Congenital absence of left pulmonary artery with collateralization from all major coronary arteries

Tüm majör koroner arterlerden collateralizasyonun olduğu doğuştan sol pulmoner arter yokluğu

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Summary– Unilateral pulmonary artery agenesis (UPA) is a rarely-observed congenital anomaly with diverse clinical signs and symptoms. It is most often diagnosed in childhood due to the high mortality rate prior to adulthood. This report describes the case of a 71-year-old woman who presented with exertional chest pain, dyspnea and palpitations and who was diagnosed as having left pulmonary artery (LPA) agenesis, hypoplastic left lung supplied by collateralization from each of the three major normal coronary arteries, and atrial fibrillation.

Unilateral pulmonary artery agenesis (UPA) is a very rare congenital malformation which may present as recurrent pulmonary infections and hemoptysis. Most UPA patients develop pulmonary hypertension (PHT). Due to the high mortality rate in childhood, patients surviving to adulthood are rarely encountered. However, as in the present case, because of atypical symptoms, some patients are diagnosed at an older age.

This report presents a 71-year-old female patient with left pulmonary artery (LPA) agenesis and an angiographically documented collateralization from each of the three major coronary arteries causing angina pectoris via coronary steal phenomenon.

CASE REPORT

A 71-year-old female patient was admitted with symptoms of exertional dyspnea, angina and palpitations. Past history was unremarkable. Physical examination showed irregular beats and a 2/6 pansystolic murmur audible at apical focus. Electrocardiography revealed atrial fibrillation with a ventricular rate of 118 beats per minute (Figure 1). Transthoracic echocardiography showed normal systolic function of the left ventricle, moderate mitral and tricuspid regurgitation with a systolic pulmonary artery pressure (sPAB) of 75 mmHg. Also, left atrial dilatation (57 mm) was detected. Coronary angiography revealed collateralization from the proximal segments of each of the three coronary arteries: the left anterior descending (LAD) artery, the circumflex (Cx) artery and the right coronary artery (RCA), into the left hypoplastic lung without any stenosis in epicardial coronary arteries (Figure 2). In order to define the exact anatomic structure of the heart, magnetic resonance angiography (MRA)

Abbreviations:
CT Computed tomography
LPA Left pulmonary artery
MRA Magnetic resonance angiography
PHT Pulmonary hypertension
UPA Unilateral pulmonary artery agenesis
was planned. LPA agenesis (Figure 3a) was detected on MRA. To define the coronary to pulmonary collateralization, computed tomography angiography was performed and showed LPA agenesis, enlarged left and right atria, and a dilated right pulmonary artery (Figure 3b).

As the patient was symptomatic and the collateralization hemodynamically significant, the patient was evaluated by a heart team for surgical intervention. However, as the MRA revealed LPA agenesis, with distal pulmonary flow being maintained by the coronary arteries via the collateralization, it was decided that surgical intervention was not a suitable option for the patient. Therefore, outpatient follow-up was scheduled with full oral medical treatment.

**DISCUSSION**

Unilateral absence of one pulmonary artery is a very rare congenital abnormality with an incidence estimated to be 1:200,000.\(^1\) It results from failure of the
The affected site is most commonly the right pulmonary artery, and is twice as common as LPA agenesis. Bockeria et al. reported that the right pulmonary artery was absent in 60% of cases among 182 cases with UPA. It has no sex predilection.

Approximately 15% of patients remain asymptomatic. Recurrent pulmonary infections, reduced exercise tolerance and exertional dyspnea are the presenting symptoms in 40% of patients. In some cases, coronary collateral circulation and coronary steal syndrome have been described. The shunt between coronary arteries and pulmonary collaterals may cause this syndrome, which may lead to myocardial hypoperfusion and thus ischemia, as in the present case. Myocardial ischemia results from large and hemodynamically significant collaterals. Coronary to bronchial collateralization is also a rare anomaly. The association of a UPA and coronary-to-bronchial artery collateralization has been described before. One reports a case complicated with coronary steal and myocardial ischemia in an elderly patient, as in the present case. Atrial fibrillation caused by an enlarged left atrium due to excess amount of blood flow from the collateral arteries is another rare complication of UPA. There are no cases in the literature regarding the development of atrial fibrillation as a result of coronary to bronchial collateralization. Another complication of this anomaly is PHT, which is observed in 25% of cases and develops due to increased blood flow to the unaffected pulmonary artery. PHT is important due to its effect on long-term survival of patients.

Chest radiography and echocardiography are helpful diagnostic tools in the diagnosis of UPA. However, anatomic details for definitive diagnosis and presence of associated anomalies can be clarified by contrast-enhanced computed tomography (CT) and MRA. In the present case, echocardiography revealed high systolic pulmonary artery pressure and an enlarged left atrium. MRA confirmed the diagnosis of absent LPA. CT angiography showed dilated right pulmonary artery and left atrium in addition to a hypoplastic left lung.

There is no consensus on treatment of UPA. The therapeutic approach should be based on symptoms, anatomy of the pulmonary artery, related cardiovascular anomalies and aortopulmonary collaterals, and PHT. When PHT is present in a patient, revascularisation can improve the patient’s condition. If revascularisation of the side with the absent artery, LPA in the present case, is not possible, therapeutic measures such as pulmonary vasodilators may be utilized. The patient in this case was put on phosphodiesterase inhibitors and oral anticoagulant as well as antianginal medication.

Our case is different in some aspects from previous cases regarding UPA. First of all, the patient was significantly older. Secondly, there was left pulmonary agenesis, which is the rare form of UPA, and coronary artery collateralization was originating from proximal segments of all three major coronary arteries. Thirdly, our patient had atrial fibrillation, pulmonary hypertension and angina pectoris, all of which were due to the LPA agenesis. Embolisation of collateral flow would have decreased blood flow to the left lung, so it was not a suitable option in this case. The patient was discharged with full oral medical treatment including phosphodiesterase inhibitors, oral anti-coagulant and anti-anginal medications and advised for follow-ups.

Although unilateral absence of pulmonary artery is a rare disorder, it may be observed in older patients. Clinicians should be alert concerning this rare condition, especially in patients with pulmonary hypertension and left atrial dilatation in echocardiography, in addition to angina pectoris and normal coronary arteries with collateral circulation to pulmonary vascular beds.

Consent

Written informed consent was obtained from the patient for the publication of this case report.

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REFERENCES


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