

# A Case of Group Five Pulmonary Hypertension with Extramedullary Hematopoiesis Secondary to Multifactorial Causes

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Extramedullary pulmonary hematopoiesis is a rare condition and an even more uncommon cause of pulmonary hypertension [1-2]. Here, a 34-year-old man with a history of Beta-thalassemia presented with progressive shortness of breath for two weeks. He was found to be in respiratory distress with significant hypoxic and hypercapnic respiratory failure requiring intubation. Initial labs were not concerning for infectious or cardiac etiology. Chest radiograph showed large pulmonary masses [Figure 1] and Chest computer-tomography revealed multi-chamber cardiomegaly with multiple paraspinal and pleural-based masses [Figure 2]. Given the patient's hematologic history, a nuclear medicine bone marrow scan was previously performed prior to this admission to evaluate the paraspinal masses which showed widespread marrow expansion and extensive extra-medullary hematopoiesis. A follow-up bone marrow biopsy was performed which revealed increased marrow activity with erythroid hyperplasia. Furthermore, the patient previously underwent a right heart catheterization which yielded a pulmonary artery pressure (PAP) of 35 mmHg, pulmonary vascular resistance (PVR) of 3.9 woods units, and a pulmonary-capillary wedge pressure (PCWP) of 10 mmHg representing a pre-capillary cause of pulmonary hypertension. The patient was supposed to be on tadalafil prior to this admission but he stopped taking it due to insurance difficulties. Treatment was initiated with intravenous milrinone, furosemide, and inhaled epoprostenol. The patient's oxygenation and ventilatory status improved and he was extubated successfully to high flow nasal cannula and further to nasal cannula before discharge. He was weaned off milrinone and inhaled epoprostenol then he was restarted on tadalafil, an oral phosphodiesterase (PDE-5) inhibitor, as well as oral furosemide upon discharge.

The likely cause of the patient's pulmonary hypertension was multifactorial. There is a precapillary component as represented by the elevated PVR value likely due to the reduced nitric oxide availability from consumption by free hemoglobin and proliferative arteriopathy. Additionally, the extramedullary hematopoiesis noted on his imaging also contributed to due to pulmonary vascular compression rather than parenchymal involvement, as the nuclear bone marrow scan did not show involvement of the lung parenchyma. No extraction or decompression treatment was planned due to the clinical improvement of the patient with medication. In all, this is a rare clinical

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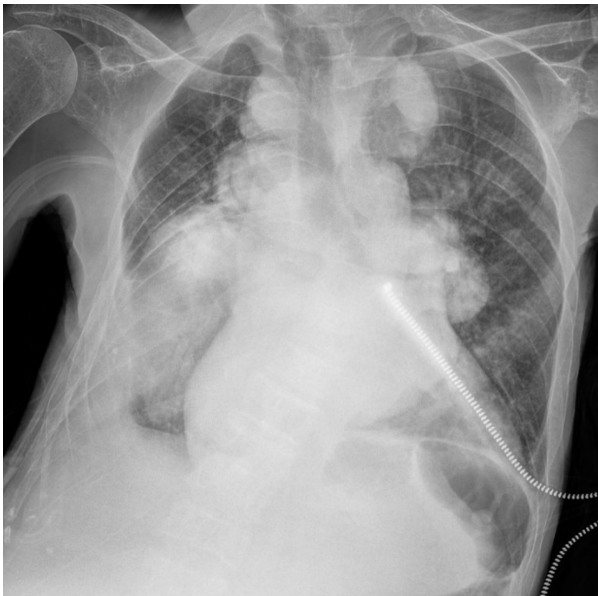
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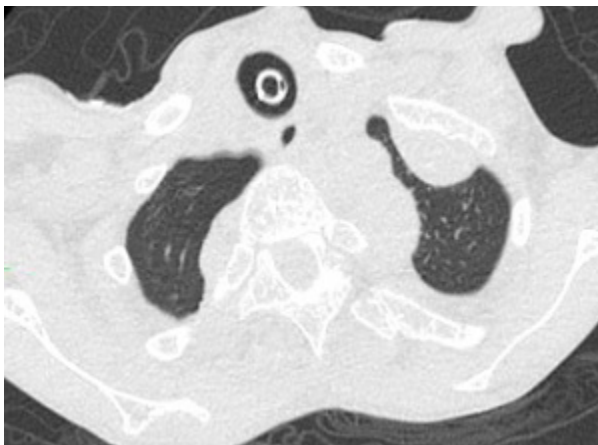
instance of pulmonary hypertension caused by hematologic variation and hematopoietic stem cell infiltration of the thoracic cavity. The mechanism by which extramedullary hematopoiesis induced pulmonary hypertension is not fully understood but is likely multifactorial including lung parenchymal restriction, thrombocytosis, hypercoagulability, and left heart failure [3].

**Figure 1.** Frontal chest radiograph demonstrating cardiomegaly and large, smooth, sharply delineated, lobulated thoracic paraspinal masses bilaterally, which signify sites of extramedullary hematopoiesis.



**Figure 2.** Axial CT images showing large paraspinal and pleural based consolidations. Images (a) and (b) demonstrate a mass extending from the anterior rib into the pleural space along with multiple paraspinal masses. Images (c) and (d) display multiple larger masses extending from the spine and causing restriction of the lung parenchyma.

**Figure 2a**



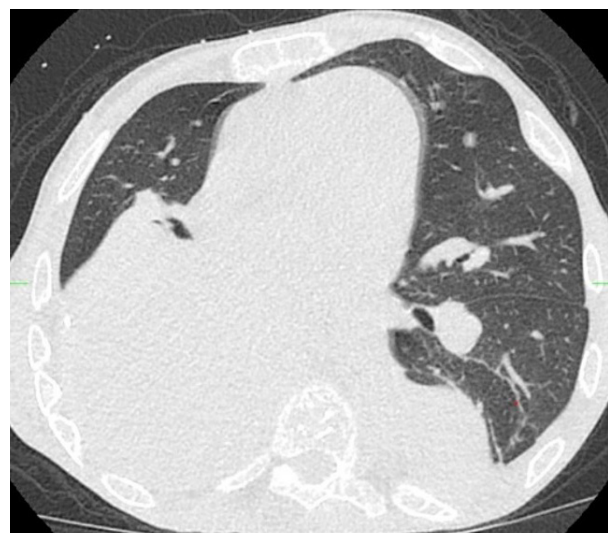
**Figure 2b**



**Figure 2c**



**Figure 2d**



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