A RARE CASE OF DYSPNEA

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ABSTRACT

Idiopathic dilatation of the pulmonary artery defines the presence of a benign enlargement of the pulmonary trunk with or without the involvement of the rest of the arterial tree in the absence of a detectable cardiac or arterial systemic disorder. An IDPA patient’s symptoms and physical examination are not significant in diagnosis confirmation. Imaging techniques such as contrast-enhanced computed tomography, right heart catheterization, and angiocardiogram constitute the foundation of IDPA diagnosis. But an extended observation period is also necessary to exclude other possible dilatation explanation. Our patient, a forty-six-year-old female, was admitted to the Pulmonology clinic of “ShefqetNdroqi” University Hospital with dyspnea, dry cough, vertigo, and fatigue for several months before admission. The patient’s contrast-enhanced Computed Tomography revealed a dilatation of the pulmonary trunk. The dilatation of the pulmonary trunk was confirmed with the Cardiac CT, and other abnormalities were excluded.

1. INTRODUCTION

Idiopathic dilatation of the pulmonary artery (IDPA) defines the presence of a benign enlargement of the pulmonary trunk with or without the involvement of the rest of the arterial tree in the absence of a detectable cardiac or arterial systemic disorder. Greene et al. (1949) The diagnosis of idiopathic dilatation of the pulmonary artery relies on specific criteria, which were first proposed by Greene et al. (1949), and later improved upon by Deshmukh et al. (1960). The latter addition included the evaluation of the right ventricle and pulmonary artery pressure to exclude pulmonary hypertension as a cause of this enlargement.

Etiologically speaking, IDPA is related to a congenital arterial wall weakness, which leads to cystic degeneration of the pars media. Balboni and Lopresti (1961)
This is the most accepted theory related to the histopathology of IDPA, but more concrete evidence is necessary.

2. CASE REPORT

Our forty-six-year-old female patient was admitted to the Pulmonology clinic of “ShefqetNdroqi” University Hospital on May 11th, 2017. She complained of exertional dyspnea, dry cough, vertigo, and fatigue for several months before admission. Physical examination was unremarkable. The arterial blood gas examination revealed a level of oxygen saturation of 94%, with hypocapnia (paCO2 32.3 mmHg) and normal oxygen arterial pressure (paO2 65.9 mmHg).

The patient’s contrast-enhanced Computed Tomography revealed a diameter of the pulmonary trunk (44 mm), right pulmonary artery (33 mm), and left pulmonary artery (25 mm). Figure 1 No significant abnormality of the lung parenchyma was noted. In the abdominal cavity, the CT revealed hepatosplenomegaly Figure 2 and a lesion in the eighth hepatic segment (hemangioma). The portal vein diameter was estimated to be 16 mm, the spleen vein diameter 15 mm, and the superior mesenteric vein 11 mm.

Trans-esophageal echocardiography revealed an interatrial septal aneurysm with a minimal left to right shunt, without contrast displacement from the right side
to the left during the Valsalva maneuver. This examination had no detectable pulmonary artery dilation or significant transvalvular gradient or regurgitation.

Because of the inconsistent findings in these imaging examinations, further investigation was required, and the patient underwent cardiac computed tomography examination and right heart catheterization. The dilatation of the pulmonary trunk was confirmed with the Cardiac CT Figure 3 and Figure 4, and its diameter was estimated to be 41 mm. In contrast, the left and right pulmonary arteries remained intact, with a diameter of 21 and 26 mm, respectively.

**Figure 3**

[Image of Cardiac CT (Frontal)]

**Figure 4**

[Image of Cardiac CT (Lateral)]

Right heart catheterization denoted a wedge pressure of 15/7 mmHg, a pulmonary artery pressure of 21/9 mmHg, and a pulmonary trunk pressure of 23/11 mmHg.

### 3. DISCUSSIONS

IDPA symptoms include dyspnea on exertion, tachycardia, exhaustion, and chest pain. Malviya et al. (2017) These symptoms are not significant in diagnosis confirmation, because they are mainly related to the disease’s complications from compression of surrounding tissues leading to massive extension, thrombosis, or even rupture. Taussig (1947), Copkiran et al. (2014), Puri et al. (2011). Myocardial infarction has been described in cases of huge dilatation of pulmonary artery related
to the compression of coronary arteries and sudden cardiac arrest may also happen. Jurado-Román et al. (2013), Andrews et al. (1993). But in most cases, IDPA is an incidentaloma, and patients either exhibit mild non-specific symptoms or have no symptoms. This often leads to underdiagnosis and misdiagnosis of IDPA. Zhao and Cheng (1992).

Physical examination also does not play a significant role in the diagnostic process. In most cases, physical examination is interpreted as inconclusive. Van Buchem et al. (1955) Experts, however, recommend acknowledging IDPA as part of the differential in a patient with minimal symptoms and systolic, diastolic, or both cardiac murmurs. Kaplan et al. (1953).

Imaging techniques such as cardiac contrast-enhanced computed tomography constitute the foundation of IDPA diagnosis. It evaluates the artery enlargement’s exact location and morphological characteristics and provides information about possible concomitant abnormalities. Nguyen et al. (2007)

The diagnosis can be confirmed only by excluding other cardiovascular or systemic causes of the dilated pulmonary artery. This information can be obtained through additional imaging techniques such as heart catheterization and cardiac magnetic resonance imaging. Futagami et al. (1987), Ugolini et al. (1999).

Even though almost ten decades have passed since IDPA was first described as an anomaly Wessler and Jaches (1923), it remains a real challenge for clinicians because of its tendency to imitate cardiac disorders and the non-specificity of its symptoms. It represents an exclusion diagnosis and requires long observation periods as some underlying pathologies come into view as the disease progresses. Ring et al. proposed including the observation period in the diagnostic criteria. Ring, and Marshall (2002).

4. CONCLUSIONS AND RECOMMENDATIONS

Patients with IDPA have a wide range of clinical presentations. The symptoms might range from being unintentionally discovered to life-threatening manifestations typically connected to serious illness complications from the extension of the dilatation.

IDPA is a diagnosis by exclusion. It necessitates a good understanding of the diagnostic criteria and sharp observational skills from a multidisciplinary team, which includes pulmonologists, radiologists, and cardiologists.

Although imagery examination, such as contrast-enhanced cardiac CT or other advanced imaging techniques may give a detailed view of the structures involved in this anomaly, an extended observation period may be necessary to rule out any secondary causes of the dilatation.

CONFLICT OF INTERESTS

None.

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