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BEYOND THE SCORES: UNMASKING A SILENT PULMONARY EMBOLISM IN ESSENTIAL THROMBOCYTHEMIA.

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Background

Pulmonary embolism (PE) is a potentially fatal event, responsible for up to 300,000 deaths annually in Europe. Over 50% of patients with proximal deep vein thrombosis have concurrent PE. The Pulmonary Embolism Severity Index (PESI) and clinical prediction scores such as Wells and Geneva are commonly used for risk stratification.

Essential thrombocythemia (ET) is a myeloproliferative neoplasm characterized by clonal thrombocytosis and increased thrombotic and hemorrhagic risk. Severe anemia and extreme thrombocytosis (≥1,000,000/μL) are associated with leukemic transformation. Bone marrow biopsy is key to defining disease stage.

Case Presentation

A 79-year-old woman with ischemic heart disease and known ET on hydroxyurea (500 mg/day) was admitted for acute respiratory failure and cardiac decompensation. She showed mild tachycardia (110 bpm), tachypnea (25 breaths/min), SpO_2 85%, and respiratory alkalosis (pH 7.54). Clinical signs suggested pulmonary congestion.

Lab results revealed hemoglobin 10 g/dL, leukocytes 30,000/μL (>80% neutrophils), platelets 780,000/μL, and BNP 572 pg/mL. Echocardiography showed preserved systolic function, moderate mitral/tricuspid insufficiency, and pulmonary artery pressure of 45 mmHg.

Despite low Wells and intermediate Geneva scores, the lack of response to therapy prompted further investigation. D-dimer was 1.75 μg/mL. CT pulmonary angiography confirmed segmental acute PE. Doppler ultrasound excluded DVT.

Fondaparinux 7.5 mg/day was initiated and later switched to apixaban 5 mg BID, with gradual clinical and respiratory improvement. During hospitalization, hemoglobin declined and platelet count increased (Table 1). No circulating blasts were found on peripheral smear. The patient declined bone marrow biopsy.

Genetic testing revealed the JAK2V617F mutation, while BCR-ABL rearrangements were absent.

Conclusion

This case highlights the need to suspect PE in ET patients with respiratory symptoms, even when embolic risk scores are low or intermediate and other diagnoses (e.g., heart failure) are plausible. ET predisposes to thromboembolic complications, particularly in the presence of leukocytosis. Anemia, severe thrombocytosis and new thrombotic-hemorrhagic events concern for possible disease progression. A bone marrow biopsy remains essential when transformation is suspected.

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	1° day	5° day	10° day
White blood cells	30.000 /μL	28.500 / μL	29.600 / μL
Hemoglobin	10 g/dL	9,3 g/dL	7,8 g/dL
Platelets	780.000 / μL	950.000 /μL	1.280.000 /μL
Lactic dehydrogenase	480 UI/L		