A Rare Case of Supracardiac Total Anomalous Pulmonary Venous Return

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Abstract

A 46-year-old female presented with a 3-month history of palpitations. Chest radiograph presented a 'calabash' configuration. Cardiac CT revealed supracardiac total anomalous pulmonary venous return, whereby all the pulmonary veins drain into vertical vein and finally to the superior vena cava. Cardiac catheterisation was consistent with anomalous pulmonary venous.

Introduction

Total anomalous pulmonary venous return (TAPVR) is a rare congenital heart defect. Anatomically, TAPVR caused by the failure of connection between left atrium and the common pulmonary vein, while resulting in persisting communication between the pulmonary and systemic veins (1). The incidence of TAPVR is 0.05 to 0.09 per 1000 live births accounting for only 1.5% of children with congenital heart disease. Patients with TAPVR are usually symptomatic at a very young age, and less than 7% of them with surgical correction can survive into adulthood (2). Herein, we reported a TAPVR adult patient underwent a successful surgical repair.

Case presentation

A 46-year-old woman presented to our hospital with a history of a palpitations and exertional dyspnea for 3 months. She was incidentally found to have a heart murmur during cough treatment at local hospital in December 2019. However, she did not look for further inspection. Palpitations correlated with atrial flutter on Holter monitoring. She was referred to our hospital for further cardiac assessment and management.

Physical examination showed a blood pressure of 105/68 mmHg in left arm, a heart rate of 106 bpm, and an oxygen saturation of 96%–98% under room air between the upper and lower limbs. Her jugular venous pulsations were normal. Auscultation revealed that her lungs were clean. Cardiac examination found a right ventricular heave, but no thrills. Cardiac auscultation revealed normal heart sounds, with no added sounds. There was no hepatomegaly, peripheral clubbing, or edema.

Investigations

Chest radiograph showed cardiac enlargement resembles a classic 'calabash' configuration (Figure 1A and 1B). A CT scan further presented anomalous pulmonary vein (PV) and to check any remaining PV drainage into the left atrium.

Three-dimensional images further delineated the supracardiac TAPVR in coronal view (Figure 2A) and sagittal view (Figure 2B). It found several pulmonary veins draining separately into the collecting vein. The collecting vein, located above the left atrium, ascended and formed a dilated vertical vein. The vertical vein drained into the superior vena cava (SVC) by a dilated left innominate vein was observed. Other CT findings

include no pulmonary venous obstruction, a large atrial septal defect (ASD), cardiomegaly with the dilated right ventricular forming the right lateral heart border.

Cardiac catheterisation revealed common drainage of pulmonary veins into a collecting vein. This vein ascended and formed a left innominate vein and finally drained into the SVC (Figure 2C and 2D). Haemodynamic data obtained as follows: aortic pressure 74/48mm Hg, main pulmonary artery pressure 28/9 (11) mm Hg, right ventricular (RV) pressure 11/4 (7) mm Hg. The elevated RV pressure with a pressure gradient of 24 mm Hg was also suggestive of pulmonary stenosis. Oxygen saturation from arterial blood gas was 96.0% under room air. The calculated pulmonary vascular resistance (PVR) was 0.87 wood unit.

The patient was referred for surgical repair. She underwent a successful surgical correction of TAPVC and ASD close. At procedure, she was diagnosed with supracardiac TAPVC and found excessively dilated SVC, innominate vein, and right heart chamber. The pulmonary artery was larger than the aorta (ratio=2.5:1). The large ASD was measured at 31mm × 22mm diameter and the vertical vein was at 22mm × 26 mm diameter. The LA was connected to the PV confluence through the anastomosis. A patch material has been used to close the large ASD. Patient was discharged well on the sixth postoperative day. No palpitations and exertional dyspnea during the half year fellow-up.

Discussion

TAPVC is a rare congenital heart defect. The incidence rate was 5 to 9 per 100 000 live births. The most common type of TAPVC is supracardiac TAPVC, accounting for approximately half of all TAPVC patients (Figure 3). Patients with TAPVC are usually symptomatic at a very young age, and less than 7% of them with surgical correction can survive into adulthood. The clinical presentation of TAPVC is mainly determined by the degree of pulmonary venous drainage obstruction and the magnitude of the left-to-right shunt (3, 4). Our patient lived with no symptoms until found at her 46-year-old because of palpitations and exertional dyspnea caused by atrial flutter.

In this case, this patient was particularly unusual in that she has been living without symptom until her nearly fifth decade of life without the need of medication. Two factors, the absence of the pulmonary venous obstruction and a large ASD, contribute to the long-time survival for this patient in supracardiac TAPVR. It explains the lack of cyanosis and normal blood pressure in this patient. Cardiac CT provides accurate anatomical delineation of pulmonary venous pathways and their connections (5). According to the CT scan of this patient, no unobstructed pulmonary venous circulation was observed. Cardiac catheterization showed no pulmonary over-circulation and a normal cardiac output. Cardiac CT scan and catheterization showed this patient had no pulmonary venous obstruction and a large ASD.

The classical 'calabash' configuration on the chest radiograph is uncommonly observed in supracardiac TAPVR. The SVC and right heart dilation formed right-sided configuration and the dilated innominate vein mainly consist of this left-sided configuration. There is also non-specific appearance of chest radiograph in some patients, and then used as such CT scan and echocardiography for accurate diagnosis of TAPVR. This patient was firstly reported as classical 'calabash' appearance on the chest radiograph. And was diagnosed by CT scan and echocardiography. Cardiac CT scan presented venous system enlargement but aortic system disuse atrophy. Cardiac hemodynamic assessment in patient showed no pulmonary hypertension and for further surgical repair.

Surgical repair is immediate need in most cases once the diagnosis of TAPVC is made. The surgical mortality is less than 5% when repair is performed in patients without obstructed pulmonary veins. This patient successfully underwent TAPVC surgical correction and closed ASD. Although surgery results in a normal circulation (the pulmonary veins returning normally to the left atrium), she appeared pulmonary edema because of aortic disuse atrophy and relatively excessive cardiac output. Several factors predict the poor outcomes such as pulmonary venous obstruction before repair, a younger age at repair, pulmonary vein size and univentricular heart (6). However, the long-term outcome after surgical repair of TAPVC is also excellent.

The authors declare no conflicts of interest.

Informed consent: Written informed consent was obtained from this patient.

Reference

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Figure legends:

1. **Figure 1.** Chest radiograph (A and B). A huge "calabash" heart presented as appearance according to chest radiograph. TAPVR=total anomalous pulmonary venous return.
2. **Figure 2.** A three-dimensional cardiac CT scan (A and B) and cardiac catheter angiogram (C and D) further delineated the supracardiac TAPVR. Pulmonary veins drained into a collecting vein. Later followed by left upper pulmonary vein to form an anomalous dilated vertical vein which eventually drains into left innominate vein and superior vena cava.
3. **Figure 3.** Images of normal heart and supracardiac TAPVR were presented.