematological abnormalities, ruling out secondary causes of intrathoracic extramedullary hematopoiesis.

Long-term follow-up is essential for monitoring potential recurrence or progression of ITEMH. This supports the curative potential of surgical resection in localized ITEMH.

Increasing awareness among clinicians about the existence of ITEMH and its clinical and radiological characteristics is crucial for early recognition and appropriate management. Reporting and documenting rare cases, such as ours, contribute to expanding the existing knowledge base and improving understanding of this rare tumor condition.

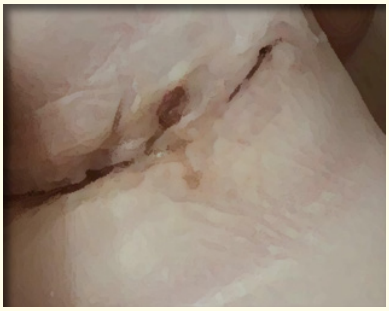

Figure 3: Pod 7wound.

Conclusions

We present a unique case of unilateral intrathoracic extramedullary hematopoiesis in a patient without underlying hematological disorders. This case highlights the importance of considering ITEMH in the differential diagnosis of intrathoracic masses, even in the absence of known hematological abnormalities. Increased awareness of this condition may aid in timely diagnosis and appropriate management, leading to improved patient outcomes. Further research and case reports are necessary to enhance our understanding of this rare tumor condition.

Disclosures

- **Human subjects**: Consent was obtained or waived by all participants in this study.
- **Conflicts of interest**: In compliance with the ICMJE uniform disclosure form, all authors declare the following:
- **Payment/services Info**: All authors have declared that no financial support was received from any organization for the submitted work.
- **Financial relationships**: All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work.
- **Other relationships**: All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

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