

Evaluating adult cor triatriatum with total anomalous pulmonary venous connections by multidetector computed tomography angiography

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A 19-year-old female patient was admitted to our hospital with dyspnea, chest pain, and shortness of breath. A chest radiograph showed mild cardiomegaly. Echocardiography revealed an extra chamber in the heart. To evaluate this abnormality, ECG-gated 16-detector-row computed tomography angiography was performed. Multidetector computed tomography (MDCT), showing cor triatriatum with total anomalous pulmonary venous connections (TAPVC), clearly revealed cardiac and vascular anatomy. ECG-gated cardiac MDCT is a useful tool for detection and characterisation of cor triatriatum and related anomalies. (Folia Morphol 2011; 70, 4: 312–314)

Key words: cor triatriatum, multidetector computed tomography, total anomalous pulmonary venous connections

INTRODUCTION

Cor triatriatum is an uncommon congenital cardiac anomaly that is found in only 0.1% of patients who have congenital heart disease [4]. This condition is associated with three cardiac chambers that have atrial morphology.

There are many congenital cardiac anomalies, such as mitral regurgitation, atrial septal defect, persistent left superior vena cava, and rarely partial anomalous pulmonary venous return, that can accompany cor triatriatum. We present multidetector computed tomography (MDCT) findings of cor triatriatum with total anomalous pulmonary venous connections (TAPVC) anomaly. To our knowledge, this is the only case in the literature that has cor triatriatum with total anomalous pulmonary venous return.

CASE REPORT

In June 2010, a 19 year-old-woman with a 5-year history of progressive dyspnea, chest pain, and shortness of breath was referred. She had no significant history of disease. On physical examination, blood pressure was 120/72 mm Hg; she had a heart rate of 73 beats/min and a regular respiratory rate of 14 breaths/min. Chest radiography showed mild cardiomegaly with normal pulmonary vascularity. An electrocardiogram showed sinus rhythm. Transthoracic echocardiography revealed a membrane that subdivided the dilated left atrium into two chambers. According to echocardiographic findings, pulmonary venous return anomaly was suspected and the patient was referred to radiology for MDCT examination.

MDCT was performed on a 16-detector-row CT scanner (Aquillon; Toshiba Medical Systems, Tokyo, Japan).

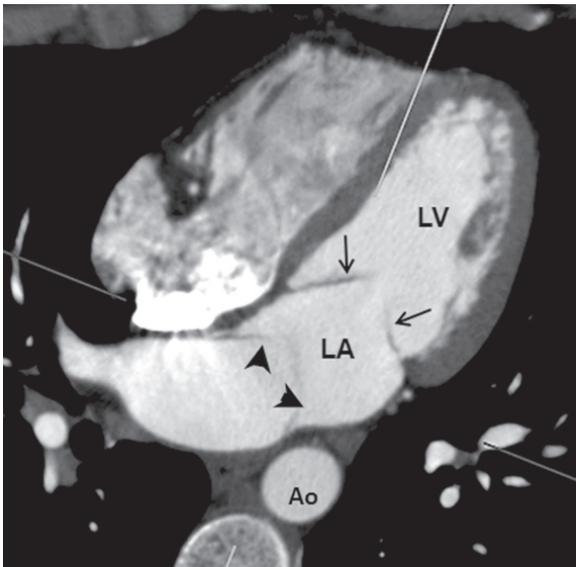


Figure 1. Axial contrast-enhanced cardiac image shows cor triatriatum; LV — left ventricle; LA — left atrium, Ao — aorta; arrows — mitral valve; black arrowheads — fibrous septa dividing left atrium.

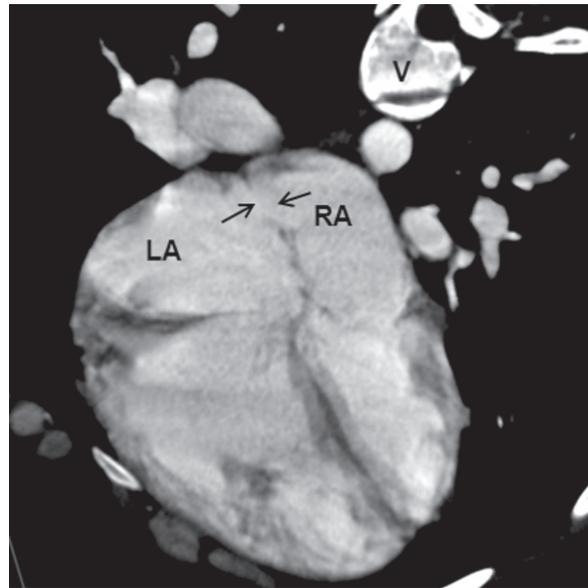


Figure 2. Coronal oblique MIP image shows atrial septal defect (arrows); LA — left atrium; RA — right atrium; V — thoracic vertebrae.

Scans were obtained with 16 0.5-mm collimation, 1.0-mm slice thickness and 1.0-mm reconstruction interval. About 90 mL iodinated contrast medium (Omnipaque, Amersham Health, Cork, Ireland) was injected intravenously at 4.5 mL/s followed by 40 mL saline at 2.5 mL/s. CT scans were obtained from 1 cm below the carina to the diaphragmatic face of the heart during a single breath-holding period with retrospective ECG gating.

MDCT showed a dilated left atrium with double-chamber conformation due to the presence of a fibrous membrane between the interatrial septum and the lateral left atrial wall (Fig. 1). The opening in the membrane was approximately 5 mm in diameter. A large atrial septal defect (ASD) was also present (Fig. 2). In addition, all pulmonary veins were returning to the right atrium via the coronary sinus (Fig. 3). The patient was referred to cardiovascular surgeon according to these findings.

DISCUSSION

Cor triatriatum is a rare condition first described by Church in 1868. Classic left cor triatriatum is characterised by a common pulmonary venous chamber separated from the true left atrium inferiorly by a fibro muscular membrane. The membrane is fenestrated to allow blood flow into the left atrium [1].

In this entity, prognosis is closely associated with the connection area between the two chambers via the fibro muscular membrane. Classification was made by Loeffler [3] in 1949, regarding the size and the number of the fenestrations. According to this classification: Group 1 lesions have no opening, and

the accessory left atrium drains into the right heart; Group 2 lesions have few and small fenestrations in the membrane, resulting in a high degree of obstruction; and Group 3 lesions have large openings in the membrane, leading to little or no obstruction.

In TAPVC, all of the pulmonary veins drain into the right atrium or a systemic venous channel which then drains into the right atrium. The essential feature of TAPVC is the failed union of the pulmonary veins with the developing left atrium, in combination with a persistent embryological connection between the pulmonary and systemic venous systems. TAPVC is classified as supracardiac (typically involving a vertical vein connecting to the brachiocephalic vein), cardiac (drainage directly to the right atrium or coronary sinus), infradiaphragmatic (usually via the portal vein), or connections at ≥ 2 levels. The onset and severity of symptoms depend on the magnitude of pulmonary artery hypertension as determined by the site of the TAPVC [5, 6].

The main imaging modalities used in the diagnosis of cor triatriatum are conventional angiography and echocardiography. However, these techniques may be inadequate in the diagnosis of such complex cardiac pathologies. In our case, cor triatriatum was revealed by echocardiography but TAPVC was not detected. MDCT was requested to identify the accompanying anomaly. Two right-sided and three left-sided pulmonary veins were present and all of them were returning to the right atrium through the sinus coronarius. The most interesting finding in our study was that the patient reached the adulthood with the condition undetected.

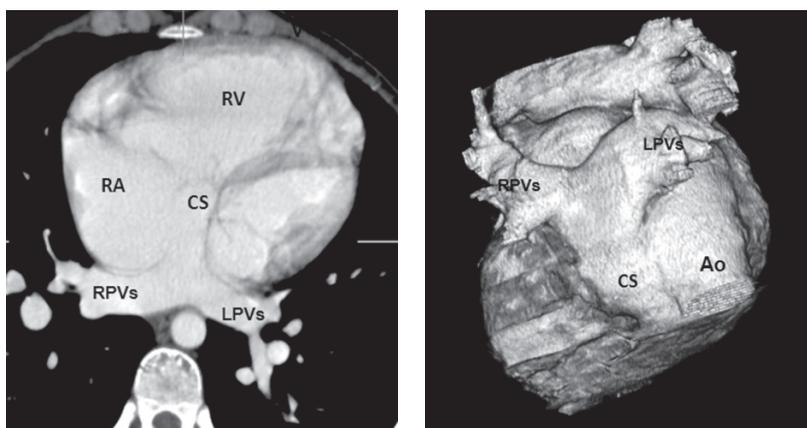


Figure 3. Axial MIP image (A) shows pulmonary veins draining into the coronary sinus, and posterior view 3D volume rendered image (B) shows total anomalous pulmonary venous connections. Two right-sided and three left-sided pulmonary veins were draining into the coronary sinus (CS); RV — right ventricle; RA — right atrium; RPVs — right pulmonary veins; LPVs — left pulmonary veins; Ao — aorta.

With 3D reconstruction and multiplanar reformat images, detailed evaluation of the cardiac anatomy was achieved and the acquired data was presented to the cardiovascular surgeon for further planning of the operation.

Several other techniques are used in the diagnosis of cor triatriatum including echocardiography, selective cardiac catheterisation, and magnetic resonance imaging. Echocardiography usually remains the primary imaging technique in diagnosis, but its overall anatomic depiction is relatively poor, especially outside of the heart. Selective cardiac catheterisation can be used in the diagnosis of such cardiac pathologies; however, it is an invasive and expensive method which requires hospitalisation. In addition, only one vascular tracing can be made in one cannulisation session. Cardiac MRI is also a technique for the evaluation of complex congenital heart disease. Cardiac MR imaging with three dimensional reconstruction has distinct advantages for pre- or postoperative assessment of pulmonary venous malformation in patients with cor triatriatum and TAPVC. However, MR imaging also has its limitations: long acquisition times may necessitate the sedation of young children and may present a problem in the examination of patients whose clinical condition is unstable [7].

CT has been used in patients with congenital heart disease, but its clinical applicability has been limited by the low temporal resolution of conventional CT. Nevertheless, novel advances in CT technology provide a non-invasive technique, offering an accurate diagnostic modality to visualise the cardiac structure by a three-dimensional display of anatomy. Multiplanar reconstructions and 3D volume rendering (VR) images can demonstrate pathologic cardiac structures in patients with congenital heart disease, even in adults [2, 8].

In our case, MDCT revealed a dilated left atrium with double-chamber conformation due to the presence of a fibrous membrane between the interatrial septum and the lateral left atrial wall. A large ASD was also present. In addition, all four pulmonary veins were returning to the right atrium.

In conclusion, ECG-gated cardiac MDCT is a useful tool for the detection and characterisation of cor triatriatum and related anomalies.

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