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IDIOPATHIC DILATATION OF PULMONARY ARTERIES AS A RARE CAUSE OF DYSPNEA

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Objective:

Introduction

Idiopathic dilatation of pulmonary arteries (IDPA) is an uncommon condition. The incidence is 0.6% in patients with congenital heart disease. IDPA has been recognised as clinical entity in 1923 by Wessler and Jaches. Even though this condition is well known for almost a century, not much is known about its etiopathology and pathophysiology. Diagnosis is set by excluding all congenital or acquired heart lessons that can lead to dilatation of pulmonary arteries.

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Case report

We report case of 67-year-old female patient who was admitted to our hospital in January of 2016 because of progressive dyspnea and chest pain. In 1985 she was operated dual arterial septal defect. Examination with CT pulmonary angiography (CTPA) and echocardiography showed heavy dilatation of truncus pulmonalis (in main diameter 58 cm), right and left pulmonary artery (33mm/40mm), as well as dilatation of right ventricular with no sign of hypertrophy. By right heart catheterization pulmonary hypertension (mPAP 24mmHg) was not confirmed. Radiocardiography showed left to right shunt of only 13% and lung perfusion scintigraphy did not show thromboembolic lungs. All immunological and infectious analysis were within normal range, as well as pulmonary function tests. She was diagnosed with IDPA. On re-evaluation in October of 2019 CTPA showed progressive truncus pulmonalis dilatation, in main diameter 64 cm. Based on her status, in compared to the previous one 2 years ago, she was diagnosed with NYHA Ⅱ and no evidence of pulmonary hypertension was established (mPAP 14mmHg). She is being ordered for follow-up in 6 months and her condition will be monitored.

Conclusion

Mean pulmonary arterial pressure of 14 mmHg shown by right ventricular catheterization and 13% of right to left shunt does not explain the massive dilatation of pulmonary arteries. In addition, there are no significant changes in status of this patient in last 4 years of follow-up thus this confirms the diagnosis of IDPA. Which is rare cause of dyspnea.

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