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A Case Report on Severe Pulmonary Arterial Hypertension (PAH)

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ABSTRACT

The pathobiological mechanisms of pulmonary arterial hypertension (PAH) are multifaceted and progressive, involving the endothelin, nitric oxide, and prostacyclin pathways. Presently available treatments for PAH focus on one of these pathways; in more extreme circumstances or when the disease is deteriorating, these treatments may be combined to several pathways concurrently. Treatment combinations can be applied upfront (using two or more therapies in patients who are not yet receiving treatment) or sequentially (intensifying from initial monotherapy). Combination therapy has long been thought of as a treatment option for PAH, but the evidence for this has usually come from registry data, expert opinion, and clinical experience.

INTRODUCTION:

A rare condition known as pulmonary arterial hypertension (PAH) causes the pulmonary vasculature to gradually remodel, increasing the risk of pulmonary vascular resistance, right ventricular failure, and eventually death.The most recent classification of pulmonary hypertension places PAH in group 1 and states that it can be idiopathic, heritable, or linked to HIV infection, connective tissue disease, congenital heart disease, portal hypertension, and exposure to toxins or medications.[1]In a physiological state, low blood pressure and low vascular resistance define pulmonary circulation. Pulmonary hypertension (PH), one of several pathological conditions linked to elevated pulmonary pressure, is one example. When it comes to left heart disease, lung disease, or pulmonary thromboembolic disease, PH can be categorized as either pulmonary arterial hypertension (PAH) or PH. In addition to being idiopathic, familial, or linked to several other conditions, PAH is characterized by the absence of the latter causes.[2] The prognosis of PAH patients has improved due to advancements in surgical techniques, physical therapy, other supportive therapies, and available PAH-specific drug therapies. Numerous large registries data show that survival has increased over time.[3]Pre-capillary pulmonary hypertension (PH), which is defined as an elevated mean pulmonary arterial pressure ($\bar{P}_{pa}$) of more than 25 mmHg at rest and no other possible cause (e.g., PH from lung diseases, chronic thromboembolic PH, or other diseases), is the basis for the diagnosis of PAH.[4]

CASE HISTORY:

A 54-year-old female patient was admitted in general medicine with chief complaints of SOB (grade 4) since 3 days, orthopnea+ , B/L pitting oedemasince 4 days (progressive) , H/O fever on and off, cough productive+, patient had a history of PTB 8 years ago used medications for 8 months , had history of covid-19, on examination the patient was conscious , coherent and tachypneic, respiratory rate: 26/min, bp: 90/45mmhg, CVS : S1S2+, RS:BAE+ B/L crepts +, Spo2 :75% on O_2, P/A: soft, CNS : NFND.

Laboratory investigations of that patient has elevated WBC levels of 18,000 cells , elevated D dimers of 3.53 mg/l, CT pulmonary angiography shows main pulmonary artery – 31mm dilated,mild pericardial effusion noted with maximum thickness of 10mm, right mild pleural effusion, patchy consolidation with bronchiectatic changes noted in anterior and posterior segments of upper lobe, basal segments of right and left lower lobes lingular segment of left upper lobe, Liver: 5mm calcified granuloma noted in right lobe of liver.

Based on the subject and objective data the patient was found to have severe PAH with lower respiratory tract infection, B/L Lt consolidation, secondary to interstitial lung disease and the patient was treated with O2 inhalation @ 2L/hr, Nebulization with duolin TID, Budecort BD, Amoxicillin potassium clavulanate 1.2mg BD, Syrup lactulose 10ml OD H/S, inj pan 40 mg OD, Tab PCM 500mg SOS, dobutamine 5cc in 45 ccs NS @3cc/hr.

DISCUSSION:

PAH is anorphan disease in which there is remodelling of the pulmonary vasculature which progressively leads to increased pulmonary vascular resistance, right ventricular failure and ultimately death. [(1)]Patients with severe PAH have a poor prognosis. According to the French registry, which included 82.6% of patients in functional class III/IV, patients enrolled in functional class IV had a 3-year survival rate of 38%, while the corresponding rates for WHO functional classes I/II and III were 80% and 60%.[(5)]The three primary biological signalling pathways in PAH that are currently known to exist are the endothelin, nitric oxide, and prostacyclin pathways. Over time, these pathways have been the focus of much-improveD treatment options.[(6)]In the recent update of the treatment algorithm for World Health Organization functional class IV in pulmonary arterial hypertension (PAH), the first-line therapy includes a range of medications such as epoprostenol, Ambrisentan, bosentan, macitentan, riociguat, sildenafil, tadalafil, and analogues of thermostable prostacyclin. Additionally, treprostinil (subcutaneous, intravenous, and inhaled) and iloprost (intravenous and inhaled) are among the recommended treatments. It's important to note that individual treatment plans may vary, and consulting with a healthcare professional is essential for personalized advice [(7)]. In this situation, the doctor could have performed standard diagnostic tests following the recommendations outlined in Vachiery's review. The diagnosis of pre-capillary pulmonary hypertension (PH) involves confirming an elevated mean pulmonary arterial pressure ($\bar{P}pa$) of over 25 mmHg at rest hile excluding other potential causes like PH associated with lung diseases, chronic thromboembolic PH, or other underlying conditions. This specific criterion serves as the foundation for identifying and diagnosing pulmonary arterial hypertension (PAH)[(4)].In this case, the diagnosis appears to have relied solely on CT pulmonary angiography. However, Paul Corris suggests therapeutic management according to the World Health Organization (WHO) functional class IV. It seems the physician opted for a treatment approach involving antibiotics, paracetamol, pantoprazole, lactulose, dobutamine, and nebulization for a respiratory tract infection, which may not align with the recommended management for WHO functional class IV[(7)]. The healthcare team needs to reassess and

consider appropriate interventions based on the patient's functional class and overall condition.

CONCLUSION:

In this situation, my suggestion is that the physician should have considered using one of the first-line therapies specified for severe pulmonary arterial hypertension (PAH) in WHO functional class IV. This approach is in line with established treatment guidelines, aiming to optimize patient care by addressing the severity of the condition through appropriate interventions. It emphasizes the importance of tailoring the treatment plan to the patient's functional class and adhering to recognized therapeutic protocols for managing PAH.

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