Total Anomalous Pulmonary Vein Drainage in an Adult Diagnosed by Helical Computed Tomography

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A 41-year-old man visited our hospital with the complaint of palpitation by atrial flutter. He was finally diagnosed as total anomalous pulmonary vein drainage by helical computed tomography (CT). This case is very unusual due to the lack of symptoms until the age of 41. The absence of pulmonary artery stenosis, and the presence of atrial septal defect providing sufficient right to left shunt flow to maintain the output of left ventricle are some of the reasons to explain the lack of symptoms and very slight impact on daily life. Helical CT, in particular 3-dimensional imaging, is very useful in diagnosing complicated cardiovascular deformation as in this case.

Key words: atrial septal defect, cardiovascular deformation, 3-dimensional imaging

Introduction

Total anomalous pulmonary vein drainage (TAPVD) is a rare congenital heart disease which has abnormal drainage of the pulmonary vein into the superior vena cava or other veins. Because almost all TAPVD patients are diagnosed and operated in childhood, it is rare to find an adult patient with such a disease.

Helical computed tomography (CT), a fourth generation CT combined with a continuously moving coach, can provide images with high continuity, and from the helical CT data we can obtain multiplanar reconstruction (MPR) or 3-dimensional reconstruction (3-D) images in static targets. For the beating heart, we developed electrocardiogram (ECG)-gating and diastolic reconstruction. By using these methods, we can clearly see how the heart and the great vessels are connected in detail.

Here, we report an adult patient with TAPVD who was not definitely diagnosed by echocardiography or angiography but was diagnosed by helical CT.

Case Report

The patient, a 41-year-old man, although he had not complained of anything in usual life, complained of palpitation and dyspnea on strenuous effort. Cardiomegaly and the heart shift to the right were pointed out in childhood. He was suddenly had a palpitation attack at midnight on October 31, 1994, and he came to the hospital by ambulance. He was conscious, although drowsy, and he had a cold sweat. Blood pressure was 86/62 mmHg, his lip was cyanotic and clubbed finger was noticed. ECG showed atrial flutter with 1:1 conduction (HR 220/min) and sinus rhythm recovered after direct current cardioversion. The ECG also indicated a right axis deviation and incomplete bundle branch block attributable to right ventricular overload. A grade 3 holosystolic murmur was audible at the left sternal border of the fourth intercostal space and fixed splitting of second sound was found.

The patient had polycythemia (red blood cells (RBC): 554 × 10^6/mm³, hemoglobin (Hb): 17.1 g/dl) and hypoxemia (pH: 7.425, partial pressure of carbon dioxide (PaCO₂): 34.8 mmHg, partial pressure of oxygen (PaO₂): 60.5 mmHg). Cardiomegaly, the heart shift to the right, an abnormal shadow (considered as vessels in the upper mediastinum) and peripheral pulmonary artery dilatation were observed on chest X-ray (Fig. 1). Echocardiography manifested the following: 1) left ventricular hypoplasia (LVDd = 38 mm, LVDs = 19 mm), 2) remarkable right ventricular enlargement, 3) atrioseptal defect, 4) dilated superior vena cava with abnormal flow and 5) small LA (LAd = 20 mm).
Adult TAPVD Diagnosed by Helical CT

Figure 1. Chest X-ray on admission. Cardiomegaly, the heart shift to the right, abnormal shadow (considered as vessels in the upper mediastinum) and peripheral pulmonary artery dilatation were observed.

Table 1. Cardiac Catheterization. The Pressure in the Right Ventricle and Pulmonary Artery was Increased and the Saturation of Oxygen in the Right Side of the Heart and Superior Vena Cava was Higher than Normal. In Addition, the Qp/Qs was 3.0.

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Based on these findings, TAPVD was suspected and the patient was referred for cardiac catheterization. The results are shown in Table 1. The pressure in the right ventricle and pulmonary artery was increased and the saturation of oxygen in the right side of the heart and superior vena cava was higher than normal. In addition, the Qp/Qs was 3.0. The coronary angiography and left ventriculography were within normal limits.

Helical chest CT showed (Fig. 2) abnormal connection of the vertical vein to the superior vena cava through an innominate vein observed at the innominate vein level (Fig. 2A) and at the level of the bifurcation of the pulmonary artery (Fig. 2B), and an abnormal connection of left and right pulmonary veins to the vertical vein at the atrial level (Fig. 2C). Furthermore, the image of MPR obtained by longitudinal slicing of the vertical vein itself showed a definite successive connection of the vertical vein to the superior vena cava through the innominate vein (Fig. 3). The 3-D images reconstructed from helical scanning more clearly demonstrated these abnormal connections of vessels than the MPR images (Fig. 4).

Discussion

With the abnormal drainage of the pulmonary vein into the superior vena cava or other veins, TAPVD is a rare congenital heart disease (1) and its incidence is only 1–3% among the total incidence of congenital heart diseases. The majority of the patients will die (2–4) during childhood unless they undergo surgical treatment (5–11). In the case presented, the 41-year-old man had no particular symptoms until admission. The absence of pulmonary vein stenosis, which enables sufficient pulmonary flow to be oxygenated in the lungs, and the presence of ASD providing enough right to left shunt flow to maintain the
output of the left ventricle are some of the reasons which explain the lack of symptoms and such slight impact on his daily life.

Although the dilated superior vena cava with some abnormal flow was detected by echocardiography and partial anomalous pulmonary vein drainage was suspected, a definite diagnosis could not be made based on echocardiographic findings only. It is quite reasonable to assume that the deformations are located behind the heart and it may be very difficult to obtain satisfactory images (12). As a rule, angiography is the most common method to confirm the diagnosis. In this case however, the contrast agent used was not sufficient to distinctly depict the dilated vessels including the superior vena cava, vertical vein and innominate vein.

3-D helical CT is very useful in diagnosing cardiovascular deformation, as shown in this case, since it can provide high resolution and high successive images within 30 seconds. In addition, motion artifacts can also be reduced when it is combined with ECG-gating and diastolic reconstruction. The images obtained are of good quality and are very easy to observe. In particular, the 3-D images displayed the abnormal relationship of the malformed vessels so clearly that it was possible to obtain a full understanding of the situation.
Adult TAPVD Diagnosed by Helical CT

Figure 3. Helical chest computed tomography (multiplanar reconstruction image). Ao: aorta, Inn V: innominate vein, LV: left ventricle, PA: pulmonary artery, RA: right atrium, SVC: superior vena cava, VV: vertical vein.

Figure 4. Helical chest computed tomography (3-dimensional image), anterior view (left side) and posterior view (right side). Ao: aorta, Inn V: innominate vein, LPA: left pulmonary artery, LPV: left pulmonary vein, RPA: right pulmonary artery, LPA: left pulmonary artery, SVC: superior vena cava, VV: vertical vein.
References


