Case Report
pISSN 1738-2637 / eISSN 2288-2928
J Korean Soc Radiol 2013;69(3):197-200
http://dx.doi.org/10.3348/jksr.2013.69.3.197

대한영상의학회지

A Case Report of Congenitally Corrected Transposition of Great Arteries: Morphologic and Functional Evaluation with Cardiac CT[1]

증례 보고: 선천성 수정 대혈관 전위의 심장 CT 소견[1]

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Congenitally corrected transposition of the great arteries (ccTGA) is a rare congenital anomaly characterized by atrioventricular and ventriculoarterial discordance. We report a case of new-onset heart failure in a 69-year-old female in whom cardiac CT demonstrated ccTGA without the associated cardiovascular anomalies. In this case, cardiac CT was useful for elucidating the rare and unexpected congenital etiologies of abrupt-onset heart failure in an old patient by the simultaneous evaluation of cardiac morphology and function as a single study (inserted).

Index terms
Congenital Heart Disease
Transposition of Great Vessels
Computed Tomography

Received May 16, 2013; **Accepted** July 3, 2013
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INTRODUCTION

Congenitally corrected transposition of the great vessels (ccTGA) is a rare congenital anomaly with a prevalence of 1 per 33000 live births, accounting for approximately 0.05% of congenital heart malformations (1). This anomaly results from leftward (L-loop) looping of the primitive cardiac tube instead of the normal rightward looping (D-loop) (2). Despite the fact that over 90% of the cases have associated cardiovascular anomalies and prognosis depends on the severity of these anomalies, no other additional cardiac lesions, except for prominent trabeculation of right ventricle, were found in our patient. We describe herein the case of an old female with new-onset heart failure in whom cardiac CT first identified the characteristic features of the systemic right ventricle and the associated functional abnormalities as well as coronary-ventricular concordance.

CASE REPORT

A 69-year-old woman was presented with exertional dyspnea and increasing fatigue for the last three days. An echocardiography was performed on the day of admission, and severe systolic dysfunction of suspected right systemic ventricle and moderate left and right sided atrioventricular valve regurgitation were described. The ejection fraction of the systemic ventricle was decreased to 30%.

Cardiac CT was then performed to assess coronary artery disease, which might be a cause of new-onset heart failure, and further elucidate the previously noted ventricular abnormality described on the echocardiography. Retrospectively, electrocardiogram (ECG)-gated cardiac CT was performed with a 64-slice multi-detector CT system (Somatom Sensation 64 Cardiac, Siemens Healthcare, Forchheim, Germany). A standardized protocol with a collimation of 64 × 0.6 mm, gantry rotation time of 330 milliseconds, and a pitch of 0.2 were utilized. Tube voltage was 120 kV with an effective tube current-time product of 770 mAs_{eff} and an ECG-dependent tube current modulation (30-80% RR-interval). The dose-length product for this CT study was 551.49 mGy·cm with an estimated effective dose of 7.72 mSv. The scan was contrast-enhanced with 65 mL of a non-ion-

ic contrast medium (Ultravist; 300 mgI/mL, Bayer, Berlin, Germany), injected at 5 mL/s through an 18 G intravenous antecubital catheter, and used a bi-phasic contrast delivery protocol.

Cardiac CT showed no coronary artery disease (Fig. 1A, B). The left main coronary artery arose from the left coronary cusp anterior to the right coronary cusp and was divided into its two characteristic branches, the anterior descending (LAD) and circumflex coronary arteries (LCX). The LAD coursed in the anterior interventricular groove and supplied the morphological left ventricle. The LCX coursed in the anterior atrioventricular groove in the position occupied by the right coronary artery in the normal heart. The right coronary artery from the right coronary cusp supplied the anterior, lateral and posterior wall of the systemic right ventricle, and continued in the posterior atrioven-

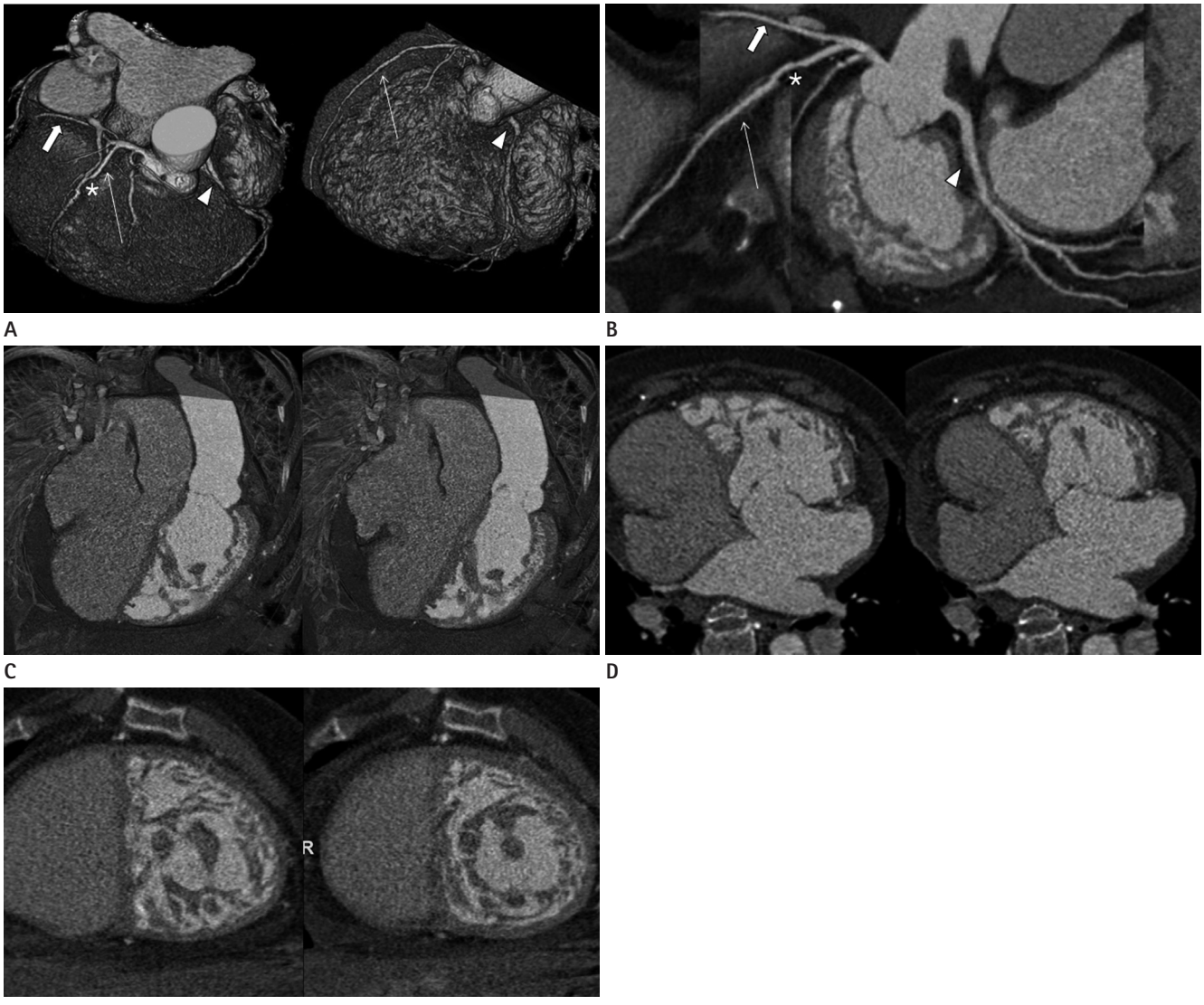

Fig. 1. Cardiac CT performed on 69-year-old female.
A. Three-dimensional volume-rendered images show spatial relationship of great arteries with ascending aorta and main pulmonary artery. Anterior descending artery (long arrow) and circumflex artery (short arrow) arise from left main coronary artery off of anterior aortic sinus. Note stair-step artifact (*) from irregular heart rhythm on mid segment of left anterior descending artery. Right coronary artery (arrowhead) originates from posterior aortic sinus.
B. Curved multiplanar reformation image demonstrates anterior descending (long arrow), circumflex (short arrow), and right coronary artery (arrowhead) without luminal stenosis. Again, stair-step artifact (*) is noted on mid segment of left anterior descending artery.
C, D. Diastolic (left) and systolic (right) reconstruction images clearly depict systemlic right ventricle and morphologic left ventricle on 3 dimensional volume rendering image **(C)** and 4 chamber view **(D)**.
E. Diastolic (left) and systolic (right) short axis reconstructions show hypertrabeculation and intertrabecular recesses filled with blood.

tricular groove. Thus, coronary ventricular concordance was confirmed.

Functional cine reconstructions revealed the thickening of the systemic ventricular wall with prominent trabeculations and deep intertrabecular recesses (Fig. 1C-E). Short axis images revealed that the intertrabecular recesses were filled with blood (Fig. 1E). Global functional parameters were obtained on an automated image processing workstation (Aquarius, Terarecon, San Mateo, CA, USA), which indicated an end-diastolic and end-systolic volume and ejection fraction of 218.65 mL, 154.14 mL, and 29.5%, respectively, all in good correlation with the echocardiography results. Because the clinical and imaging evaluation supported the diagnosis of heart failure associated with ccTGA and right systemic ventricular dysfunction without significant coronary artery stenosis, the patient received supportive treatment with diuretics, digitalis, and dobutamine infusion for systemic right heart failure; but unfortunately, she progressed to refractory cardiogenic shock and died.

DISCUSSION

ccTGA occurs in less than 1% of all forms of congenital heart disease. The associated anomalies in ccTGA include a ventricular septal defect in approximately 80% cases, pulmonic obstruction in 30-50% cases, left-sided Ebstein's anomaly, left atrioventricular (AV) valve regurgitation in 30-80% cases, conduction abnormalities and coronary anomalies. The finding of ccTGA without significantly associated anomaly is much less frequent (2, 3). Prognosis depends on AV conduction, arrhythmias, structural abnormalities, and degree of hemodynamic disturbance. Systemic right ventricular dysfunction in adulthood is also well known. Further, right ventricular failure can develop over time. This may be related to the coronary perfusion mismatch in the systemic right ventricle supplied by a single coronary artery as well as from the differences in the right ventricular fiber orientation, geometry, and microscopic structural features when functioning as the systemic ventricle. A multicenter study with congenitally corrected transposition of the great arteries demonstrated that 25% of patients without associated cardiac lesions and 67% of patients with other cardiac abnormalities developed congestive heart failure by age 45 (4).

In our case, extensive trabeculation was found in the systemic right ventricle without additional associated cardiac abnormalities. However, although debated, the right ventricle inherently has greater trabeculation than the left and marked trabeculation of the right systemic ventricle in response to systemic pressure and volume overload, which may be features of ccTGA (2, 5, 6).

Traditionally, echocardiography has been used to establish a diagnosis of ccTGA. In our case, cardiac CT, which was performed for the evaluation of new-onset heart failure in this patient, enabled a quantitative and qualitative assessment of global and regional ventricular function from the same dataset in addition to the morphological assessment for coronary artery and general cardiac morphology. Accordingly, the integrative nature of cardiac CT suggests an advantage of this modality over other imaging tests, such as echocardiography and cardiac magnetic resonance in this setting (7). In this patient with ccTGA, no significant coronary artery stenosis and no other intracardiac abnormalities were revealed by cardiac CT. Thus, the right ventricular dysfunction is most likely due to the effect of systemic pressures on the morphologic right ventricle along with atrioventricular regurgitation, as described in the initial echocardiography (8). Although the function of the systemic right ventricle tends to deteriorate gradually after the second decade of life, in this patient, however, the ventricular function was adequate to maintain a "normal" activity level into old age in the setting of ccTGA without significant associated abnormalities, such as interventricular communication and pulmonary outflow tract obstruction.

In conclusion, we report a rare case of ccTGA in a female patient who was presented with systemic right ventricular failure developed later than usual without the associated cardiac anomalies. In this case, cardiac CT was useful for elucidating the rare and unexpected congenital etiologies of heart failure by the simultaneous evaluation of cardiac morphology and function as a single study.

REFERENCES

1. Wallis GA, Debich-Spicer D, Anderson RH. Congenitally corrected transposition. *Orphanet J Rare Dis* 2011;6:22
2. Kharge J, Prasad MR, Ramegowda RT. An unusual case of congenitally corrected transposition of the great arteries associated with noncompaction-like remodeling of the

morphological right ventricle. *Echocardiography* 2011;28:E212-E214
3. Nagle JP, Cheitlin MD, McCarty RJ. Corrected transposition of the great vessels without associated anomalies: report of a case with congestive failure at age 45. *Chest* 1971;60:367-370
4. Graham TP Jr, Bernard YD, Mellen BG, Celermajer D, Baumgartner H, Cetta F, et al. Long-term outcome in congenitally corrected transposition of the great arteries: a multi-institutional study. *J Am Coll Cardiol* 2000;36:255-261
5. Patrignani A, D'Aroma A, Cicogna S. Unusual association between "congenitally corrected transposition of the great arteries" and "noncompaction" of the right systemic ventricle. *Int J Cardiovasc Imaging* 2009;25:551-553
6. Purvis J, Barr S. An appearance of "non-compaction" of the right systemic ventricle is common in congenitally corrected transposition of the great arteries. *Int J Cardiovasc Imaging* 2009;25:751-752
7. Ruzsics B. Integrative computed tomography imaging of ischemic heart disease. *J Thorac Imaging* 2010;25:231-238
8. Chang DS, Barack BM, Lee MH, Lee HY. Congenitally corrected transposition of the great arteries: imaging with 16-MDCT. *AJR Am J Roentgenol* 2007;188:W428-W430

증례 보고: 선천성 수정 대혈관 전위의 심장 CT 소견[1]

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선천성 수정 대혈관 전위는 방실 및 심실 대혈관 불일치연결을 특징으로 하는 매우 드문 선천성 심장질환이다. 저자 등은 69세 여자 환자에서 심부전을 평가하기 위해 시행된 심장 CT에서 우연히 발견된 선천성 수정 대혈관 전위와 이에 동반된 형태학적 우심실의 기능부전을 보고하고자 한다. 본 증례는 다른 기형이 동반되지 않아 비교적 늦게 심부전이 발생한 경우로 CT는 심장의 형태 및 기능의 이상을 동시에 평가함으로써 갑작스러운 심부전을 일으킨 본 질환을 진단하는 데 유용하였다.

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