Incidentally detected right pulmonary artery agenesis with right coronary artery collateralization

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Abstract

Unilateral pulmonary artery agenesis (UPAA) with pulmonary hypoplasia is a rare congenital anomaly. We describe a 71-year old male who was incidentally diagnosed with the right UPAA and a hypoplastic right lung supplied by collateralized right coronary.

Keywords: Pulmonary artery agenesis • Coronary collateralization • Hypoplasia

INTRODUCTION

Unilateral pulmonary artery agenesis (UPAA) with pulmonary hypoplasia is a rare congenital anomaly caused by failure of the sixth aortic arch to connect to the pulmonary trunk. The underlying aetiology is not known. Prevalence of this abnormality is estimated to be approximately 1:200 000 in young adults. While patients may remain asymptomatic until adulthood, most present with a variety of complaints during childhood [1].

Among 108 cases of UPAA reported between 1978 and 2000, 14 were asymptomatic at the time of diagnosis. The median age of diagnosis was 14 years (range 0.1–58 years). Common symptoms at presentation included frequent pulmonary infections, dyspnoea or limited exercise tolerance, or haemoptysis. Pulmonary hypertension was present in 44% of the patients. Cases of high-altitude pulmonary oedema were also described [1].

UPAA was associated with other cardiovascular defects such as patent ductus arteriosus, atrial and ventricular septal defect, truncus arteriosus and tetralogy of Fallot. Most of these symptomatic patients required surgical correction [2].

CASE REPORT

A 71-year old male with a medical history of hypertension and dyslipidaemia was referred by his primary cardiologist for elective coronary angiography due to a 2-month history of recurrent retrosternal chest pain and dyspnoea on exertion. Prior to development of symptoms, he denied any chest pain, shortness of breath and other respiratory symptoms. He was living in a low socioeconomic community in a rural area. He had a sedentary lifestyle and did not participate in any sport, though he did not have any limitation in his daily activity.

Physical exam revealed normal vital signs. Cardiovascular examination revealed normal heart sounds without murmurs, rubs or gallops. The point of maximal impulse was not displaced, and there were no heaves or lifts. The jugular venous pressure was normal. Pulses were strong and symmetric in all extremities. There was mildly decreased air entry into the right lung and clear breath sounds were appreciated. An electrocardiogram revealed sinus rhythm with left ventricular (LV) hypertrophy. A chest film revealed a small right hemithorax and rightward shift of the mediastinum (Fig. 1). Coronary angiography demonstrated >75% left main artery stenosis (Fig. 2A). The right coronary artery was measured 3 mm in diameter and provided tortuous collateral circulation to the distal right pulmonary artery (Fig. 2B). Left ventricular systolic function...
was moderately decreased in angiography. Magnetic resonance imaging of the thorax confirmed the diagnosis of an absent right pulmonary artery and right lung hypoplasia. Echocardiography showed decreased LV ejection fraction with normal right ventricular size and function. The estimated pulmonary arterial systolic pressure was within normal limits. The patient underwent left main coronary artery bypass surgery. The congenital anomaly was not corrected.

**DISCUSSION**

To the best of our knowledge, there are only five case reports published describing UPAA and coronary collateral circulation supplying the affected lung in the setting of pulmonary artery agenesis [1–4]. Overall right UPAA is more common than left [5]. Reports of left pulmonary artery agenesis with left coronary artery collateral circulation are even scarcer [3]. Three of these five cases reported the absence of the right pulmonary artery with right coronary collateral arteries [1, 2, 4]. Most cases of UPAA presented with pulmonary symptoms like recurrent respiratory infections, haemoptysis or pulmonary hypertension. Only a few patients with UPAA remain asymptomatic [5]. In some cases, coronary collateral circulation and coronary steal syndrome were described. The shunt between coronary arteries and the pulmonary collaterals may result in steal phenomena which can cause myocardial hypoperfusion [1].

The case we report is significantly older at presentation than any of those previously reported and without pulmonary or cardiovascular symptoms related to this anomaly [2, 5]. The patient was incidentally diagnosed with right UPAA after developing critical left main coronary artery stenosis. Although no physiological test was performed to rule out ischaemia in the right coronary artery territory, it is a non-dominant artery and therefore unlikely to have contributed to the patient’s symptoms. It is likely that oxygen demand of the right ventricular myocardium supplied by the right coronary artery is met despite the shunting of blood flow to the pulmonary circulation.

In conclusion, we describe the oldest known presentation of coronary artery collateralization of a hypoplastic lung due to UPAA. The collateral vessel origination from a non-dominant coronary artery explains why the patient was asymptomatic throughout his life.

**Conflict of interest:** none declared.

**REFERENCES**


