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Surgical Treatment of Total Anomalous Pulmonary Venous Drainage with Atrial Fibrillation in an Adult: A Case Report and Literature Review

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ABSTRACT

Total anomalous pulmonary venous drainage (TAPVD) is caused by the absence of the congenital connection between the pulmonary vein and left atrium. This causes blood drainage into the right atrium, resulting in poor development of the left atrium and increasing the burden for the right atrium. It is accompanied by an atrial septal defect. TAPVD mostly is diagnosed during the fetal period and rarely is reported in adults. Atrial fibrillation (AF), a common arrhythmia, originates primarily from the joint of the pulmonary vein and left atrium, whereas AF originating from the right atrium has not been documented. Herein, we report the case of a 45-year-old male diagnosed with TAPVD accompanied by AF. After the correction of TAPVD and radiofrequency ablation (RFA) for AF performed under general anesthesia and cardiopulmonary bypass, the patient returned to normal sinus rhythm and showed no AF recurrence during two years of follow up.

INTRODUCTION

Total anomalous pulmonary venous drainage (TAPVD) is a rare congenital heart disease that accounts for approximately 1-3% of all congenital heart diseases [Hossein 2019; Yanqiang 2019; Yong 2017]. It can be divided into supra-cardiac, intra-cardiac, infra-cardiac, and mixed [Czekajska-Chehab 2018]. Few patients with TAPVD survive to adulthood. Most had cyanosis, dyspnea, congestive heart failure and other symptoms, which were associated with a worse prognosis [Yong 2019]. Atrial fibrillation is the most common arrhythmia in clinic. It is mainly caused by the left atrium, but rarely caused by the right atrium [Seungyup 2020; Fukamizu 2012]. Herein, we report the case of a 45-year-old male patient with TAPVD accompanied by AF, and we review past literature.

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CASE REPORT

The patient, a 45-year-old male weighing 37 kg, was admitted to the hospital with "20+ years of cardiac murmur and 1 month of chest distress and shortness of breath." Compared with his peers, the patient had low exercise capacity and body mass. He had bilateral lower extremity edema not accompanied by paroxysmal nocturnal dyspnea. Physical examination at admission revealed cyanosis of the lips and clubbing of fingers and toes. No changes were heard in both lungs, and there was a III/6 systolic murmur in the intercoastal space between ribs 3 and 4, increased P2 and fixed splitting, heart rate of 78 bpm, absolute arrhythmia, and unequal intensity of heart sounds. Electrocardiograph identified atrial fibrillation. Ultrasonic cardiogram revealed supracardiac total anomalous pulmonary venous drainage, huge atrial septal defect (35mm × 40mm), right-to-left shunt, pulmonary artery markedly widened (approximately 40 mm in diameter), severe pulmonary hypertension (75 mmHg), poor development of the left atrium (the left atrial dimension was 34mm) and left ventricle (the left ventricular end-diastolic dimension was 37mm), and thin ascending aorta (approximately 17 mm in diameter). The mitral valve Z score was about -1.9. Computed tomography angiography documented the left and right pulmonary veins afflux into the left innominate vein through vertical vein after the formation of a common pulmonary venous trunk in the rear of the left atrium, the superior vena cava presented a tumor-like dilation, a significant widening of the main pulmonary artery as well as the left and right pulmonary artery, a small inner diameter of the aorta, and severe enlargement of heart shadow. Supracardiac TAPVC was considered. (Figure A) The right-sided heart catheterization showed the mean pulmonary artery pressure was 47 mmHg, pulmonary arterial wedge pressure was 22 mmHg, and pulmonary vascular resistance was 4.36 wood units. At admission, the patient was diagnosed with congenital heart disease, supracardiac TAPVC, atrial septal defect, AF, pulmonary arterial hypertension, and level III cardiac function.

In May 2018, the patient was given TAPVD correction and RFA for AF under low-temperature (28°C) cardiopulmonary bypass under general anesthesia. The surgery revealed significant thickening of the innominate vein, superior vena cava, and pulmonary artery, hypertrophy of the right atrium and right ventricle, and thin aorta. The diameter of the pulmonary artery was found to be 2.5 times larger than that of the aorta. (Figure B) After the establishment of cardiopulmonary bypass, blocking the ascending aorta, and perfusion

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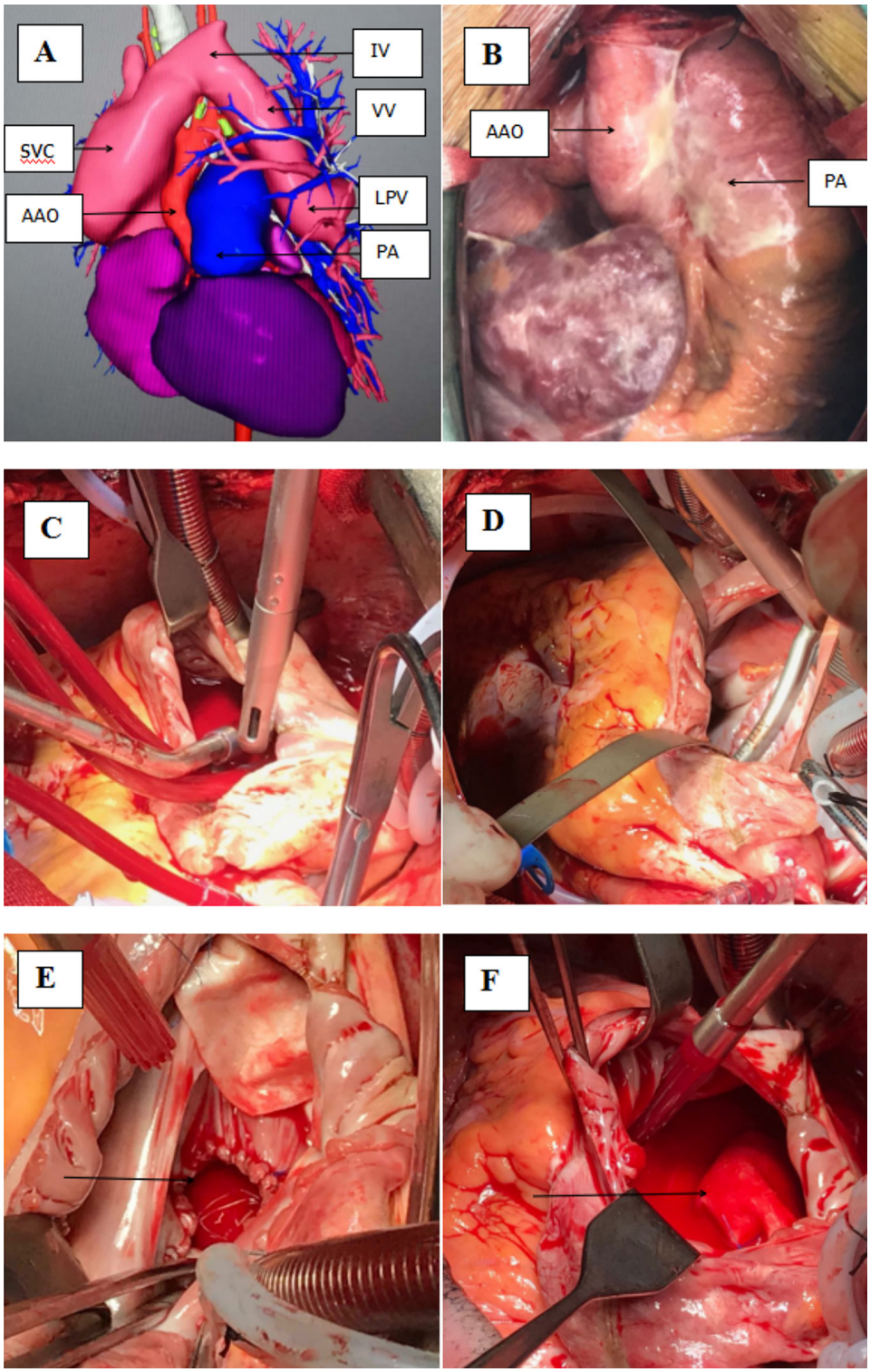

Fig. A: Three-dimensional reconstruction of heart structure; Fig. B: the diameter of the pulmonary artery seen after sternotomy was approximately 3 times larger than that of the aorta; Fig. C: a huge atrial septal defect was found after the incision of the right atrium, and no opening of the pulmonary vein was found in the left atrium; Fig. D: ablation with AtriCure bipolar radiofrequency ablation forceps; Fig. E: the common pulmonary venous trunk was dissected and continuously anastomosed to the wall of the left atrium with a 4-0 Prolene suture (arrow shows the anastomotic); Fig. F: pericardial patch with a 0.5 cm opening in the center was used to repair the atrial septal defect (arrow shows the repaired atrial septum).

of the myocardium with Thomas II myocardial protective solution, the right atrium was dissected. A huge atrial septal defect borderless with superior vena cava and inferior vena cava, and the absence of the opening of the pulmonary vein in the left atrium were identified. (Figure C) Upon turning the heart over, a common venous trunk in the rear of the left atrium was found. The dissection of the left pleura revealed the vertical vein originating from the left and right pulmonary venous trunk and connecting to the left innominate vein. Subsequently, an ablation with AtriCure bipolar radiofrequency ablation forceps was performed in the anterior and posterior part of the right auricle, the middle part of the right atrium to the inferior vena cava isthmus of the right atrium. Through the large size of the huge atrial septal defect, part of the posterior wall of the left atrium was removed to create an anastomotic that was wider than the opening of the mitral valve. Ablation was carried out from the right atrium to the right lateral margin of the incision of the dissected left atrium, then from the left lateral margin of incision in the left atrium to the left auricle (Figure D), and finally in the left auricle. (Figure D) The common pulmonary venous trunk was dissected and continuously anastomosed to the wall of the left atrium with a 4-0 Prolene suture. (Figure E) To avoid a pulmonary hypertensive crisis, the atrial septal defect was repaired using a bovine pericardial patch with a 0.5 cm opening in the center. (Figure F) Subsequently, the vertical vein was ligated, and the incision of the right atrium was closed. After the de-airing of the left and right heart, the ascending aorta was opened, and the heart resumed an autonomous sinus rhythm. Aortic root continued to de-airing until the esophagus ultrasound indicated that there was no air in the heart. With stable circulation, the extracorporeal circulation machine was withdrawn, and surgery was concluded routinely after closure of the chest.

The patient recovered smoothly from the operation and was discharged after 14 days. He was treated with amiodarone (0.2 g qd) for six months. During the two years of follow up, the patient did not show chest distress or shortness of breath, his cardiac function improved, cyanosis of the lips disappeared, and ECG documented normal sinus rhythm. No thrombosis or mass was found in the atria and ventricles.

The present case report was approved by the Ethics Committee of the Affiliated Hospital of North Sichuan Medical College, and written informed consent was obtained from the patient for publication of this case report and the accompanying images.

Literature review: The search for "total anomalous pulmonary venous drainage" and "atrial fibrillation" in the Pubmed and Medline databases did not identify any reported cases of surgical treatment of TAPVD with AF.

DISCUSSION

The incidence of TAPVD was very low and had no self-healing, and operations were the only effective method [Yong 2017]. Among patients with untreated TAPVD, 80% die from congestive heart failure before reaching one year of age, and very few survive to adulthood [Yong 2020]. Thus, the patient reported here who was 45 years old is a very rare case. Currently, AF originating in the joint of pulmonary veins and left atrium is the accepted version, and the main reason is considered to be the extension of the pulmonary veins myocardial sleeve to the left atrium [Ehrlich 2020; Lau 2019]. In the most simplified radiofrequency ablation for AF, a large box is established in four openings of pulmonary veins in the posterior wall of the left atrium, which can result in a 90% efficacy [Poynter 2010]. The right atrial isthmus is regarded as the primary site producing atrial flutter [Byounghyun 2020; Hołda 2020]. However, the discussed patient did not have a connection between the pulmonary veins and the left atrium with a significantly enlarged right atrium and a smaller left atrium. These findings implied that AF was generated entirely by the right atrium. A possible pathological basis of AF is the remodeling of myocardial structure and electrical remodeling of the right atrium caused by a long-term shunt and significantly enlarged right atrium in adults with congenital heart disease [Hugh 2020; Jean-Baptiste 2020].

The RFA strategies for TAPVD accompanied by AF may be different from other RFA. To prevent an excessive enlargement of the right atrium after the surgery, we performed left auricle ablation in the rear wall of the left atrium following the right atrial ablation, connected the common pulmonary venous trunk and incision in the posterior wall of the left atrium to create a complete ablation line, and divided the left atrium into the top and bottom parts to avoid the formation of an AF link.

In summary, we provide the first clinical evidence of AF caused by the right atrium and the method of RFA in a patient of TAPVD with AF. Since AF can be triggered in the right atrium alone, we suggest performing RFA in both atriums for the treatment of AF. However, we did not clarify the specific mechanism of AF caused by the right atrium, which needs further study in the future.

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