Right Pulmonary Artery to Left Atrial Fistula Confirmed by 320-slice Computerized Tomography

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To the Editor: Right pulmonary artery-left atrial fistula (RPALAF) is an extremely rare congenital cardiovascular malformation that may be difficult to diagnose by clinical symptoms. High resolution computed tomographic (CT) gives a clear view of cardiovascular and can provide an accurate diagnosis, and it has not been reported in the last literature date though 320-slice CT scan giving these clear views. On the base of a case of this disease, we describe an 18-year-old girl with RPALAF confirmed by 320-slice CT. This case demonstrates that the 320-slice CT scan is a sensitive and reliable technique for establishing the diagnosis of RPALAF when it cannot be visualized by clinical symptoms.

An 18-year-old adolescent girl was hospitalized because of progressive cyanosis of lips and skin, and shortness of breath after daily exertion and had no hemoptysis, squatting phenomenon, seizures, fever, night sweats, weight loss, or chest pain. Moreover, she had no family history of cardiac problems or hypertension. Physical examination disclosed that she had cyanosis of the lips and limbs with clubbing of the fingers. Heart examination showed that the girl's heart rate was regular at 82 beats/min, and her breath sounded clear, with a respiratory rate of 21 breaths per minute. The cardiac examination exhibited a slight systolic murmur (grade 2/6) was auscultated in right parasternal surface. Liver and spleen were not palpable, peripheral pulses were symmetric. And resting oxygen saturation was 80% in room air. An electrocardiogram revealed left atrial enlargement. Chest radiography showed increased pulmonary vascularity with left atrial enlargement. An echo Doppler study showed situs solitus, normal pulmonary and systemic venous drainage, and normal cardiac chambers and valves. Transthoracic echocardiography examination showed immediate and dense opacification of the left atrial, suggesting the RPALAF communication [Figure 1]. However, it was hard to make a firm diagnosis. Then a 320-slice CT scan was performed, which revealed a large communication as the shape of ">" between the right pulmonary artery to the left atrium [Figure 2]. The upper branch originated from ostiums of right pulmonary artery and the lower branch drained into the left atrium directly. The bending part of this anomaly was enlarged as round shapes [Figure 3]. And there were no branches proceeded from this abnormal vessel. All pulmonary veins are normal that return directly to the left atrial, and RPALAF with type I was diagnosed. Unfortunately, the patient rejected the subsequent surgery because of personal reasons.

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RPALAF is a direct connection between branches of the right pulmonary artery and left atrium through a thin-walled aneurysm. They act as direct right-to-left shunts, resulting in central cyanosis, clubbing, exceptional dyspnea, and polycythemia, when the shunt is large. In addition, because the RPALAF bypasses the capillary bed, the lung loses its filtering function, thus allowing emboli and bacteria to pass directly into the systemic circulation. These would result in some complications, such as stroke, cerebral abscess, cerebral and systemic emboli, rupture of the fistula, and so on. [1] RPALAF has been classified into four types. [2] Type I is communication with normal veins, type II is aneurysm with abnormalities of lobulation and absence of pulmonary veins, type III is all veins draining into the communication, and type IV

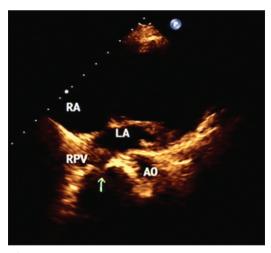


Figure 1: Transthoracic echocardiography examination showing an immediate and dense opacification of the left atrial.

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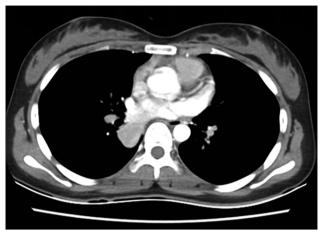


Figure 2: Horizontal plane of 320-slice computed tomographic angiography scan showing an abnormal fistula.

is aneurysm with the right veins connecting with it. Since the first case which was operated and described 60 years ago, only a few cases of RPALAF have been reported around the world. Hence, it was difficult to diagnose by clinical symptoms. High-resolution CT gives a clear view of cardiovascular details and can provide an accurate diagnosis, and it has not been reported in the last literature date though 320-slice CT scan giving these clear views. On basis of this, we reported this case of RPALAF confirmed by 320-slice CT, which obtained a clear view of cardiovascular malformation details. The patient's all pulmonary veins are normal that return directly to the left atrial, and was classified into type I RPALAF.

The traditional treatment is close the abnormal communication with surgical ligation by median sternotomy. However, there are high risk, huge trauma and lots of complications, particularly systemic embolism, infective endarteritis, cerebral abscess, and rupture of aneurysmal communications, with this operation. ^[5] In recent years, transcatheter coil occlusion of RPALAF was successfully performed in many cases and appears to be a safe and effective alternative to surgical treatment. ^[1] Unfortunately,

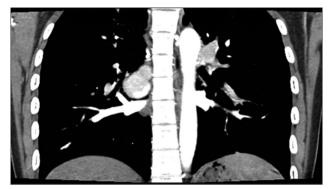


Figure 3: Frontal plane of 320-slice computed tomographic angiography scan showing an aneurysmal communication.

the patient rejected the subsequent surgery (because of personal reasons) even though we are confident and have rich experience to use intervention technology closing the abnormal communication.

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Conflicts of interest

There are no conflicts of interest.

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