Retrograde venography and three-dimensional mapping of a great cardiac vein with separate drainage into the high right atrium in a patient with Wolf-Parkinson-White syndrome

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ABSTRACT

A 13-year-old boy presenting with palpitations was diagnosed with Wolf-Parkinson-White syndrome. We elected to bring him to the electrophysiology lab for evaluation of the accessory pathway and potential ablation. Structural assessment of the coronary sinus with electro-anatomical mapping and retrograde venography led to the discovery of a great cardiac vein having a separate insertion from the coronary sinus into the high right atrium. The accessory pathway was localized to the left lateral portion of the mitral annulus and treated successfully with radiofrequency ablation. Such venous anatomical variant has been seldom reported, and its association with an accessory pathway is described here for the first time. Its recognition can have important implications when performing procedures that require access into the coronary venous system.

KEYWORDS Congenital heart disease; great cardiac vein; retrograde venography; Wolf-Parkinson-White

The coronary sinus (CS) drains most of the venous blood from the heart. Many of its anatomical variants have been described, and some of them have implications for procedures like cardiac resynchronization therapy, heart surgery, mapping and ablation of accessory pathways, and ventricular tachycardias. The great cardiac vein (GCV) and middle cardiac vein are tributaries to the CS and have been shown in imaging and autopsy studies to be very consistent morphologically. We present a patient with a GCV that drained separately from the CS into the right atrium that was found incidentally during mapping and ablation of an accessory pathway.

CASE DESCRIPTION

A 13-year-old boy was referred to the cardiology clinic with a complaint of palpitations. His resting 12-lead electrocardiogram was normal. A Zio XT (iRhythm Technologies, Lincolnshire, IL) monitor revealed multiple episodes of symptomatic supraventricular tachycardia with

preexcitation, leading to a diagnosis of Wolf-Parkinson-White syndrome. A transthoracic echocardiogram was negative for structural abnormalities. We proceeded with mapping and ablation of the accessory pathway. Catheter manipulation in the right atrium led to engagement of two separate venous ostia. One had the typical location of the CS and drained the middle and posterior cardiac veins. The second ostia was in the superior aspect of the right atrium and drained the GCV. This was demonstrated by retrograde venography (*Figures 1 and 2*) and three-dimensional electro-anatomical mapping using EnSite Velocity NavX (model Z600, St. Jude Medical, Inc., St. Paul, MN) (*Figure 3*). To reach the accessory ostia, the GCV had to course through the anterior aspect of the transverse sinus of the pericardium. The accessory pathway was localized at the left lateral portion of the mitral annulus. Radiofrequency ablation was successful at this site and the patient was able to leave home the following day.

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Received May 17, 2018; Accepted June 18, 2018.

Figure 1. Retrograde venogram of the great cardiac vein (left) and the coronary sinus (right) observed from a right anterior oblique 30° angle. Drainage of the great cardiac vein to the superior aspect of the right atrium as well as its course through the anterior wall of the left ventricle can be appreciated.

Figure 2. Retrograde venogram of the great cardiac vein (left) and the coronary sinus (right) observed from a left anterior oblique 30° angle. The great cardiac vein is seen traveling toward the interventricular sulcus, and the coronary sinus has its expected course through the atrioventricular groove toward the lateral mitral annulus.

Figure 3. The anatomy of the great cardiac vein and the coronary sinus was also reproduced with electro-anatomical mapping. The great cardiac vein appears in blue and the coronary sinus in yellow. On the left is a right anterior oblique view and on the right a left anterior oblique view.

DISCUSSION

Abnormalities in drainage of the GCV are not often heard of and are likely to be underreported, with most cases found incidentally in cardiac imaging and autopsy studies.¹ This vein normally originates at the apex of the heart as the anterior interventricular vein and becomes the GCV as it reaches the lower to middle third of the interventricular sulcus.² It then becomes the CS at the level of the Vieussens valve.² This vein and the middle cardiac vein appear to be morphologically the most consistent of the coronary venous anatomy.²

Cases reporting drainage of the GCV at a separate ostium in the right atrium are very limited, and these also describe an anatomical course similar to that of our patient.^{3,4} It first travels through the conus arteriosus and then the transverse pericardial sinus to finally reach the right atrium. In a similar manner, the GCV has also been found to drain into the superior vena cava.^{5,6} A review of 620 human heart specimens did not reveal abnormalities of the GCV drainage, indicating how difficult these can be to find.⁷ Another review of 337 human heart specimens only found one GCV draining into the superior vena cava.⁸

Abnormal drainage of the GCV into other structures has been reported. In a review of 250 hearts, there was a case of a GCV draining into the anterior cardiac veins, which drain the anterior wall of the right ventricle.⁹ Drainage into the left internal thoracic vein was found during surgery in a 17-month-old girl who was undergoing repair of a ventricular septal defect.¹⁰ Drainage into the left atrium was incidentally discovered during computed tomography of the coronaries in a patient with chest pain and in another patient who underwent surgical revascularization.¹¹

For practitioners who perform procedures involving the venous system of the heart, it is imperative to understand the variants that could be encountered. Failure to recognize them can lead to complications, and recognition can be a challenge because of their infrequency. For example, retrograde cardioplegia during cardiac surgery is performed through the coronary sinus.¹² If a GCV with separate drainage is not recognized, a large amount of myocardial mass can be compromised. The alternative in this case would be to perform antegrade cardioplegia through the coronary arteries. In a cardiac resynchronization therapy procedure, we would still have at our disposition branches from the CS for placement of a left ventricular lead at the most lateral region possible. If an adequate target in the CS cannot be found, then the GCV variant could be engaged, with placement of the lead in an anterolateral branch.

In theory, ablation of a mitral isthmus flutter could become difficult because the GCV may not reach the epicardial portion of the lateral mitral annulus, which is often necessary to achieve a line of complete block.¹³ Mapping and ablation of premature ventricular contractions initiating from the left ventricular summit may be more

challenging because the GCV offers the region of closest proximity to this structure.¹⁴

This case also represents the first description of a GCV draining separately into the right atrium that is associated with an accessory pathway. Other coronary venous anomalies such as coronary sinus diverticula are known to be linked with accessory pathways.¹⁵

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