An impressive image of unilateral pulmonary artery agenesis associated with coronary collateralization in an adult

Introduction

Isolated unilateral absence of pulmonary artery (UAPA) is a very rare congenital anomaly in adults. The prevalence of this anomaly is about 1 in 200,000 live births. It is usually diagnosed
in early childhood and the average age of patients with the condition is 14 years (0–58 years) (1, 2). Clinical symptoms of the patient, anatomical variation of the pulmonary artery (PA), and additional cardiovascular anomalies together with their associated collateral circulation and pulmonary hypertension (PHT) are important factors to consider in the choice of treatment (3).

**Case Report**

A 63-year-old female was referred to our clinic for evaluation of progressively-worsening dyspnea over the last 1 year by a pulmonology clinic. We learned that she had exercise-induced dyspnea for 20 years. She was also hypertensive and had been diagnosed with atrial fibrillation for the past 2 years. Her physical examination and laboratory findings were within the normal range. An elevated right hemidiaphragm with hypoplasia of the right lung and a shift of the heart to the right side was seen on a routine chest X-ray. Her electrocardiogram revealed atrial fibrillation with normal ventricular rate response. Echocardiography revealed normal left ventricular systolic function and a normal left atrial diameter. We detected an increase in the systolic PA pressure (PAP=60 mm Hg) with normal right ventricular function. Lung scintigraphy showed a heterogeneous and decreased perfusion in the right hemithorax compared to the left side. We decided to perform computed tomographic pulmonary angiography (CTPA), since the chest X-ray was abnormal, to exclude the chronic thromboembolic pulmonary hypertension (CTEPH). CTPA revealed an absence of the right PA and dilated left and main PAs. Magnetic resonance angiography of the aorta and PA, we confirmed the absence of the right PA and its branches, together with any additional anomalies. We performed right heart catheterization and coronary angiography to exclude coronary artery disease. The PAP as measured was 55/25 mm Hg with a mean value of 36 mm Hg and the pulmonary capillary wedge pressure was 15 mm Hg. Pulmonary angiography also showed an absence of the right PA with a dilated left PA and the vasoreactivity test was negative. Coronary angiography revealed normal coronary arteries, although we detected advanced collaterals arising from the conus branch of the right coronary artery, supplying the right PA’s branches (Figs. 1, 2, Videos 1, 2). The patient was discharged with sildenafil and oral anticoagulant therapy. At the three-month follow-up, there was a significant improvement in the clinical condition. We did not replace her therapy with an endothelin receptor antagonist or riociguat.

**Discussion**

Cardiovascular anomalies such as septal defects, Fallot’s tetralogy, patent ductus arteriosus, and right aortic arch may accompany UAPA (4). The isolated absence of the right PA is more common than that of the left (63%). Right-sided UAPA is less associated with other cardiac anomalies and patients with this condition may survive to an advanced age (2). According to the literature, hemoptysis was present in 20%, recurrent pulmonary infections in 37%, limited exercise tolerance in 40%, and PHT in...
44% of patients with isolated UAPA. The condition also affects both sexes equally (2). Echocardiography is useful for monitoring the right heart function and the development of PHT. Pulmonary angiography is the gold standard for the diagnosis of pulmonary angiography agenesis and the evaluation of collateral circulations (5). Another method that may be used in the differential diagnosis is magnetic resonance imaging, which is very valuable in terms of clearly showing the underlying etiologies and comorbidities if it can be done routinely. Mortality is usually associated with progressive PHT and the resulting right ventricular failure, respiratory failure, and massive hemoptysis. The overall mortality rate is approximately 7% (3). Although the treatment is unclear, the development of PHT may be the only indication for treatment (3). Swyer–James–Macleod syndrome, lobar atelectasis, post lobectomy status, and chronic pulmonary thromboembolism are the main differential diagnoses (6). On the other hand, pathologies that may cause deterioration of the clinical condition (acute pulmonary embolism, CTEPH, anemia, etc.) should not be ignored in such patients who have been asymptomatic for many years. When there is the presence of coronary-artery-to-pulmonary or -bronchial artery collaterals such as in our case, CTEPH should be kept in mind as a differential diagnosis. However, our case is important as, it shows that a congenital anomaly may be present in advanced age, like in the case of our patient. Kepez et al. (7) demonstrated that the rate of occurrence of coronary to PA collateral circulation was 18.1%, especially in patients with total occlusions at the level of the main pulmonary arteries (7). UAPA with coronary collaterals is a very rare anomaly, and many cases of collateral circulating are linked with RCA (8).

In a case report similar to ours with coronary-to-pulmonary collateral circulation, endothelin receptor antagonists (bosentan) for PHT were started by the authors and they mentioned this therapy is effective (9). We also showed that phosphodiesterase V inhibitors may be effective over a short period. However, considering the mechanism of development of PHT in this patient group, riociguat, which is indicated for groups 1 and 4 PHT, may be considered as the first-line treatment. One of the interesting aspects of our patient is that she is at advanced age and is similar to cases of Group 2 PHT phenotypically due to her accompanying additional comorbidities. Another interesting aspect is that her case suggests CTEPH in Group 4 PHT due to risk factors such as age, hypertension, and atrial fibrillation.

**Conclusion**

UAPA should be kept in mind before closing coronary-to-pulmonary-artery collaterals via surgical or percutaneous interventions.

**References**

1. Bouros D, Pare P, Panagou P, Tsintiris K, Siafakas N. The varied manifestation of pulmonary artery agenesis in adulthood. Chest 1995; 108: 670-6.
2. Ten Harkel AD, Blom NA, Ottenkamp J. Isolated unilateral absence of a pulmonary artery: a case report and review of the literature. Chest 2002; 122: 1471-7.
3. Kruzlik P, Syamasundar RP, Novak M, Pechanova O, Kovacova G. Unilateral absence of pulmonary artery: pathophysiology, symptoms, diagnosis and current treatment. Arch Cardiovasc Dis 2013; 106: 448-54.
4. Currarino G, Williams B. Causes of congenital unilateral pulmonary hypoplasia: a study of 33 cases. Pediatr Radiol 1985; 15: 15-24.
5. Griffin N, Mansfield L, Redmond KC, Dusmet M, Goldstraw P, Mittal TK, et al. Imaging features of isolated unilateral pulmonary artery agenesis presenting in adulthood: a review of four cases. Clin Radiol 2007; 62: 238-44.
6. Muthusami P, Ananthakrishnan R, Elangovan S. Incidentally detected unilateral pulmonary artery agenesis with pulmonary hypoplasia in a 67 year old woman. J Radiol Case Rep 2010; 4: 32-7.
7. Kepez A, Mutlu B, Paudel A, Ileri C, Atas H, Yildizeli B. Prevalence of Coronary Artery to Pulmonary Artery Collaterals in Patients with Chronic Thromboembolic Pulmonary Hypertension: Retrospective Analysis from a Single Center. Thorac Cardiovasc Surg 2018; 66: 180-6.
8. Darwazah AK, Alhaddad IA. Pulmonary artery agenesis associated with coronary collaterals among adults. J Cardiothorac Surg 2016; 11: 109.
9. Ghanbari H, Feldman D, David S, Saba S. Unilateral absence of a left pulmonary artery: successful therapeutic response to a combination of bosentan and warfarin. Circ Cardiovasc Imaging 2009; 2: e46-8.

**Address for Correspondence:** Dr. Yakup Alsancak, Necmettin Erbakan Universitesi, Meram Tıp Fakültesi, Kardiyojıloji Anabilim Dalı, Konya- Türkiye
Phone: +90 506 910 14 04
E-mail: dryakupalsancak@gmail.com

**Informed consent:** Written informed consent to publication was obtained from the patient.

**Video 1-2.** Right coronary angiography demonstrates a normal right coronary artery with collaterals from the conus branch to the right lung tissue.