A Case of Mistaken Identity: Differentiating Pulmonary Hypertension Subtypes in a Complex Presentation

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Pulmonary hypertension (PH) is a heterogeneous condition defined by elevated pulmonary artery pressures that encompass five distinct subgroups under the World Health Organization (WHO) classification system. It effects 1% of the global population. Specifically, PH is defined by a mean pulmonary artery pressure (mPAP) of greater than or equal to 20 mmHg at rest measured on right heart catheterization. Diagnosis and classification can be challenging due to overlapping clinical features and the nonspecific nature of presenting symptoms. This case highlights the diagnostic complexity of pulmonary hypertension in a patient with multiple comorbidities.

Case

We present a middle-aged female with a past medical history of pulmonary hypertension, recurrent pulmonary emboli including a saddle embolus, pregnancy losses, obesity, OSA, hypertension, laparoscopic sleeve gastrectomy, and rheumatoid arthritis who presented to the ER with progressive dyspnea on exertion, chest pain, fatigue, and bilateral lower extremity edema.
A few weeks prior to this admission, she was treated in a different hospital system for bilateral pulmonary emboli and underwent catheter directed thrombolysis and was discharged on Eliquis.

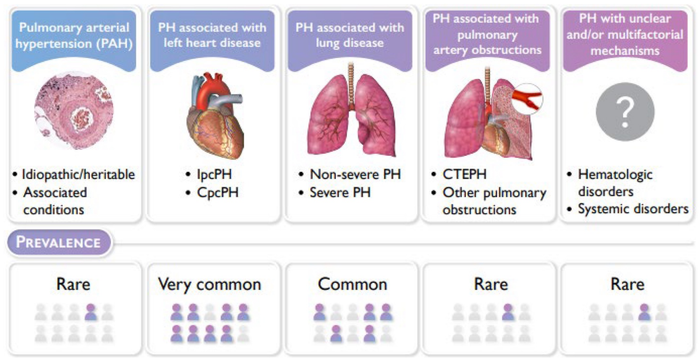
Workup and Discussion

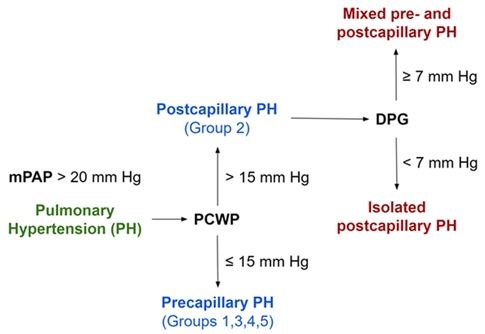
Upon admission, patients blood pressure was 142/92, she was afebrile, had a pulse of 93, and an SpO2 of 100% on 4L of oxygen via nasal cannula. BMI was 54.9. Labs performed showed a Troponin of 18, anti-factor Xa of 1.70, and BNP of 1239. Additionally, she had a protime of 22.2, an INR 2.0, a HGB 9.5, and a MCV of 68.5.

Portable chest x-ray showed cardiomegaly. CT angiogram of the chest showed filling defects of the branches of both pulmonary arteries, an enlarged heart with a RV/LV ratio of 1.53, a small pericardial effusion, and a small left plural effusion. Additionally, a wedge shaped subpleural consolidation was seen in the left lower lobe and mosaic attenuation was seen in the bilateral lungs which was interpreted as potential small airway disease.

The patient started IV diuresis with Bumex as well as IV heparin therapy. She was subsequently hospitalized in the ICU step-down unit.

During hospitalization, ANA, anti-dsDNA, anticardiolipin, B2-glycoprotein, lupus anticoagulant, and protein c/s levels were ordered. Anti-dsDNA antibodies were positive. 2D echo was performed and showed right heart strain with an EF 60-65% with dilated RA and PA pressure of 87 mmHg. Cardiology was consulted and they performed a right heart catheterization. This showed a PA pressure of 97/40 (63) mmHg and a pulmonary capillary wedge of 21 mmHg. She was diagnosed with pulmonary hypertension groups 2, 3, and 4.





Upon discharge from this hospitalization the patient was placed on Ricoiquat for treatment of proposed CTEPH and was followed up closely in the outpatient pulmonary office locally. She was also seen at a tertiary care center pulmonary hypertension clinic.

The patient experienced worsening of symptoms on Ricoiquat. Further workup was conducted by the outpatient pulmonary hypertension clinic 7 months from hospitalization that included RHC and V/Q scan. RHC showed PA pressure of 97/40 (63) mmHg and a pulmonary capillary wedge of 21 mmHg. V/Q scan was without evidence of PE. Original diagnosis of CTEPH was reconsidered and Riociquat was discontinued. Treatment is now focused on her OSA and HF for her pre- and post-capillary pulmonary hypertension.

Conclusion

Accurate differentiation between these subtypes is essential, as management strategies and prognoses vary significantly.

The overlapping clinical features among PH subtypes emphasize the need for multidisciplinary and comprehensive evaluation to guide appropriate management.

Utilizing alternative diagnostic modalities to differentiate subgroups of PH should be emphasized in future practice when overlapping risk factors exist.

Additionally, ongoing re-evaluation of the diagnosis as the patient's condition evolves is crucial to avoid diagnostic anchoring and to ensure accurate identification of PH.

References