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Unilateral Absence of Main Branch of Pulmonary Artery

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Abstract

Background: Unilateral absence of a pulmonary artery (UAPA) is an uncommon anomaly with reported incidence of 1 in 200,000 young adults and a diverse clinical presentation. It may be diagnosed as an isolated congenital anomaly or in combination with other heart defects. The present study aimed to investigate the clinical course and characteristics of 40 patients with UAPA the diagnosis of whom was first established in childhood or adulthood.

Materials and Methods: From January 2002 to April 2021, 40 patients with UAPA were diagnosed at our institution. The medical records of these patients were retrospectively reviewed in detail, and information covering the patients' demographics, associated anomalies, imaging studies, and operation data were elicited.

Results: According to the results, all 40 patients were between 2 months and 25 years old (mean age: 6.45 years) at the first presentation, weighing 4.70-59.00 kg (mean weight: 19.17 kg). Regarding gender, 25 (62.5%) patients were female. Left sided UAPA was found in 60% (n=24) of cases, and 40% of patients had occult right pulmonary artery (RPA). Moreover, a right-sided aortic arch was found in 4 (10%) patients. Tetralogy of Fallot was diagnosed in 16 patients, whereas complex cardiac anomalies were found in 13 cases.

Conclusion: Clinicians should consider the undiagnosed cases of UAPA, especially in adults with suspicious symptoms, such as unexplained hemoptysis. The application of different available imaging techniques for early diagnosis and treatment of isolated UAPA cases is important to prevent the devastating complications of pulmonary hypertension and massive hemoptysis in the long term.

Keywords: Symptoms, Surgical treatment, Unilateral absence of pulmonary artery

Introduction

The absence of unilateral or intra-pericardial branch pulmonary arteries (UAPA) initially reported by Frentzel in 1868 is an extremely rare entity with a wide spectrum of presenting symptoms that may be diagnosed as an isolated cardiac anomaly or in combination with other heart defects (1, 2). It is usually diagnosed in childhood; however, in isolated cases, it may be incidentally found in adult patients. The reported incidence of UAPA is estimated at 1 in 200,000 population with only 12% being diagnosed in infancy (3, 4).

It is hypothesized that a UAPA is caused by the growth alteration of the proximal sixth aortic arch with interruption of connection to the pulmonary trunk during embryogenesis, and in two-thirds of patients, the RPA affected the contrary side of the aortic arch (5-7).

It may occur in isolation; nevertheless, it is frequently associated with a variety of congenital heart anomalies, such as Tetralogy of Fallot (TOF), TOF with atresia of pulmonary artery, transposition of great arteries with pulmonary atresia, and truncus arteriosus (2). Approximately, 30% of cases with UAPA have isolated form with no associated cardiovascular anomalies, and most of them present with mild clinical courses and symptoms, followed by survival into adulthood. Conversely, patients with associated cardiac abnormalities are diagnosed in infancy or childhood due to related symptoms (8). Although the early clinical course is seemingly benign, the majority of untreated cases will lead to pulmonary hypertension, followed by pulmonary infections and hemoptysis (9).

The diagnosis of UAPA needs a high clinical suspicion and can subsequently be established using pulmonary angiography, as the gold standard, or non-invasive imaging modalities, such as ventilation-perfusion scintigraphy, which shows the absent perfusion on the affected side, computed tomography (CT) scan, or magnetic resonance imaging (MRI) (10,11). Although surgical reconstruction of the affected pulmonary artery may be technically possible, the satisfying outcome is not always guaranteed due to the pathological changes in the parenchyma of the affected lung (12).

This report presents 40 patients of UAPA with a diverse clinical presentation the diagnosis of whom was first established in childhood or adulthood.

Materials and Methods

From January 2002 to April 2021, 40 patients with UAPA were diagnosed at our institution. The medical records of these patients were

this series were those with unilateral agenesis of one lung. The diagnosis was based on history, clinical examinations, and imaging studies, including chest X-ray, trans-thoracic echocardiography, CT scan, ventilation-perfusion lung scan, MRI, and digital subtraction angiography (DSA). In all cases, at least one DSA examination was performed, most of cases having serial studies (Figures 1, 2). On imaging studies, native and alternative sources of circulation created by surgery to the affected pulmonary parenchyma were also evaluated. Patients presenting with dyspnea and cyanosis were immediately treated with oxygen therapy, diuretics, and anti-pulmonary hypertension crisis depending on their hemodynamic stability. The final purpose of surgical treatment was to find the absent pulmonary artery branch and its reconstruction by connecting it to the main PA or the systemic circulation. Retrospective data were analyzed by expert list reviewers using standard data-sheaths. The information included the patients' demographics, coexisting anomalies, as well as imaging and operation data. All the data were collected retrospectively and the IBM SPSS (version 21) was used for statistical analyses.

retrospectively reviewed in detail, and information covering the patients' demographics, associated anomalies, imaging studies, and operation data were elicited. Excluded patients from

Results

All 40 patients were between 2 months and 25 years old (mean age: 6.45 years) at first presentation, weighing 4.70-59.00 kg (mean weight: 19.17 kg). Regarding gender, 25 (62.5%) patients were female. All patients had tachypnea at the time of presentation, and PH with symptoms and signs of right-sided heart failure was present in 10 (25%) patients. The mean pulmonary artery systolic pressure of these patients was obtained at 55 (range: 42-75) mmHg. The absence of LPA was found in 60% (n=24) of patients, and 40% of cases had occult right pulmonary artery. Furthermore, right-sided aortic arch was detected in 4 (10%) patients, and the TOF was diagnosed in 16 patients, whereas complex cardiac anomalies were found in 13 cases. Associated cardiac anomalies are presented in Table 1. In total, 19 (47.5%) patients underwent at least one surgical operation. Treatment strategies of patients are described in Table 2. During the mean follow-up of 8.20 years, 52.5% of patients survived, 5 cases expired, and 14 patients missed the follow-up.

Table 1: Associated cardiac anomalies of patients with UAPA

Anomaly	No (%)
Complex anomaly	13 (32.5%)
ASD	5 (12.5%)
PAPVC	2 (5%)
Hetrotaxia	7 (17.5%)
TOF	16 (40%)
PS	3 (7.5%)

PAPVC: Partial anomalous pulmonary venous connection, ASD: Atrial septal defect, TOF: Tetralogy of Fallot, PS: Pulmonary stenosis

Table 2: Treatment strategy of patients with UAPA

Variable	No (%)
Medical	18 (45%)
Surgery	19 (47.5%)
Intervention	1 (2.5%)
Surgery and intervention	2 (5%)

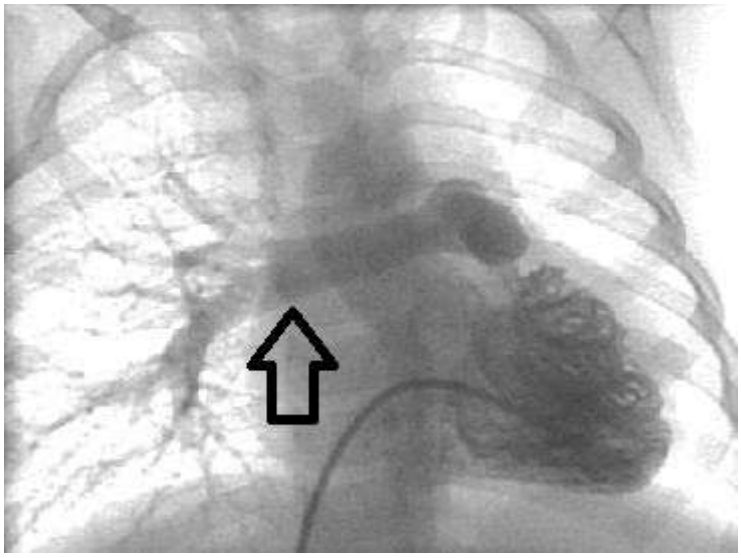

Figure 1: Cardiac catheterization showing normal RPA (arrow) and absence of LPA

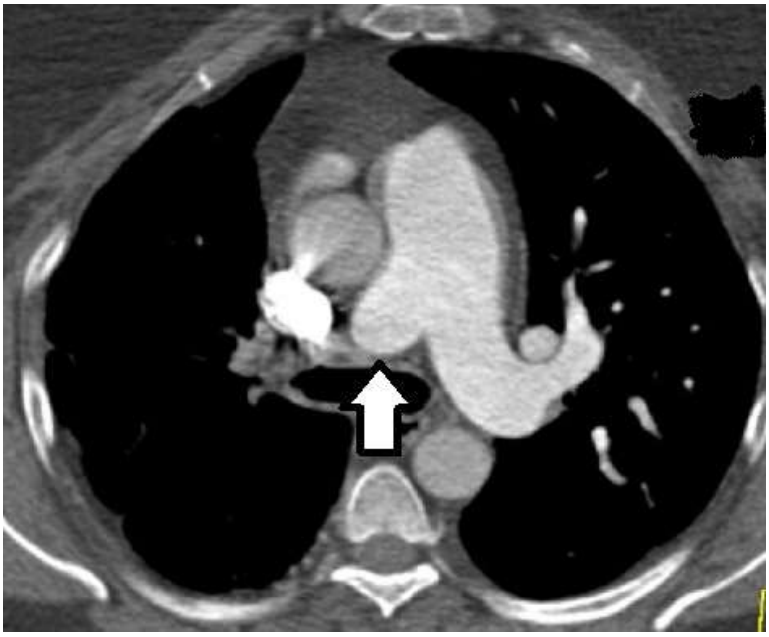

Figure 2: CT angiogram showing absent RPA (white arrow), normal pulmonary trunk, and normal LPA

Discussion

Congenital UAPA was first diagnosed in 1868 and is a rare anomaly that is described as the complete agenesis of the intra-pericardial segment of one of the pulmonary artery branches. So far, 420 UAPA cases have been reported in the literature (13, 14). The major embryologic explanation of UAPA results from early deterioration of the sixth aortic arch which consequently ends in the involution of the proximal branch of the PA and instead of the persistent relation of ductus arteriosus or contributions from systemic collaterals and bronchial arteries to the hilar pulmonary (9,15). Congenital absence of right PA is more common, while left-sided UAPA is commonly associated with complex cardiovascular anomalies (16).

Despite the fact, the prevalence of left-sided UAPA in our case series was 60%, and the diagnosis was set at the mean age of 6.5 years, whereas the overall median age of diagnosis is 14 years (17). The mortality rate of UAPA is about 7%, and both pulmonary hypertension and hemoptysis negatively affect long-term patients' survival (18). The most common causes of mortality include respiratory failure, right heart failure, massive hemoptysis, and pulmonary edema (6, 18). The distal vascular bed of

the affected is supplied by ductus arteriosus or abnormal collaterals branches from bronchial, intercostals, subclavian, as well as coronary and sub-diaphragmatic arteries (18). When the PDA gradually closes, the blood supply of the affected pulmonary artery will be decreased, diminish in size, and cannot be detected by echocardiographic or angiographic studies (9).

Pulmonary hypertension in UAPA occurs as a result of remodeling of pulmonary arterial vasculature due to high flow circulation to the other lung with an increase in the injury to the intimal endothelium. A similar event happens in some patients after pneumonectomy, who present PH (10, 19). Ten Harkel showed that PH was present in 44%, limited exercise tolerance in 40%, recurrent pulmonary infections in 37%, and hemoptysis in 20% of patients with untreated isolated UAPA which determines patients' long-term survival (6).

The initial diagnosis of isolated UAPA is usually suspected on a plain chest X-ray with a small size of the involved lung, compensatory hyperinflation of the contralateral lung and mediastinal shift, elevated ipsilateral hemidiaphragm, and difference of the parenchymal vascular markings in the two sides (12). Although ventilation-perfusion mismatch in pulmonary perfusion scintigraphy is useful for the diagnosis of UAPA, the differentiation among pulmonary artery branch stenosis, thrombotic occlusion, and pulmonary artery agenesis is not possible by this imaging study. When suspicious presentations are found on a chest X-ray, contrast-enhanced CT scan or magnetic resonance angiography can definitively make the diagnosis of UAPA by demonstrating the absence of left or right PA branch terminating within 2 cm of its predictable starting point from the main pulmonary artery and different bronchial or systemic collateral arteries (8, 20).

Definitive diagnosis is established by pulmonary angiography or digital subtraction angiography as the gold standard showing the complete absence of one of the main PA branches. Injection of the entire aorta is mandatory to identify the blood supply of the affected lung and find collateral circulation originating from the aorta which also helps to perform interventions, such as revascularization or embolization (21).

Treatment of isolated UAPA depends on the patient's age and clinical conditions and can vary from medical treatment to restoration of physiologic pulmonary circulation in selected pediatric cases or endovascular embolization of collateral circulation in adults, ending with surgical pneumonectomy in cases with no response to the previously performed therapies (22). Therefore, treatment is indicated in symptomatic cases, especially with recurrent or massive hemoptysis due to devastating effects on long-term survival (6).

Early surgical reconstruction of an isolated UAPA restores the physiologic pulmonary circulation, normal distal growth of pulmonary vascular bed, and regression of pulmonary hypertension (9). Several techniques have been described for the reconstruction of absent PA branch, including main PA flap, use of interposition graft, anastomosis of a saphenous vein graft or pericardial roll, and homograft interposition (2). However, the majority of these techniques have limited long-term durability since there is no potential for conduit growth.

Conclusion

In conclusion, clinicians should bear in mind the undiagnosed cases of UAPA, especially in adults with suspicious symptoms, such as unexplained hemoptysis. The application of different available imaging techniques for early diagnosis and treatment of isolated UAPA cases is important to prevent the devastating complications of pulmonary hypertension and massive hemoptysis in the long term.

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