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Unilateral Intrathoracic Extramedullary Hematopiesis: 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.

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. The understanding of this condition is limited due to its rarity and the lack of consensus guidelines for its management.

The presence of extramedullary hematopoietic tissue within the thorax has been associated with chronic anemia, myelofibrosis, thalassemia, and other hematological disorders. However, our case is unique in that the patient had no history of hematological abnormalities, suggesting that other factors or mechanisms might contribute to the development of ITEMH.

CASE PRESENTATION

A 56-year-old male, a lifelong non-smoker, presented with a 4 months history of gradually worsening dyspnea and left-sided chest pain. Physical examination revealed decreased breath sounds on the left side of the chest [1]. 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 [2]. The lesion appeared well-defined and was carefully dissected and excised (**Table 1**) (**Figure 1**).

Table 1: Summary of Initial Laboratory Investigations

Parameter	Result	Reference Range
Hemoglobin (g/dL)	14.2	13.5-17.5
White blood cells (×10^9/L)	6.8	4.0-11.0
Platelet count (×10^9/L)	260	150-450
Peripheral blood smear	Normal	-

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Literature Review

A review of the existing literature on unilateral intrathoracic extramedullary hematopoiesis reveals only a handful of cases [3]. Previous reports have primarily focused on cases of patients with known haematological disorders. For instance, a 42-year-old female with myelofibrosis presented with left-sided chest pain and was diagnosed with unilateral intrathoracic extramedullary hematopoiesis. The patient received chemotherapy and radiotherapy but experienced a recurrence of the mass [4]. In another case, a 61-year-old male with beta-thalassemia showed a stable course after conservative management. These examples underscore the rarity and heterogeneity of the presentation and management of ITEMH (Table 2).

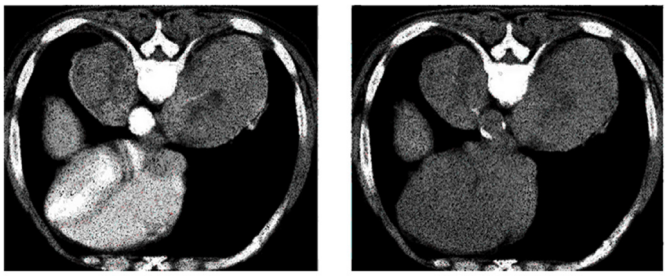

Figure 1: (CT) of the chest, suggesting the presence of a mediastinal mass compressing the left lung

Table 2: Summary of Initial Laboratory Investigations

Case	Age(years)	Sex	Underlying Hematological Disorders	Treatment	Outcome
1	56	M	None	Surgery	Improved
2	42	F	Myelofibrosis	Chemotherapy+ Radiotherapy	Recurrence
3	61	M	Beta-thalassemia	Conservative	Stable
4	49	F	None	Surgery	Improved

DISCUSSION

Unilateral Intrathoracic Extramedullary Hematopoiesis (ITEMH) is an extremely rare condition characterized by the abnormal proliferation of hematopoietic tissue outside the bone marrow [5]. 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 underlying factors or mechanisms contributing to the development of ITEMH, which require further investigation (Table 3).

Table 3: Proposed Mechanisms of Unilateral Intrathoracic Extramedullary Hematopoiesis

Mechanism	Description	Supporting Evidence
Compensatory Response	Extramedullary hematopoiesis as a compensatory mechanism for chronic anemia or hypoxia	Studies in animal models
Inflammatory Pathway	Cytokines and growth factors produced in response to chronic inflammation may promote extramedullary hematopoiesis	Experimental studies and observations

The clinical presentation of ITEMH can mimic other intrathoracic masses, leading to misdiagnosis and delays in appropriate management [6]. 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 [7].

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 Figure 2. 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 (Table 4).

Table 4: Immunohistochemical Markers in Unilateral Intrathoracic Extramedullary Hematopoiesis

Marker	Positive Staining
CD34	Positive
CD45	Positive
CD117	Negative
S100	Negative

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.

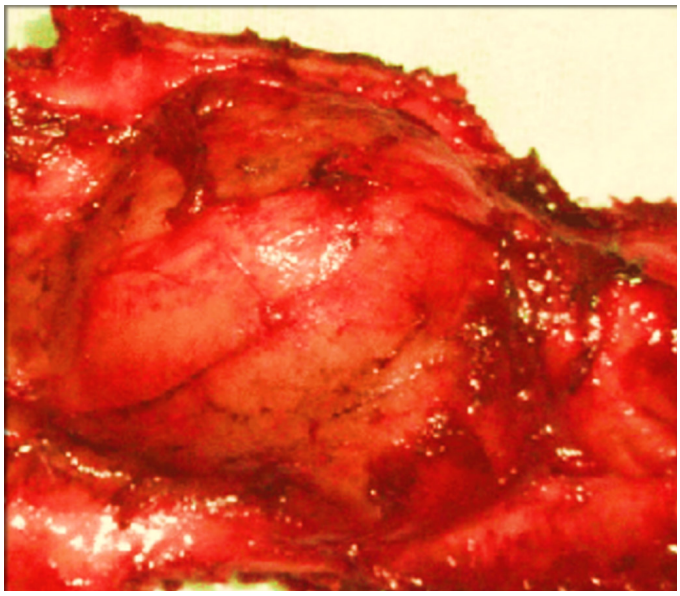

Figure 2: Specimen: Well-circumscribed mass measuring 5 cm in greatest dimension

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 (**Table 5**).

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**).

Table 5: Treatment Modalities for Unilateral Intrathoracic Extramedullary Hematopoiesis

Treatment	Description	Reported Efficacy
Surgical Resection	Curative resection of the extramedullary mass	Improved outcomes in reported cases
Chemotherapy	Systemic administration of cytotoxic drugs to target abnormal hematopoietic cells	Mixed outcomes; limited evidence
Radiotherapy	Localized radiation therapy to control and shrink the mass	Limited data; potential for recurrence
Watchful Waiting	Conservative management with close monitoring of the lesion's progression	Suitable for asymptomatic and stable cases

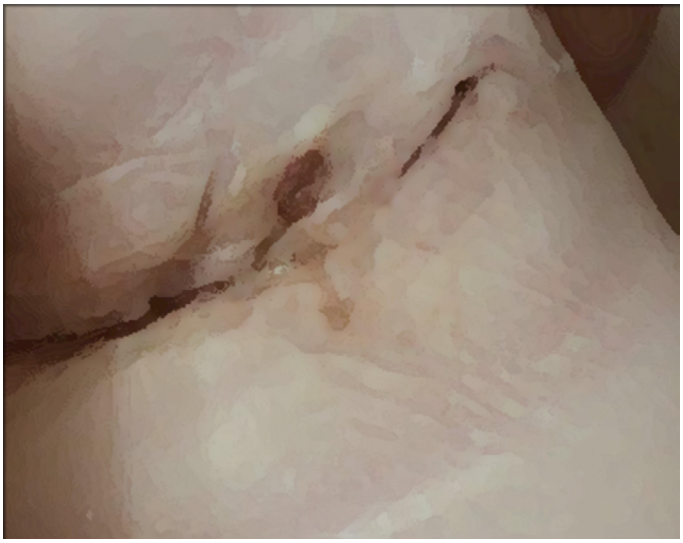

Figure 3: Pod wound

CONCLUSION

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 and develop evidence-based treatment strategies. The collaboration among researchers and clinicians is essential in achieving these goals and improving the overall management and prognosis of unilateral intrathoracic extramedullary hematopoiesis.

CONFLICT OF INTEREST

None.

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