Case Report

Idiopathic dilatation of pulmonary artery

Rahul Kumar Sharma, Deepak Talwar, Sameer K Gupta, Shobhit Bansal
Metro Centre for Respiratory Diseases, Metro Multispeciality Hospital, Noida, Uttar Pradesh, India

ABSTRACT

Idiopathic dilatation of pulmonary arteries (IDPA) is a rare abnormality of pulmonary arteries, the reported incidence in literature being as low as 0.007% in autopsy samples. With the improvement in diagnostic modalities, antemortem diagnosis of IDPA has been increasingly established by excluding diseases that induce pulmonary arterial enlargement. Here, we present a rare case of idiopathic dilatation of the pulmonary artery admitted with shortness of breath where IDPA was diagnosed as an incidental finding using computed tomography pulmonary angiography and cardiac catheterization.

KEY WORDS: Cardiac catheterization, idiopathic pulmonary artery dilatation, pulmonary hypertension

INTRODUCTION

Dilatation of pulmonary arteries is a well-known clinical entity commonly encountered in many congenital and acquired heart lesions. Idiopathic dilatation of pulmonary arteries (IDPA) was first recognized as a benign anomaly in 1923 by Wessler and Jaches.[1] The incidence of this disease is 6 cases per 1000 cases of congenital cardiac disorders.[2] Diagnosis of IDPA is made after excluding all possible causes that can induce pulmonary artery (PA) dilatation. It is usually an incidental finding not fully explained by the patient symptoms and clinical condition. Treatment is usually not required as it is considered to be a benign condition, although literature on long-term follow-up is lacking. We here report a case of symptomatic IDPA diagnosed on cardiac catheterization.

CASE REPORT

A 68-year-old female presented with insidious onset gradually progressive shortness of breath from mMRC Stage 1 to Stage 3 for the last 10 years, which further increased over the past 2 months. She had no associated history of a cough, fever, palpitations, chest pain, pedal edema, orthopnea, or paroxysmal nocturnal dyspnea.

On clinical examination, her vitals included pulse rate - 94/min, respiratory rate - 24/min, blood pressure - 126/80 mmHg, and O₂ saturation - 91% in room air. There was no pallor, icterus, cyanosis, pedal edema, or raised jugular venous pressure. Rest of the general physical and systemic examination was essentially normal.

Patient's investigations revealed hemoglobin - 12.8 g/dl, total leukocyte count - 5600/dL with a differential count of neutrophils - 72%, lymphocytes - 10%, eosinophils - 1%, and monocytes - 7%. Serum C-reactive protein level was normal (0.24 mg/dl) with elevated serum brain natriuretic peptide levels (162 pg/ml). Anti-nuclear antibodies along with anti-neutrophil cytoplasmic autoantibody levels and thyroid function tests were within normal limit, and there was no evidence of infectious diseases such as tuberculosis or syphilis.

X-ray chest revealed mild cardiomegaly of the right ventricular (RV) type with mild right atrial dilatation.

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and massive enlargement of the main PA with normal pulmonary vascular markings [Figure 1].

Echo-cardio-graphic findings revealed a normal-sized left ventricle and left atrium with 55% ejection fraction. RV was dilated with adequate systolic function, with dilated right atrium, mild tricuspid regurgitation, and mild PA hypertension (mean PA pressure 36 mmHg). There was no intra- and extra-cardiac shunt.

A computed tomography (CT) pulmonary angiography with high-resolution CT was done [Figures 1 and 2] which revealed dilated main PA with a maximum diameter of 49.75 mm, dilated right main trunk with a diameter of 37.76 mm, and dilated left trunk with a diameter of 36.96 mm. There was no filling defect ruling out pulmonary embolism. Lung parenchyma revealed areas of ground glass opacities. Pulmonary function test revealed restrictive airway defect with a normal diffusing capacity of carbon monoxide. She was made to walk for 6 min in which she covered 216 meters without desaturation but had moderate fatigue and severe breathlessness at the end of the test.

Cardiac catheterization was done through right femoral vein/right femoral artery approach. RV angiogram showed the adequate systolic function of RV with normal pulmonary valve and massively dilated the main PA. Hemodynamic data are given in Table 1.

On the basis of mild symptoms and pulmonary artery pressure (PAP) < 30 mmHg, a massive dilation of PA could not be explained so the diagnosis of IDPA was made after ruling out all possible causes.

**DISCUSSION**

IDPA is a rare disease with little known about its etiology and pathogenesis. Kaplan et al. postulated it as mal-development of the entire pulmonary tree and congenital weakness of the arterial wall whereas Assman thought of it as an unequal division of truncus arteriosus communis. In 1949, Greene et al. and associates established the following pathologic criteria for its diagnosis:

1. Simple dilatation of the pulmonary trunk with or without involvement of the rest of arterial tree
2. Absence of intra-cardiac or extracardiac shunts
3. Absence of chronic cardiac or pulmonary disease
4. Absence of arterial disease, such as syphilis, or more than minimal atheromatosis or arteriosclerosis of the pulmonary vascular tree.

In 1960, Deshmukh et al. presented report of 13 patients in whom right heart catheterization was performed aimed at elaborating its clinical and physiological aspects and to further draw attention to this anomaly. They opined that normal pressure in the

| Site                                | Pressure (mmHg) | Saturation (%) |
|-------------------------------------|-----------------|----------------|
| Right atrium                        | 10 (mean)       | 78             |
| Right ventricle                     | 35 (mean)       | 78             |
| Pulmonary artery                    | 35/25 (28)      | 79             |
| Pulmonary capillary wedge pressure  | 12              |                |
| Left ventricular end-diastolic pressure | 18              |                |

**Table 1: Results of right heart catheterization**

Figure 1: (a) Chest X-ray showing dilated pulmonary artery, (b) contrast-enhanced computed tomography chest showing enlarged main pulmonary artery (49.75 mm), (c) contrast-enhanced computed tomography chest showing dilated left pulmonary artery (36.96), (d) contrast-enhanced computed tomography chest showing dilated right pulmonary artery (37.76 mm), (e) contrast-enhanced computed tomography showing no evidence of pulmonary embolism, (f) computed tomography pulmonary artery reconstruction image showing dilated pulmonary artery

Figure 2: High-resolution computed tomography sections of lung showing ground glass opacities in lung parenchyma.
RV and PA should be introduced as the fifth criteria in the diagnosis.

Our case showed massive dilatation of PA with no apparent intra- or extra-cardiac shunt or any significant heart or lung disease. On right heart catheterization, PAP was only 28 mmHg which does not explain the massive dilatation of PA in our case thus confirming the diagnosis.

In spite of the well-marked clinical, radiologic and electrocardiographic features, the diagnosis of idiopathic dilatation of the PA is one of exclusion.

Cardiac catheterization is superior to echocardiography as a diagnostic tool. The absence of shunt, the normal relationship in cardiac chambers and normal oxygen saturation are important findings.

IDPA is considered a benign and nonprogressive condition. Long-term prognosis of IDPA is usually excellent compared to pathological dilatation of PA. However not much data on long-term follow-up of these patients exist. Management is also not well established, and any patient with an apparent diagnosis of idiopathic pulmonary hypertension needs to be followed up for a long time to know its benign course.

Our case adds to the existing literature and spreads physician awareness about this entity and its diagnosis, which should be established after ruling out all possible causes of pulmonary dilatation.

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Conflicts of interest
There are no conflicts of interest.

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