The Heart Surgery Forum #2014-446
18 (3), 2015 [Epub June 2015]
doi: 10.1532/hsf.1311

Online address: http://journal.hsforum.com

Mitral Valve Replacement in an Adult with Left Pulmonary Agenesis

Huseyin Saskin, MD,[1] Mustafa Idiz, MD,[2] Cagri Duzyol, MD,[1] Huseyin Macika, MD,[3] Rezan Aksoy, MD[1]

[1]Clinic of Cardiovascular Surgery, Derince Education and Research Hospital; [2]Cardiovacular Surgery Clinic and [3]Anesthesiology and Reanimation Clinic, Acibadem Hospital, Kocaeli, Turkey

INTRODUCTION

Pulmonary agenesis is associated with the absence of pulmonary vessels, bronchi, or parenchyma. This condition usually occurs between the 4th and 5th week of gestation during the embryonic phase. Etiopathogenic factors associated with pulmonary agenesis are not fully understood. In the literature, genetic and teratogenic factors, viral infections, and vitamin-A deficiency are shown to be associated with pulmonary agenesis [Malcon 2012]. This condition may be seen unilaterally or bilaterally. Although the precise rate of incidence is unknown, it is estimated to occur in one of every 10,000 to 12,000 live births [Yetim 2011]. There is a 1.3:1 female predominance with unilateral agenesis [Halilbasic 2013].

This condition is usually diagnosed in the first few years of life. However, it may rarely be diagnosed in adulthood. Pulmonary agenesis may be isolated or associated with other abnormalities (cardiovascular, skeletal, gastrointestinal, genitourinary, etc.) with a rate of 50% [Rappaport 1994]. Recurrent symptoms of pulmonary infections and dyspnea are commonly observed in cases of pulmonary agenesis. However, some patients may remain asymptomatic until adulthood [Gunbey 2014]. The clinical findings may vary according to the presence or severity of other accompanying congenital abnormalities. Prognosis in right pulmonary agenesis is usually worse than in left [Canıtez 2013].

Posteroanterior chest X-ray reveals contralateral deviation of mediastinum, ipsilateral narrowing of intercostal spaces, and ipsilateral elevation of hemidiaphragm. Computerized tomography (CT) with intravenous contrast or magnetic resonance imaging (MRI), bronchography, or angiography may be used for diagnosis [Deniz 2004].

Total atelectasia, congenital hernia of the diaphragm, congenital cystic adenomatoid malformation, pulmonary sequestration, chylothorax, bronchogenic cyst, and tumors may be considered for differential diagnosis [Pahuja 2013].

In this paper, we present a case of an adult patient who was coincidentally diagnosed with left pulmonary agenesis in the preoperative period, who was undergoing mitral valve replacement under cardiopulmonary bypass due to severe mitral valve regurgitation.

Received November 4, 2014; received in revised form February 9, 2014; accepted April 24, 2015.

Correspondence: Huseyin Saskin, MD, Derince Education and Research Hospital, Clinic of Cardiovascular Surgery, Kocaeli, Turkey; (e-mail: sueda_hs@yahoo.com).

CASE REPORT

A twenty-four-year-old male patient was referred to the cardiology outpatient clinic with a complaint of dyspnea and fatigue for one year. Transthoracic echocardiography revealed enlarged left ventricular sizes with an end-systolic diameter of 4.4 cm, end-diastolic diameter of 6.4 cm, and an ejection fraction of 58%. The mitral valve was found to have a myxomatous degeneration with posterior leaflet prolapse for 0.5 cm into the left atrium during systole. Severe eccentric mitral regurgitation through the interatrial septum was also observed. The estimated pulmonary arterial pressure was 38 mmHg. Based on these findings the patient was referred to our clinic for operation.

The patient had a complaint of dyspnea and wheezing during effort, becoming intense particularly in cold weather. He had no history of smoking. Compared with the right, the left thorax wall was found to be relatively depleted during physical examination. Respiratory sounds were reduced during auscultation on the upper, middle, and basal areas of the left lung when compared with the right. Systolic regurgitation of 5/6 severity on the mitral focus extending to the left axillary area were auscultated. Heart rate was 70 bpm and arterial blood pressure was 120/70 mmHg. Electrocardiography revealed normal sinus rhythm and findings of left ventricular hypertrophy. The examinations of other systems were normal.

During the preparation period for operation, the patient's whole blood count and routine biochemical tests were within normal range. There was a homogenous density increase filling the left hemithorax and total loss of aeration of the left

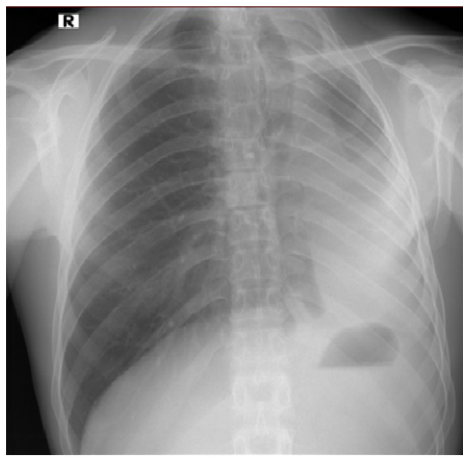

Figure 1. Preoperative posteroanterior X-ray image.

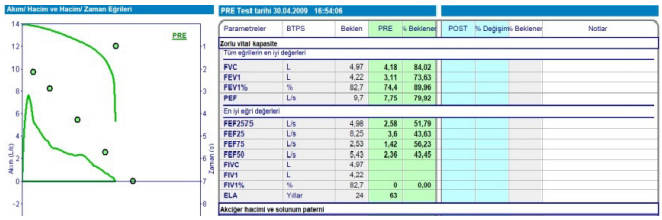

Figure 2. Preoperative respiratory function test.

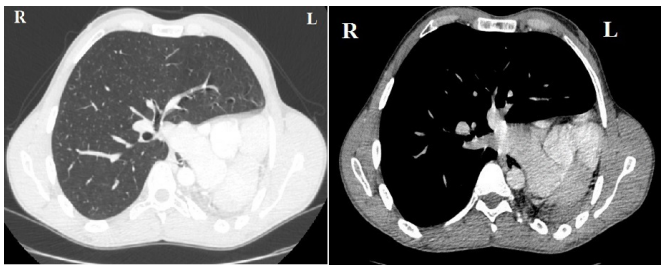

Figure 3. Preoperative CT with intravenous contrast (axial cross section).

lung seen in the posteroanterior X-ray imaging. The right lung was significantly hyperplastic where the heart and other mediastinal vascular structures were significantly deviated to the left (left lung agenesis or severe hypogenesis) (Figure 1).

The respiratory function test was performed with spirometry which was consistent with loss of volume in the left lung, showing FVC: 4.18 L (Pred 84.02%), FEV1: 3.11 L (Pred 73.63%), FEV1/FVC: 74.4% (Pred 89.96%) (Figure 2). The thorax CT with intravenous contrast showed the absence of the left lung parenchyma, the left bronchus, and left lung vascular structures (agenesis). Therefore, the trachea, mediastinal structures, and heart were markedly deviated to the left and there was a compensatory hyperplasia of the right lung (Figures 3 and 4). Mitral valve replacement operation was planned.

The mediastinum was reached with median sternotomy under general anesthesia. The right lung was found to be totally filling the anterior portion of the mediastinum. When the pleura of the left lung was opened, the left hemithorax was found to be completely empty and the heart and other mediastinal structures were deviated to the left hemithorax. The pericardium was opened and lifted. Cardiopulmonary bypass (CPB) was initiated following aortic and bicaval cannulation. Systemic hypothermia of 28°C was used. An aortic cross clamp was placed. Cardiac arrest was obtained by antegrade blood cardioplegia. The mitral valve was visualized by left atriotomy. The posterior mitral leaflet was prolapsed. The mitral valve was totally excised and a bileaflet mechanical mitral valve (29 mm SJM Master; St. Paul, MN, USA) was implanted to its position with one by one pledgeted sutures. The left atriotomy was closed. The aortic cross clamp was removed. CPB was terminated following stable hemodynamics. The operation was ended following placement of one mediastinal and two thoracic drains. The aortic cross clamp duration was 62 minutes and total CPB duration was

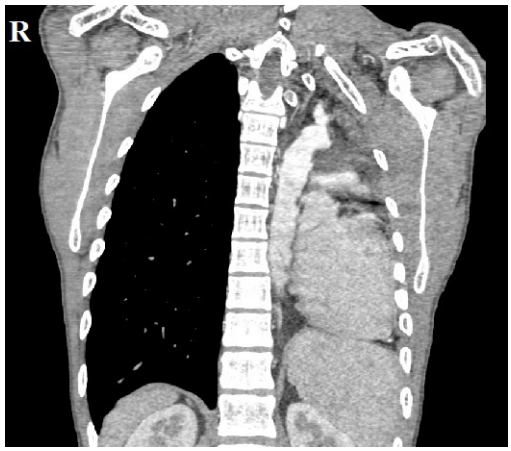

Figure 4. Preoperative thorax CT with intravenous contrast (coronal cross section).

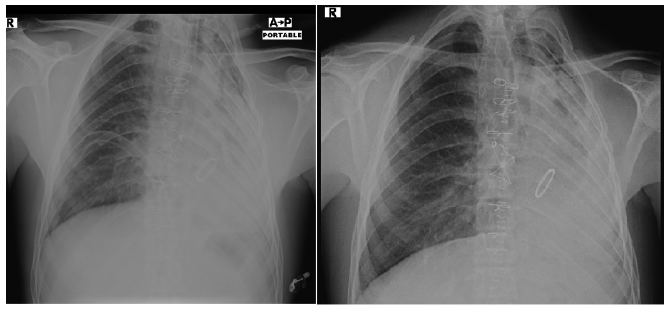

Figure 5. Posteroanterior thorax X-ray image in the 1st and 12th postoperative days.

83 minutes. No blood products were used during operation. The patient, who had normal blood gas analysis and no drainage, was extubated on the 2nd postoperative hour. There were no findings other than the presence of the artificial mitral valve and drains on the posteroanterior chest X-ray image in the first postoperative day (Figure 5). The patient, who had stable hemodynamics without an inotropic support and had a total drainage of 300 mL, was taken to the inpatient room following removal of the drains. No respiratory or cardiac problems occurred in the follow-up period. Following proper adjustment of prothrombin time with oral anticoagulant, the patient was discharged on postoperative day five. The skin sutures of the patient were removed on postoperative day 12 and he has been followed up without any problems since then.

DISCUSSION

Pulmonary agenesis is one of the rare congenital abnormalities that may be associated with cardiovascular, gastrointestinal, neuro-musculo-skeletal, and genitourinary malformations for approximately 50% [Rappaport 1994]. Pulmonary agenesis is the absence of lung vessels, bronchi, and parenchyma.

The pathology for pulmonary agenesis has been categorized with the following classification:

1. Agenesis-complete absence of a lung and bronchi, with no blood supply to the effected side;
2. Aplasia-presence of a rudimentary bronchus with complete absence of parenchyma;
3. Hypoplasia-lobar agenesis and hypoplastic lung [Malcon 2012].

Pulmonary agenesis may be seen unilaterally or bilaterally. Although the precise rate of incidence is unknown, it is estimated to occur in one of every 10,000 to 12,000 live births [Yetim 2011]. Although the etiopathogenic factors are not clearly understood, genetic and teratogenic factors, viral infections, and vitamin-A deficiency may play a role [Malcon 2012]. There was no history of antenatal viral infections, vitamin-A insufficiency, or exposition to teratogenic factors in our case.

The clinical presentation may vary between individuals; while some of them may have signs of pathology in the neonatal period, some may be asymptomatic until adulthood and may be diagnosed coincidentally [Gunbey 2014]. Our case was asymptomatic until the adulthood period and the diagnosis was made coincidentally.

Cardiovascular (primarily patent ductus arteriosus and foramen ovale), gastrointestinal (tracheoesophageal fistula and duodenal atresia), skeletal (hemi vertebra, absence of costa and radius), genitourinary (horseshoe kidney), and facial abnormalities were described in patients with unilateral pulmonary agenesis. The prognosis is usually related to accompanying cardiovascular, gastrointestinal, and genitourinary system abnormalities [Canıtez 2013]. The prognosis of right pulmonary agenesis is usually worse than the left, regarding the coexisting cardiac abnormalities, and distortions of great vessels, trachea, and bronchi due to cardiac shifting [Ootaki 2004].

The diagnosis of pulmonary agenesis is mainly based on imaging methods. Loss of aeration on the side of agenesis and enlargement on the healthy side due to hyperaeration can be seen in chest X-ray imaging and thorax CT. Displacement of the heart and midline structures secondary to hyperplasia of the healthy lung, and loss of visualization of vessels and bronchi on the agenetic side are helpful findings for diagnosis. Echocardiography for detection of accompanying cardiac abnormalities, CT-angiography, and magnetic resonance imaging for visualization of other vascular malformations, and bronchoscopy for confirmation of the diagnosis may also be used [Deniz 2004].

For differential diagnosis of cases detected in the neonatal period, congenital hernia of the diaphragm, congenital cystic adenomatoid malformation, pulmonary sequestration, chylothorax, pulmonary hypoplasia, bronchogenic cyst, and tumors such as neuroblastoma, fibrosarcoma, and teratoma should be discussed [Pahuja 2013].

As a result, patients with unilateral pulmonary agenesis, like our case, may be asymptomatic for a long period of time if there are no coexisting abnormalities. The diagnosis may be coincidental in adulthood for these patients. As in our case, CPB may be planned for patients with the diagnosis of pulmonary agenesis if the respiratory functions are adequate. Mitral valve replacement with CPB can be safely performed with collaborative work of anesthesia and cardiovascular surgery teams. Assistance of a pulmonologist and respiratory physiotherapist should be obtained to minimize pulmonary complications. In such cases in which respiratory parameters are well in the early postoperative period, patients should be mobilized early and pulmonary rehabilitation should be done following extubation.

REFERENCES

Canıtez Y, Cekic S, Gurpınar A, Sapan N. 2013. Right lung agenesis; isolated and with accompanied anomalies. J Curr Pediatr 11:134-7.

Deniz O, Tozkoparan E, Ciftci F, et al. 2004. Left pulmonary aplasia (agenesis): case report. Gulhane Med J 46:56-8.

Gunbey HP, Gunbey E, Sayit AT, Bulut T. 2014. Unilateral right pulmonary agenesis in adulthood. J Clin Diag Res 8:RD01-RD02.

Halilbasic A, Skokic F, Hotic N, et al. 2013. Unilateral pulmonary agenesis and gastric duplication cyst: a rare association. Case reports in Pediatrics. Article ID 608706:1-3.

Malcon MC, Malcon CM, Cavada MN, Caruso PEM, Real LF. 2012. Unilateral pulmonary agenesis: Case report. J Bras Pneumol 38:526-9.

Ootaki Y, Yamaguchi M, Yoshimura N, Oka S. 2004. Pulmonary agenesis with congenital heart disease. Pediatr Cardiol 25:145-8.

Pahuja A, Lee S. 2013. Persistent unilateral white-out in preterm infant. J Paediatr Child Health 49:E87-9.

Rappaport DC, Herman SJ, Weisbord GL. 1994. Congenital bronchopulmonary diseases in adults: CT findings. AJR Am Roentgenol 162:1295-9.

Yetim TD, Bayaroğullari H, Yalcin HP, Arıca V, Arıca SG. 2011. Congenital agenesis of the left lung: a rare case. J Clin Imaging Sci 1:47.