Unilateral pulmonary artery agenesis (UPAA) is a rare condition. It is caused by an embryogenic malformation of the 6th aortic arch of the affected side (1,2). The first case was reported in 1868 and the estimated prevalence is 1/200.000 people (3-5). Right-sided UPAA is twice as common as the left-sided (6). However, cardiac malformation is more common in the left sided agenesis (5). When cardiac malformation coexists with UPAA, the patients are more symptomatic and diagnosed during childhood (7). The most common symptoms in UPAA are recurrent pulmonary infections, hemoptysis, and dyspnea (6). If the agenesis is isolated, the symptoms may be minor or absent, the diagnosis can be made in the adulthood (5). A study by Bouros D. et al reported that 30% of the adult patients with UPAA haven’t any symptoms (5). In this study, 75% of the left-sided UPAA was associated with congenital cardiac anomalies, whereas right-sided agenesis was not associated with any cardiac malformation (5). The most common associated cardiovascular abnormalities are tetralogy of Fallot, septal defects, patent ductus arteriosus, coarctation of the aorta and transposition of great vessels (4). Mitral valve prolapse (MVP) association with UPAA is very rare. In the literature, there are only two cases (5,8). To the best of our knowledge, it is the third reported right-sided UPAA case with coexisting MVP. Here, we present a rare case of right-sided UPAA and severe MVP diagnosed at adult age.

A 43-year-old man was referred to pulmonary medicine outpatient clinic for investigation of dyspnea after the prosthetic ring valvuloplasti. He was on anti-hypertensive treatment for a year. One month prior to his routine evaluation for cardiology, he reported exertional dyspnea and orthopnea. He was non-smoker. Had no other systemic symptoms. On the physical examination he had 4/6 systolic murmur at mitral area. In the echocardiographic evaluation, there was grade 3-4 mitral regurgitation, mitral valve prolapse, and chordal rupture. Systolic pulmonary arterial pressure was 50 mmHg, the left ventricular ejection fraction was 65%. The coronary angiography was normal.
A rare case of right sided pulmonary artery agenesis associated with congenital mitral valve prolapse

The ventriculography showed severe mitral regurgitation and the patient was decided to be operated to repair mitral valve. There was mediastinal shift to the right hemithorax, the left lung volume and the cardiothoracic index was increased at chest X-Ray (Figure 1). Chordal repair and prosthetic ring valvuloplasty was done surgically. There was no postoperative complication. Mitral valve ring was functioning. There was not any mitral regurgitation after the operation. Because of ongoing exertional dyspnea and abnormal chest radiography, the patient was evaluated for pulmonary diseases. The pulmonary function test was as follows: FVC = 3870 mL (95% of predicted), FEV₁ = 2900 mL (86% of predicted), FEV₁/FVC = 74.9%. CT revealed hypoplastic right lung, compensatory enlargement of the left lung, cardiomegaly and absent right main pulmonary artery (Figure 2). The right lung showed only minimal paranchymal perfusion activity at the superior region (Figure 3). This might be due to superposition of the left lung herniation across the midline.

The diagnosis of UPAA is based on chest X-Ray, CT, pulmonary ventilation-perfusion scintigraphy, and echocardiography. The gold standard for the diagnosis is pulmonary angiography. However, in recent years the angiography is made only preoperatively and in cases to make selective bronchial artery embolisation for hemoptysis (5). Our patient had characteristic findings on the chest X-Ray consisting of the following:

a) ipsilateral cardiac and mediastinal displacement;

b) mediastinal shift to the right hemithorax, increased left lung volume and cardiothoracic index.

c) hypoplastic right lung, compensatory enlargement of the left lung, cardiomegaly and absent right main pulmonary artery.

d) minimal paranchymal perfusion activity at the superior region of the right lung area, and normal perfusion of the left lung. This minimal paranchymal activity at the right might be due to superposition of the left lung herniation across the midline.

Figure 1. Plain chest X-Ray shows mediastinal shift to the right hemithorax, increased left lung volume and cardiothoracic index.

Figure 2. Thoracic computed tomography revealed hypoplastic right lung, compensatory enlargement of the left lung, cardiomegaly and absent right main pulmonary artery.

Figure 3. Ventilation-perfusion lung scan showed only minimal paranchymal perfusion activity at the superior region of the right lung area, and normal perfusion of the left lung. This minimal paranchymal activity at the right might be due to superposition of the left lung herniation across the midline.
b) smaller hemithorax; c) absent pulmonary artery shadow; and d) contralateral lung hyperinflation and herniation beyond the midline. Thorax CT was also supporting the chest X-ray findings. Ventilation-perfusion scan showed absent perfusion in the right lung. The possible diagnosis was UPAA in this patient with the combination of symptoms and characteristic findings on CT and ventilation-perfusion lung scan.

Congenital UPAA was commonly coexists with structural cardiac defects (4). However, severe MVP has been documented only in two cases as being associated with UPAA (5,8). MVP is the displacement of mitral valve leaflet into the left atrium during systole. The pathogenesis of this rare malformation is unknown. The prevalence is estimated at 2-3% of the population (9,10). Prolapsed mitral valves are classified as classic, non classic (10). Nonclassic form of MVP carries a low risk of complications and it has generally benign prognosis. In severe cases of classic MVP, complications include mitral regurgitation, infective endocarditis and congestive heart failure. Persistent pulmonary hypertension (PH) is reported in 46% of the patients after the mitral valve operation (10). The most common finding associated with UPAA is also PH and it defines the survival (1). In cases of severe cardiac malformations or PH, revascularization can be a treatment option. Asymptomatic and mildly symptomatic patients do not usually need any treatment for UPAA. Our case had a classic MVP and he was asymptomatic until the formation of severe mitral regurgitation caused by chordal rupture. He had mild dyspnea on exertion and it did not relieve completely after mitral valvuloplasty. Although he had UPAA and severe mitral valve regurgitation, PH disappeared after mitral valvuloplasty. So we thought that PH was related to valvular disease, and it was not related to UPAA.

In conclusion, UPAA is a rare congenital disease. It may be asymptomatic and remain undiagnosed until advanced age. It should be suspected in patients having congenital cardiac anomalies. Radiologic examination with a high index of suspicion may help clinician for diagnosis of UPAA.

REFERENCES