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Case Report

Idiopathic Pulmonary Arterial Hypertension Was Diagnosed Initially by the Computed Tomographic Angiogram

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Idiopathic pulmonary arterial hypertension (IPAH) is a rare and progressive disease with non-specific signs and symptoms. A 50-year-old woman with IPAH presented to the emergency department (ED) with a complaint of episodic dyspnea that had persisted for the previous two months. Based on the findings of the initial chest computed tomographic angiography conducted in the ED, we suspected pulmonary hypertension. IPAH was eventually confirmed following a series of investigations, including right heart catheterization. The history of this interesting case is reported with a review of the relevant literature.

Key words: *computed tomographic angiograms, dyspnea, pulmonary arterial hypertension*

Introduction

Idiopathic pulmonary arterial hypertension (IPAH) is a rare, progressive and fatal disease; characterized by elevated pulmonary vascular resistance, with non-specific symptoms and signs. Because of the vague symptoms, IPAH may take several months to diagnose.[1] We describe a case of IPAH presenting to the emergency department (ED) with history of episodic dyspnea for two months. However, IPAH was initially suspected by the findings of chest computed tomographic angiogram (CTA). The objective of this report is to create awareness of this life threatening condition so that IPAH will be considered in the differential diagnosis for the common symptom, like dyspnea.

Case Report

A previously healthy 50-year-old woman was brought to our ED due to progressive increase in the extent of dyspnea for two months. Her past history was negative for major systemic diseases. At ED, the patient was alert. Blood pressure: 126/87 mmHg, heart rate: 78 beats/min, respirator rate: 25 beats/min, and body temperature was 36.5°C. Her initial oxygen saturation by pulse oximetry was 87% on room air. Her physical examinations were unremarkable except increased pulmonary second heart sound on cardiac auscultation. Chest was clear on auscultation. Initial laboratory studies including complete blood count and biochemistry data were not remarkable. Electrocardiogram (ECG) showed right axis deviation, prominent R wave at V1, which implied right ventricle hypertrophy (Fig. 1). Chest film revealed borderline bulging of left pulmonary conus but without cardiomegaly (Fig. 2). The SpO_2 increased to 95% with O_2 therapy (2 L/min) via nasal cannula. Pulmonary embolism was suspected and chest CTA was done, which showed no remarkable filling defect in major pulmonary arteries.

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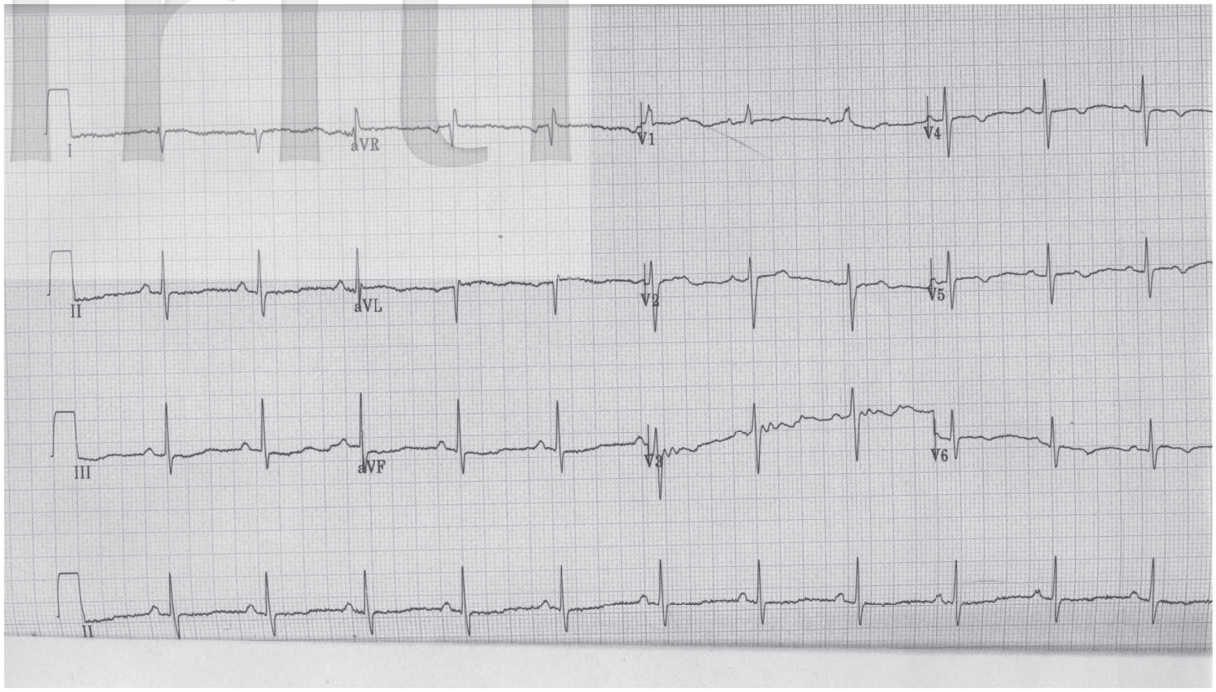

Fig. 1. The 12-lead electrocardiography showed sinus rhythm, right axis deviation, R/s > 1 in V1, which suggesting right ventricular hypertrophy.

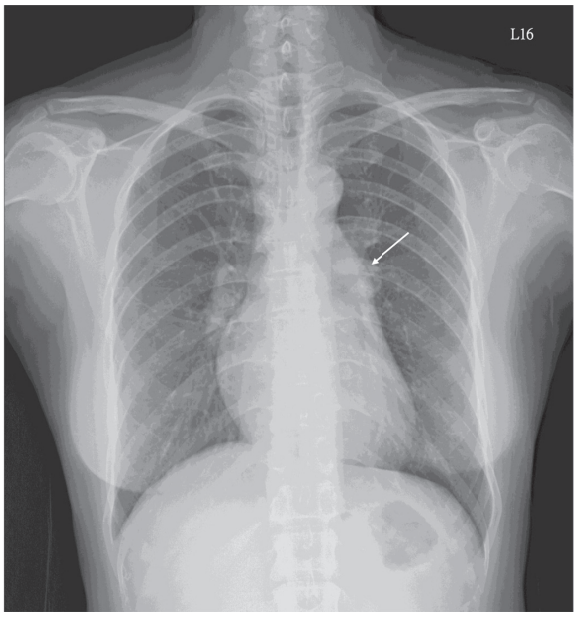

Fig. 2. Chest X-ray depicted borderline bulging of left pulmonary conus (arrow), and normal cardiac size and lung markings.

However, enlarged pulmonary arteries were noted, which suggested the presence of pulmonary hypertension (PH) (Fig. 3). Patient was then admitted for further diagnostic workup.

After admission, the blood testing for connective tissue diseases and human immunodeficiency virus (HIV) titers, abdominal ultrasound, polysomnography, pulmonary function test, and lung perfusion studies, all showed no remarkable findings. The B-type natriuretic peptide level at ward (two days later) was 128.3 pg/mL. (reference range < 100). Transthoracic echocardiography demonstrated enlarged right ventricle and right atrium (RA) and D-shape of left ventricle (Fig. 4). Doppler echocardiography revealed mild pulmonary regurgitation, and moderate tricuspid regurgitation. The estimated maximum instantaneous pulmonary artery (PA) systolic pressure was 135 mmHg and mean PA pressure was 72 mmHg. The cardiac catheterization revealed a normal left ventricular angiogram, mean RA pressure was eight mmHg, PA pressure was 105/42 mmHg, mean PA pressure was 60 mmHg, pulmonary capillary wedge pressure (PCWP) was seven mmHg. Cardiac index was 1.99 L/min/m^2 and pulmonary vascular resistance 20.15 wood units. She was eventually diagnosed as IPAH by exclusion of other known causes.

The patient began to receive subcutaneous infusion treprostinil sodium (Remodulin®) treatment on day eight days after admission. She was discharged with some symptomatic improvement after 11 days hospital-

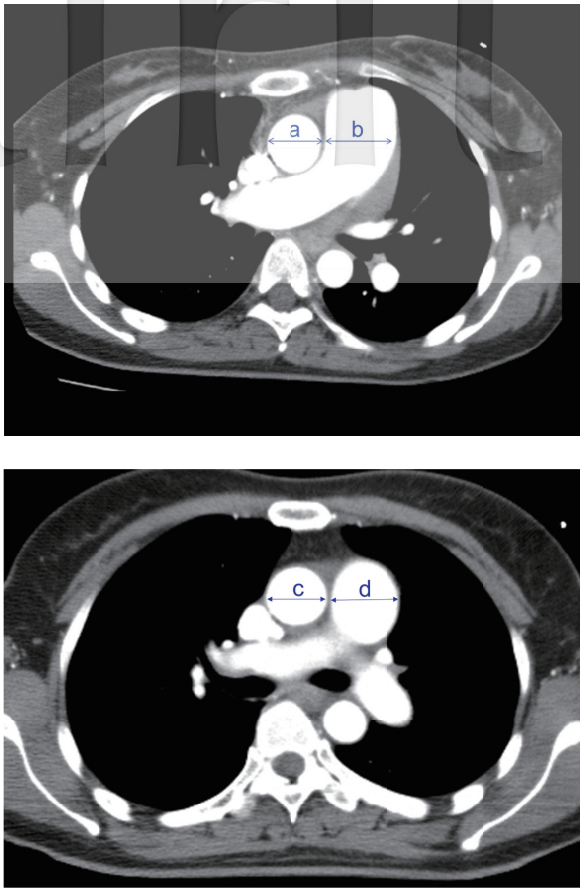

Fig. 3. Two levels of computed tomography (CT) angiograms showed enlarged pulmonary arteries, the pulmonary trunk diameter (b = 3.3 cm, d = 3.1 cm) is greater than that of ascending aorta (a = 2.6 cm, c = 2.7 cm).

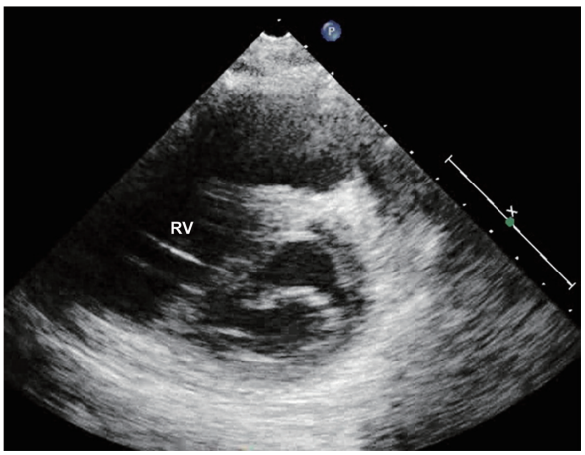

Fig. 4. Two-dimensional echocardiography showed enlarged right ventricle (RV), and D-shape of left ventricle.

ization. The patient received regular outpatient follow up after discharge and remained stable up to date.

Discussion

IPAH is a rare disease with a prevalence of 15 per million.[2] If left untreated, IPAH has a mean survival rate of 2.8 years from the time of diagnosis.[3] PH is confirmed when the mean pulmonary artery pressure is ≥ 25 mmHg at rest.[4] PAH is diagnosed when PCWP or left atrial pressure is < 15 mmHg in presence of PH. Our case fit the criteria of IPAH with following evidences: (1) PCWP 7 mmHg to exclude PH owing to left heart disease (i.e., group 2 PH); (2) chronic lung diseases and other causes of hypoxemia are excluded by history, pulmonary function test, polysomnography and computed tomography (CT) angiogram (to exclude group 3 PH); (3) pulmonary thromboembolic disease is excluded by CTA, and lung perfusion study (to exclude group 4 PH).

Doppler echocardiography is a recommended method to estimate PA pressure and to assess for right ventricular and RA enlargement, pericardial effusion, left ventricular systolic dysfunction, and valvular disease as part of the initial evaluation of a patient suspected of having PAH.[5] Echocardiography is a noninvasive, widely available, reproducible, and relatively inexpensive modality. Besides, the lack of radiation exposure makes it a valuable examination for serial follow-up studies in the PAH patients. The classic chest CTA findings of PH include dilated pulmonary arteries, dilated right heart, and the presence of thromboembolism in the main or peripheral pulmonary arteries.[4] In our case, the diameter of main PA (Fig. 3) was greater than the upper limit of control of Wisconsin Lung Transplant Group report (29 mm)[6] and the main PA diameter larger than that of the ascending aorta were consistent with the CT signs of PH.[7]

In summary, this report emphasizes that patients with PH often have vague symptoms at presentation. IPAH may take several months to diagnose. A high index of suspicion is crucial for the correct diagnosis of IPAH.

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