Case Report

J Korean Soc Radiol 2021;82(4):982-987
https://doi.org/10.3348/jksr.2020.0104
eISSN 2288-2928

대한영상의학회지

A Case of Coronary Sinus Atresia with a Total Anomalous Cardiac Venous Drainage to the Left Atrium without Persistent Left Superior Vena Cava: Imaging Findings on Cardiac CT

지속성 좌상대정맥을 없이, 좌심방으로 연결되는 전관상정맥환류이상과 동반된 관상정맥동 입구폐쇄 1예: 심장전산화단층촬영 소견

Sang Hun Baek, MD, Eun-Ju Kang, MD*, Ki-Nam Lee, MD

Department of Radiology, Dong-A University Medical Center, Dong-A University College of Medicine, Busan, Korea

The coronary sinus (CS) is the venous drainage system of the heart. CS ostium atresia is a rarely seen cardiac malformation. Congenital atresia of the CS is usually found together with persistent left superior vena cava (LSVC) and other cardiac malformations. However, isolated congenital atresia of the CS is very rare. We present a rare case of isolated congenital atresia of the CS connecting the left atrium and coronary veins without persistent LSVC in a 58-year-old female.

Index terms Coronary Sinus; Congenital Abnormalities; Atresia; Heart; Computed Tomography, X-Ray

Received May 24, 2020
Revised September 11, 2020
Accepted October 29, 2020

***Corresponding author**
Eun-Ju Kang, MD
Department of Radiology,
Dong-A University Medical Center,
Dong-A University
College of Medicine,
26 Daesingongwon-ro, Seo-gu,
Busan 49201, Korea.

Tel 82-51-240-5367
Fax 82-51-253-4931
E-mail medcarot@dau.ac.kr

ORCID iDs
Sang Hun Baek
https://orcid.org/0000-0001-7453-1515
Eun-Ju Kang
https://orcid.org/0000-0003-0937-3607
Ki-Nam Lee
https://orcid.org/0000-0003-0848-3935

INTRODUCTION

The coronary sinus (CS) is the venous drainage system of the heart. The CS generally opens into the right atrium (RA) and returns blood from almost all areas of the heart (1). Although there have been many reported congenital cardiac malformations and abnor-

malities of the CS, they have not attracted much interest from researchers because they are probably not a big problem clinically or functionally, and also they are extremely rare. However, exact knowledge of abnormalities of the CS play an important role in various cardiac interventions, such as percutaneous transluminal coronary angioplasty and radiofrequency cardiac ablation (2-4).

There are many kinds of CS abnormalities that have been reported, and among them congenital CS ostium atresia is rarely seen CS abnormality and cardiac malformations (5). Congenital atresia of the CS is normally associated with single ventricle, hypoplastic left heart syndrome, atrial septal defect, systemic and pulmonary venous return anomalies including persistent left superior vena cava (LSVC) and other situs anomalies, particularly heterotaxy syndromes that involve the heart and other organs such as the stomach, intestine, liver, lungs and is often associated with complex congenital cardiac disease (1, 6). Especially, atresia of the CS is often accompanied by a persistent LSVC. However, isolated congenital atresia of the CS without persistent LSVC is very rare (5). We present a rare case of an isolated congenital atresia of the CS that cardiac veins are drained to the left atrium (LA) without persistent LSVC. It was incidentally found during cardiac CT.

CASE REPORT

A 58-year-old female was admitted to our cardiology clinic with an intermittent atypical chest pain for ten years. The patient denied having any systemic disease in the past. Her physical examination, vital signs, and the results of her laboratory tests were unremarkable. Electrocardiography was in sinus rhythm and there was no any ST-T segment change. Transthoracic echocardiography revealed normal functional parameters of the left ventricle without any regional wall motion abnormalities, and the cardiac chambers and valvular structures appeared normal. To rule out acute coronary syndrome and other heart problems that could cause chest pain, further evaluation was arranged in the form of a cardiac CT examination.

On the cardiac CT, curved multiplanar reformation (MPR) images showed the three major epicardial coronary arteries (left anterior descending, left circumflex, and right coronary arteries) were normally originating from the aortic sinus, and the diameters of each one was in the normal range without any obvious abnormality such as plaques or stenosis. The transaxial images show a completely obliterated CS with a remnant blind pouch, and the cardiac veins were drained to the LA through a common trunk, without any connection with the RA (Fig. 1A). On the short axis MPR images, show merging three cardiac veins that consist of great cardiac vein, posterolateral vein and middle cardiac vein. These cardiac veins was connected to the inferior wall of the LA, and there was no connection with the RA (Fig. 1B). Three-dimensional volume-rendered images gave a better view of the obliterated CS and anomalous venous connection just before communicating with the LA (Fig. 1C).

Because this cardiac venous anomaly was not thought to be the main cause of the patient's chest pain and also lack of clinical needs for treatment, we decided to follow up. Later, at request of the patient, the patient is being followed up at local hospital.

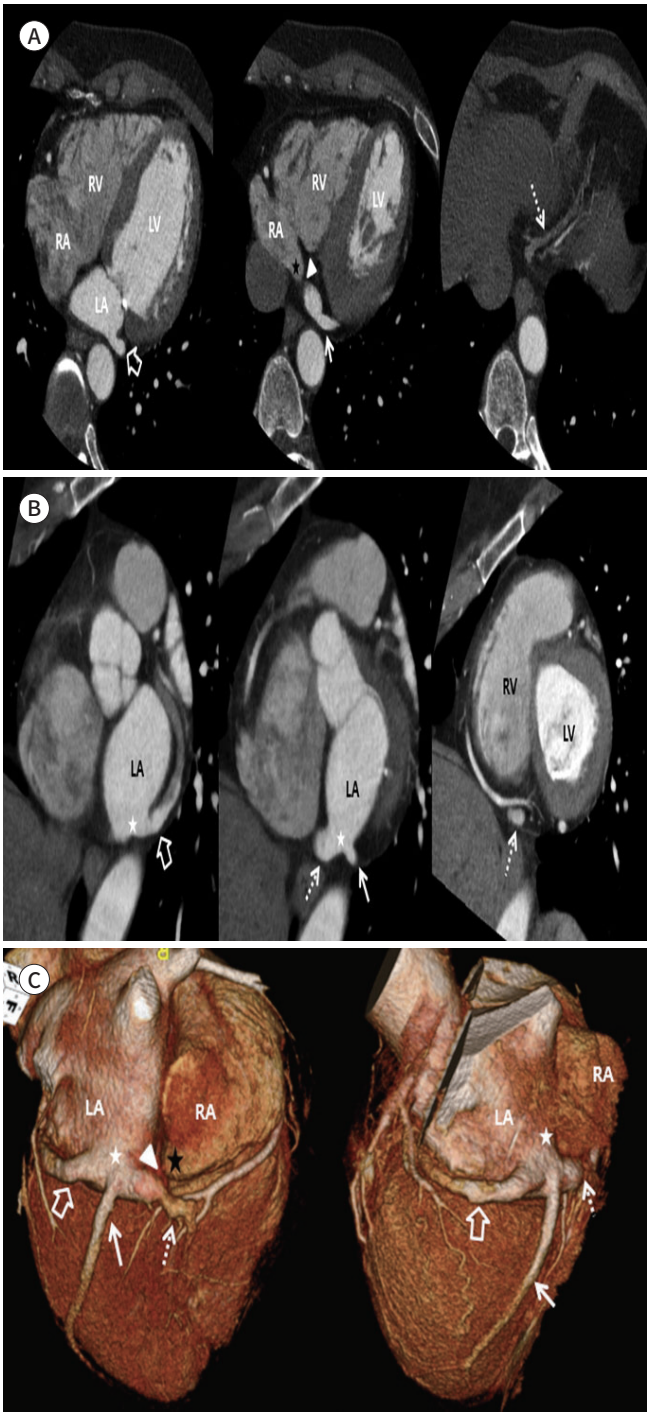

Fig. 1. Coronary sinus atresia with a total anomalous cardiac venous drainage to the LA without persistent left superior vena cava in a 58-year-old female.
A, B. Serial transaxial (A) and short-axis (B) multiplanar reformatted images show that the cardiac veins are connected with the LA by the merging of three cardiac veins (white stars): the great cardiac vein (open arrows), the posterolateral vein (white arrows), and the middle cardiac vein (dotted arrows). The coronary sinus is obliterated (arrowhead) without any connection to the cardiac veins, and the residual atresic right atrial ostium is shown (black star).
C. Three-dimensional volume-rendered images show a schematic view of the coronary sinus atresia and the abnormal communication between the merging cardiac veins (white stars) (great cardiac vein = open arrows; posterolateral vein = white arrows, middle cardiac vein = dotted arrows) and the LA. There is no connection (arrowhead) between the cardiac veins and the RA (black star).
LA = left atrium, LV = left ventricle, RA = right atrium, RV = right ventricle

DISCUSSION

Normal fetal venous development is a procedure of progression and then regression of the three major paired venous systems. By the 4th week of gestation, the ipsilateral horn of the sinus venosus receives blood from the common cardinal, umbilical, and vitelline veins (7). During the fifth week of development, the left vitelline vein degenerates and both the left

common cardinal vein and the left anterior cardinal vein form the LSVC. During the 7th week of gestation, the LSVC involutes following development of the left innominate vein and the LSVC becomes the ligament of Marshall. By the tenth week of gestation, the left common cardinal vein is occluded, and the left horn of the sinus venosus and the left common cardinal vein are retained as the CS in the adult. Moreover, the right horn of the sinus venosus persists as the RA (7). During this series of processes, the persistent LSVC commonly drains into the CS through embryologic communication of the left horn of the sinus venosus and the left common cardinal vein. Furthermore, in cases of an absence of CS or atresia of the CS ostium, functional communication of a persistent LSVC and the LA can occur (8).

In most previous published articles, CS abnormalities were found by necropsy, echocardiography, MRI, and coronary angiography (1, 7). However, recently, coronary CT has become widely used for screening and ruling out acute coronary syndromes. As a result, CS anomalies are now being found incidentally.

A classification of CS anomalies was proposed by Mantini et al., including: 1) enlargement of the CS with/without a left-to-right shunt, 2) absent CS, 3) atresia of the CS ostium, and 4) hypoplasia of the CS. These abnormalities are regarded as the result of incorrect embryonic development that occurs in the heart (6).

In the case of atresia of the CS ostium, the CS usually has a blind pouch with atresia or severe CS ostium stenosis. In this situation, venous blood cannot enter the RA through the CS ostium. According to previously reported literature, there are three major pathways for blood to enter the RA (Supplementary Fig. 1 in the online-only Data Supplement): 1) the venous blood returns through a functional persistent LSVC; 2) the venous blood returns through a gross communication that connects the CS and the LA with a persistent LSVC. Although there is a persistent LSVC, the venous blood does not flow that way; and 3) the venous blood returns through multiple communications between the CS and the related atria without a persistent LSVC. In such a case, there have been venous collateral pathways such as intraseptal veins, cardiac vein branches, and direct communications between the CS and LA or RA (6, 9).

Unroofed CS is a rare congenital anomaly, a kind of congenital intra-cardiac shunt (atrial septal defect), and is produced by the failure of separation of the superior wall of the CS with the atrium, forming a communication between both atria. There are very rare occasions when atresia of the CS ostium is accompanied with unroofed CS, which may seem similar to our case in the hemodynamic aspect (6). However, limited radiologic images of unroofed CS with ostial atresia have been reported in previous literature by echocardiographic findings, and the morphologic appearance in our case is different from that in several previously reported cases of unroofed CS with ostial atresia (10).

There are few reported cases of congenital atresia of CS communication to the LA and most of them exist via alternative pathways such as a window to the LA, multiple enlarged thebesian veins, and through the levoatriocardinal veins (1, 6). However, in our case, the cardiac veins were communicating to the LA without persistent LSVC or other alternative pathways mentioned above (Supplementary Fig. 1 in the online-only Data Supplement). We speculated that there may be other CS outflow routes and the other blood flow to RA through the thebesian veins hidden within myocardium, so it may not produce a significant hemodynamic effect, even though it is still a possible systemic embolic source (6, 9).

In conclusion, we reported a rare case of incidentally detected isolated congenital atresia of the CS, where cardiac veins drained to the LA without persistent LSVC on cardiac CT. To our knowledge, this is the first report of this anomaly in cardiac CT images with various kind of reconstruction, which can help to understand the anatomic appearance more clearly.

Supplementary Materials

The online-only Data Supplement is available with this article at http://dx.doi.org/10.3348/jksr.2020.0104.

Author Contributions

Conceptualization, K.E.; investigation, B.S.H.; methodology, L.K.; supervision, K.E.; visualization, B.S.H.; writing—original draft, B.S.H.; and writing—review & editing, K.E., L.K.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

Funding

None

REFERENCES

1. Janssen DR, Graham TP, Doyle TP, Moore JD. Occlusion of the coronary sinus ostium: a danger for cavopulmonary anastomosis. *Catheter Cardiovasc Interv* 2005;64:108-111
2. Muster AJ, Naheed ZJ, Backer CL, Mavroudis C. Is surgical ligation of an accessory left superior vena cava always safe? *Pediatr Cardiol* 1998;19:352-354
3. Ruengsakulrach P, Buxton BF. Anatomic and hemodynamic considerations influencing the efficiency of retrograde cardioplegia. *Ann Thorac Surg* 2001;71:1389-1395
4. Saremi F, Muresian H, Sánchez-Quintana D. Coronary veins: comprehensive CT-anatomic classification and review of variants and clinical implications. *Radiographics* 2012;32:E1-E32
5. Foale RA, Baron DW, Rickards AF. Isolated congenital absence of coronary sinus. *Br Heart J* 1979;42:355-358
6. Mantini E, Grondin CM, Lillehei CW, Edwards JE. Congenital anomalies involving the coronary sinus. *Circulation* 1966;33:317-327
7. Miraldi F, Di Gioia CR, Proietti P, De Santis M, D'Amati G, Gallo P. Cardinal vein isomerism: an embryological hypothesis to explain a persistent left superior vena cava draining into the roof of the left atrium in the absence of coronary sinus and atrial septal defect. *Cardiovasc Pathol* 2002;11:149-152
8. Geva T, Van Praagh S. *Anomalies of the pulmonary veins.* In Allen HD, Driscoll DJ, Shaddy RE, Feltes TF, eds. *Moss and Adams' heart disease in infants, children, and adolescents: including the fetus and young adult.* 7th ed. Philadelphia: Lippincott Williams & Wilkins 2008
9. Chou MC, Wu MT, Chen CH, Lee MH, Tzeng WS. Multidetector CT findings of a congenital coronary sinus anomaly: a report of two cases. *Korean J Radiol* 2008;9:S1-S6
10. Kim SJ, Ann SH, Kim YG, Park GM, Won KB, Lee SG. A case of unroofed coronary sinus with coronary sinus orifice atresia: use of multimodality imaging. *J Cardiovasc Imaging* 2018;26:103-105

J Korean Soc Radiol 2021;82(4):982-987

대한영상의학회지

지속성 좌상대정맥을 없이, 좌심방으로 연결되는 전관상정맥환류이상과 동반된 관상정맥동 입구폐쇄 1예: 심장전산화단층촬영 소견

백상훈 · 강은주* · 이기남

관상정맥동은 심장의 정맥 배출 체계이다. 관상정맥동 입구의 폐쇄는 드문 심장 기형이다. 선천적인 관상정맥동 입구의 폐쇄는 주로 지속성 좌상대정맥을 동반하며 그 외에 다른 심장 기형과 동반되어서 나타난다. 하지만 선천적인 관상정맥동 입구의 폐쇄만 단독적으로 나타나는 경우는 아주 드물다. 이 논문에서는 58세 여성에서 발견된 지속성 좌상대정맥을 동반하지 않고, 좌심방과 심장정맥이 연결되는 선천적인 관상정맥동 입구의 폐쇄만 단독적으로 나타나는 드문 증례를 보고하고자 한다.

동아대학교 의과대학 동아대학교의료원 영상의학과

https://doi.org/10.3348/jksr.2020.0104