Clinical Case: Living Breathless

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Abstract

Pulmonary hypertension is a hemodynamic state defined by a resting mean pulmonary artery pressure ≥ 25 mmHg, calculated by right cardiac catheterization. Its main symptom is progressive effort dyspnea, which can sometimes lead to misdiagnosis and delays in proper treatment.

We present the case of a 35-year-old woman diagnosed with pulmonary arterial hypertension after 4 months of progressive dyspnea and exertional syncope. Patient condition improved following treatment.

Keywords: Pulmonary Hypertension; Dyspnea; Exertional Syncope

Abbreviations

PH: Pulmonary Hypertension; mPAP: Mean Pulmonary Artery Pressure; PAH: Pulmonary Arterial Hypertension; CTEPH: Chronic Thromboembolic Pulmonary Hypertension; PWP: Pulmonary Wedge Pressure; ED: Emergency Department; ECG: Electrocardiogram; TTE: Transthoracic Echocardiogram; V/Q: Ventilation/Perfusion; PE: Pulmonary Embolism; RCC: Right Cardiac Catheterization; IPAH: Idiopathic Pulmonary Arterial Hypertension; RV: Right Ventricle; CT: Computed Tomography

Introduction

The definition of pulmonary hypertension (PH) is mainly hemodynamic, determined by a resting mean pulmonary artery pressure (mPAP) ≥ 25 mmHg [1]. PH may be found in multiple diseases with different physiopathology, treatment and prognosis. Thus, etiological diagnosis is paramount.

PH classification includes 5 groups [2]: 1) Pulmonary arterial hypertension (PAH), including idiopathic PH, heritable forms, PH secondary to drugs and toxins, PH associated with other diseases, such as congenital heart disease, HIV infection or connective tissue diseases, and pulmonary veno-occlusive disease and pulmonary hemangiomatosis; 2) PH secondary to left heart disease; 3) PH secondary to pulmonary diseases/hypoxia; 4) Chronic thromboembolic pulmonary hypertension (CTEPH) and other pulmonary artery obstructions; and 5) PH with unclear and/or multifactorial mechanisms.

PAH patients feature precapillary PH, defined by pulmonary wedge pressure (PWP) ≤ 15 mmHg in the absence of other precapillary PH, such as PH caused by pulmonary diseases, CTEPH or other diseases [1,3].
PH diagnosis starts with clinical suspicion, requires confirmation, determining etiology, assessing functional repercussion, severity and specific treatment.

Clinical Case

35-year-old woman with no significant medical history and treated with oral contraceptives consults with the Emergency Department (ED) in our hospital for dyspnea at rest and exertional syncope. Four months earlier, she had consulted with her primary care doctor for dyspnea on moderate exertion; at that time, the physical examination did not show alterations, and the tests (blood test, chest x-ray and spirometry) were normal. Thus, assessment by the Cardiology Unit was requested, which took place three months later. The electrocardiogram (ECG) showed right axis deviation, right bundle branch block and right ventricular hypertrophy, and the transthoracic echocardiogram (TTE) showed right-side overload and PH data; the day before her consultation with our ED, the patient underwent a ventilation/perfusion (V/Q) lung scan, showing multiple hypoperfusion areas in her left upper lobe, lingula, left lower lobe, right upper lobe, left lung parenchyma and basal segments of the right lower lobe, with normal pulmonary ventilation. The test is reported as compatible with bilateral, multisegmental pulmonary embolism (PE) in a subacute stage and the patient’s admission is decided. The physical examination does not show significant findings, except for tachycardia, tendency to arterial hypotension and low oxygen saturation for the first 24 hours. The study is completed with: 1) full blood test, including hormonal, autoimmunity and tumor marker studies, showing a PO2 value of 55 mmHg and a Pro-BNP value of 1478 pg/ml; 2) CT angiography of the chest, showing PH data affecting the right chambers, but no endoluminal filling defect in pulmonary arteries and branches suggesting PE; and 3) right cardiac catheterization (RCC) showing mPAP of 46 mmHg and PWP of 9 mmHg. Severe idiopathic PAH in functional class IV is diagnosed, and treatment starts with tadalafil (40 mg/day), ambrisentan (10 mg/day) and acenocoumarol, with good clinical and functional response.
Clinical Case: Living Breathless


Figure 2

Figure 3
Clinical Case: Living Breathless

Discussion

In Europe, PAH prevalence and incidence are 15 - 60 and 5 - 10 cases/million people/year, respectively [4]. Idiopathic PAH (IPAH) is a sporadic disease, with no family history of PAH nor known triggering factor, with an average age of 50 - 65 years at the time of diagnosis.

Dyspnea of unclear cause requires considering the PH diagnosis. PH symptoms are unspecific and mainly related to progressive right ventricle (RV) failure. Symptoms include dyspnea, fatigue, weakness, angina and syncope; less frequently, dry cough, nausea and vomiting induced by exertion may occur. The physical examination may show an accentuated pulmonary component of the second heart sound, a third RV sound, a pansystolic murmur of tricuspid regurgitation and a diastolic murmur of pulmonary insufficiency. Patients in the advanced stage of the disease show high jugular venous pressure, hepatomegaly, ascites and peripheral edema. Pulmonary auscultation is usually normal [4].

ECG findings may include P pulmonale, right axis deviation, RV hypertrophy, right bundle branch block and QTc prolongation, indicating a severe disease in this case [5]. A normal ECG does not exclude diagnosis.

Only 10% of all IPAH patients show a normal chest x-ray at diagnosis. Findings include central pulmonary arterial dilation and RV and atrial enlargement [6].

TTE allows to see PH effects on the heart and estimate PAP with continuous-wave Doppler measurements. The test should be run whenever PH is suspected, although it is not enough to make therapeutic decisions, and should be supported by cardiac catheterization [4].

V/Q lung scan is the screening method of choice for its greater sensitivity compared to CT angiography of pulmonary arteries, especially in hospitals with less experience [7]. A normal or low-probability V/Q lung scan effectively excludes PE as a cause of PH [8]. V/Q lung scans may show small perfusion defects in PH, but also in other diseases, such as pulmonary veno-occlusive disease [4]. A pathological scan does not exclude running other complementary tests.

CT is a widely available imaging technique allowing to assess pulmonary arterial or RV dilation, and to identify the cause of PH [9]. The test also provides information on the lung parenchyma and vessels, and it allows to confirm PE [8].

RCC is necessary to confirm PAH and CTEPH diagnosis [3]. The test also allows to assess the severity of the disease and provides prognostic information.

World Health Organization Functional Class is used to determine PH prognosis, both at diagnosis and follow-up [10].

The goal of PAH treatment is the clinical stabilization of the patient, avoiding right heart failure and disease progression. The recommended initial treatment for high-risk patients is a combined therapy including an endothelin receptor antagonist, a phosphodiesterase-5 inhibitor and an intravenously-administered prostaglandin analog [3].

Survival is higher in patients with oral anticoagulant therapy [11]. The high prevalence of thrombotic lesions in studies with IPAH patients, alterations in coagulation and unspecific venous thromboembolism risk factors such as immobility justify this therapy [12,13].

Bibliography


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