ATRIAL FIBRILLATION

COMPLEX CASE STUDY

Ablation of Anomalous Pulmonary Veins Triggering Atrial Fibrillation: A Novel Association and Approach to Ablation

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ABSTRACT. Atrial fibrillation ablation is commonly targeted at pulmonary vein isolation because in the majority of patients the veins serve as an arrhythmogenic trigger. In a small number of patients, anomalous pulmonary venous anatomy may exist, including pulmonary vein drainage directly into the right atrium. The frequency with which patients presenting for atrial fibrillation ablation have anomalous pulmonary veins is unknown. Furthermore, these anomalous veins have been previously reported to not have electrical connection to the heart. We present a case of a patient in whom an anomalous right superior pulmonary vein was found draining into the right atrium and was a trigger for atrial fibrillation. Furthermore, challenges related to isolation of this vein including use of epicardial access to reduce the risk of phrenic nerve injury are discussed.

KEYWORDS. atrial fibrillation, ablation, phrenic nerve, anomalous pulmonary vein.

Introduction

Pulmonary vein isolation is a well-established treatment for atrial fibrillation. Anisotropy of cardiac musculature at the pulmonary vein–atrial interface, a high density of autonomic innervation in the region of the pulmonary vein ostia, and other unique electrophysiologic characteristics of pulmonary vein tissue are thought to contribute to their arrhythmogenicity. In the majority of patients with recurrent atrial arrhythmias after prior pulmonary vein isolation, electrical reconnection to the atrium of one or more pulmonary veins is usually implicated as the cause. However, anatomic variations in the number and location of pulmonary vein ostia are possible. While anatomic variability commonly exists in the number of ostia entering into the left atrium, when performing wide area circumferential ablation such variability is unlikely to impact the likelihood of procedural success. However, in rare patients, one or more pulmonary veins may enter the right rather than the left atrium, most commonly the superior vena cava (SVC), but also directly into the right atrium when they are often associated with a sinus venosus defect or into the inferior vena cava. Termed partial anomalous pulmonary venous connection, it may be seen in as many as 0.5% of patients and is often not diagnosed until adulthood when pulmonary hypertension and right ventricular dysfunction develop due to the increased right-sided venous return.

While it is known that the pulmonary veins serve as a principal arrhythmogenic trigger in atrial fibrillation, it is

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not as clear what role, if any, anomalous pulmonary veins may have in the pathogenesis of atrial fibrillation. While the putative mechanism by which the pulmonary veins trigger atrial fibrillation is thought to be electrical anisotropy at the vein–atrial interface, it is unclear if anomalous pulmonary veins retain similar electrical connections to the heart. Four prior case reports of atrial fibrillation ablation in patients with anomalous pulmonary veins have suggested that they are electrically silent.9–12 We present a case of a patient in whom anomalous return of a right superior and right middle pulmonary vein to the right atrium demonstrated electrical connection to the heart, served as the trigger for atrial fibrillation and we further discuss the potential hurdles to achieving electrical isolation and novel solutions to overcoming these obstacles.

Case report

A 55-year-old woman without significant past medical history was referred for further management of atrial fibrillation diagnosed 2 years previously in the setting of heart failure symptoms attributed to rapid ventricular rates. She was cardioverted after her first episode of atrial fibrillation and started on sotalol, despite which she had recurrent atrial fibrillation requiring repeat cardioversions. Given her recurrent, symptomatic atrial fibrillation in spite of antiarrhythmic drug therapy, the decision was made to refer her for catheter ablation.

Prior to ablation, magnetic resonance imaging (MRI) was performed as part of routine pre-ablation imaging. An anomalous right superior and middle pulmonary vein entering the right atrium posteriorly near the SVC/right atrial junction was noted (Figure 1A). Electroanatomic mapping and angiography (Figure 1B) confirmed the anatomy of the anomalous connection of a right middle and right superior pulmonary vein to the right atrium. The remaining pulmonary veins (the right inferior, left superior, and left inferior) all entered normally into the left atrium. No septal defect was identified by intracardiac echocardiography or MRI in association with the anomalous veins.

A baseline right heart catheterization was performed and demonstrated a pulmonary artery systolic pressure of 39 mmHg with a Qp:Qs of 1.9. A transseptal puncture was performed and a complete left and right atrial map was created. Triggers for atrial fibrillation were seen from the anomalous right pulmonary vein. The left atrial pulmonary veins (i.e., the right inferior pulmonary vein and both the left superior and left inferior pulmonary veins) were isolated first with documentation of entrance and exit block. During attempt at isolation of the anomalous right pulmonary vein, large areas of phrenic capture during pacing with outputs of 20–40 mA were seen along the lateral aspect of both the vein and the SVC, obviating the ability to form a circumferential lesion set without risking phrenic injury. Attempts to map electrically active regions inside the vein to attempt to disconnect the vein without creating a circumferential lesion set were still limited due to capture of the phrenic nerve with pacing. Thus, a partial lesion set was created and a decision was made to refer for further hemodynamic and subsequent surgical evaluation for possible repair of the anomalous pulmonary venous connection which could also serve as a means of electrically isolating the arrhythmogenic vein from the atrium.

Given repeat MRI revealing a Qp:Qs of 1.4 and a lack of pulmonary hypertension, right heart dysfunction, or heart failure symptoms when in sinus rhythm, the risk of surgical repair of her anomalous pulmonary venous return was felt to outweigh potential benefits. She continued to have episodes of symptomatic atrial fibrillation requiring cardioversion despite sotalol and dofetilide over the ensuing months, and thus the decision was made to take her back for a repeat attempt at ablation. Prior to the procedure, the potential for epicardial access to attempt to avoid phrenic injury by separating off the phrenic nerve using a balloon or air was discussed with the patient.

During the second procedure, an electroanatomic shell (CARTO, Biosense, Diamond Bar, CA) of both the right and left atrium was created. Chronic entrance and exit block of the left atrial veins (i.e., the left superior and inferior pulmonary veins and the right inferior pulmonary vein) was documented. During catheter manipulation in the left atrium, heparin was given and titrated to an activated clotting time (ACT) of 350–400 (pre-procedure international normalized ratio (INR) was 2.3). There was apparent electrical connection of both anomalous right veins and the SVC consistent with the partial isolation previously performed. Signals triggering atrial fibrillation and frequent premature atrial depolarizations arising from the anomalous pulmonary vein were seen (Figure 2). Repeat mapping by pacing at outputs ranging from 20 to 40 mA showed a large area of phrenic capture obviating safe endocardial ablation without risking phrenic injury similar to the prior ablation.

Epicardial access was obtained without complication after fully reversing the ACT with protamine and a 8.5 French bidirectional sheath was advanced into the pericardial space. A Tyshak-X balloon (8 mm diameter, 4 cm length; NuMed, Inc., Hopkinton, NY) was advanced via the bidirectional sheath and positioned behind the area of the anomalous right vein/SVC. Maneuvering the balloon was difficult at first and positioning to sufficiently separate the phrenic nerve off of the epicardial surface of the heart was not achieved. Thus, air was also injected into the pericardial space to assist in maneuvering of the balloon as well as to allow for increased separation of the pericardium and phrenic nerve off of the epicardial surface. A total of 30 mL of air was injected into the space and there was a 10 mmHg drop in systolic blood pressure which then stabilized. This allowed for better maneuverability of the balloon and phrenic capture at 20–40 mA output in regions of prior capture was no longer seen.

Ablation was then performed circumferentially around the SVC and anomalous right veins as a common ostium. Pacing at 20 mA was performed prior to each lesion to...
Figure 1: The anomalous right veins in relation to the superior vena cava (SVC) entering the right atrium. (A) Magnetic resonance imaging of where the anomalous right veins enter the right atrium (white arrows). (B) Angiography performed using a long bidirectional steerable sheath placed in the right atrium in left anterior oblique and right anterior oblique views (top two images are of the SVC and the bottom two are of the anomalous right veins). (C) Intracardiac echocardiography delineating the SVC from the anomalous right veins.
Figure 2: (A) The electroanatomic map of the left and right atrium and the relative catheter positions during isoproterenol infusion prior to ablation. (B) The electrograms with the catheters as shown with atrial arrhythmias arising from the anomalous right vein.
Figure 3: (A) The left anterior oblique and right anterior oblique (RAO) views of the epicardial placed, contrast-infused balloon positioned between the superior vena cava/anomalous right vein junction within the right atrium and the overlying phrenic nerve and pericardium. Air was also infused (clear space best seen on RAO view) to help separate the phrenic nerve off the heart surface. (B) The final lesion set (red spots represent ablation lesions placed without epicardial balloon or air; white spots represent areas of phrenic capture with pacing; purple spots represent ablation lesions placed while the epicardial balloon and air was used.)
Isoproterenol was then given up to a dose of 30 mg/min of the SVC or anomalous right veins. Adenosine was used entrance and exit block with no evidence of reconnection. After a 30-min waiting period, there was persistent atrial fibrillation and thus require terminating energy delivery early. Isoproterenol was then given, and there was no evidence of reconnection. Isoproterenol was then given up to a dose of 30 mg/min of the SVC or anomalous right veins. Adenosine was used entrance and exit block with no evidence of reconnection. After a 30-min waiting period, there was persistent atrial fibrillation and thus require terminating energy delivery early.

The patient did well over a 4-month follow-up period and has remained free of atrial fibrillation and off antiarrhythmic medications.

Discussion

We present a unique case in which an anomalous pulmonary vein served as the trigger for atrial fibrillation requiring electrical isolation. Part of the uniqueness of the case involves the need for epicardial access to physically separate the phrenic nerve off of the right atrium to achieve electrical isolation. To our knowledge, this has not been reported in the literature. Four prior published case reports in which patients presented with anomalous pulmonary venous return to the SVC or the right atrium suggested the absence of myocardial sleeves extending into these anomalous veins and thus that they were unlikely to play a role in triggering atrial fibrillation or other atrial arrhythmias seen. However, our case suggests that this is not always the case, and it is indeed possible for these veins to retain electrical connection to the heart.

The difficulties that may arise in treating these patients include 1) recognizing the presence of an anomalous vein; and 2) determining whether the anomalous pulmonary vein, the SVC, or both, should be targeted for isolation. Recognition of the presence of an anomalous pulmonary vein may be difficult and require preablative imaging including either MRI or computed tomography scan, which are commonly performed prior to or after ablation. Even when an anomalous vein is present, the SVC may serve as an independent trigger for atrial fibrillation and it needs to be delineated whether the SVC or the anomalous vein or both are serving as triggers for atrial fibrillation. Depending on the anatomic association, it may be reasonable to target isolation of both the SVC and the anomalous vein together or independently. In prior case reports, isolation of the SVC was performed in spite of the lack of evidence for electrical activity within the anomalous pulmonary vein. However, in our case there was clear electrical connection and triggers inducing atrial fibrillation were seen in the anomalous pulmonary vein as well as anatomic proximity to the SVC. Both the SVC and the anomalous right veins were targeted as a common ostium for isolation given that the anatomic location of the SVC relative to the anomalous right veins would have made isolation of the anomalous right veins independently from the SVC difficult.

Another important consideration during atrial fibrillation ablation, especially when in the region of the SVC or the right-sided veins, is the risk of phrenic nerve injury. Various methods including pacing to identify the anatomic course of the phrenic nerve prior to ablation have been used to avoid phrenic injury when creating septal lesions to the right-sided pulmonary veins in the left atrium or when isolating the SVC. However, when there is an extensive area of phrenic capture that obviates safe electrical isolation as seen in our patient, options may be limited. One option is to pace the phrenic nerve during each ablation lesion and immediately terminate the lesion upon loss of capture. However, there is still risk associated with this method as it requires delivering an insult to the phrenic nerve sufficient that conduction ceases prior to terminating ablation, and there is a lack of data on the safety of this approach. Another option is to pace around the region to try and identify the anatomic course of the nerve based on where capture is achieved. However, the relationship of the ability to capture the phrenic nerve at a specific pacing output relative to the likelihood of phrenic injury with a lesion delivered at that location is unknown. In other words, higher pacing outputs are associated with a larger “virtual electrode” that may lead to more far-field capture and thus may or may not identify structures that would be affected by a lesion given at the same spot. Whether there is a threshold pacing output at which phrenic capture can be achieved but ablation would not result in injury is unknown. Thus, consideration of whether or not to ablate needs to take into consideration all of these factors and realization that phrenic injury is possible.

The role of an intrapericardial balloon or infusion of air or saline to separate the phrenic nerve off the epicardial ventricular surface to perform epicardial ablation for ventricular tachycardia has been previously described. However, during endocardial ablation for ventricular tachycardia, it is unlikely that ablation would result in phrenic injury due to the thickness of ventricular tissue. This is not the case in atrial tissue where the relative thin walls make it more likely to affect the phrenic nerve during endocardial ablation. Thus, the same methods used for separating the phrenic nerve off the epicardial surface during epicardial ventricular ablation may be useful in complicated cases of endocardial atrial ablation, such as ours.

The limitations to epicardial access during atrial fibrillation ablation, however, must be taken into consideration. Generally, during atrial fibrillation ablation, large amounts of anticoagulation are used while catheters are in the left side of the heart. The risk of thrombus formation is also high immediately after ablation when the endocardial surface of the atrium is denuded and thus becomes relatively more thrombogenic. Thus, a decision needs to be made after having ablated in the left side of the heart.
atrium about the relative risk of thromboembolism from a prolonged duration off anticoagulation versus the risk of epicardial access. In the case of our patient, during the second procedure when epicardial access was obtained, the INR was therapeutic and no ablation was done in the left atrium likely allowing for safer reversal of heparin to allow for epicardial access. However, the risk of epicardial access at therapeutic INRs and the relative thromboembolic risk if epicardial access is immediately after extensive left atrial ablation is unclear.

In summary, we present the first case to our knowledge of a patient with atrial fibrillation presenting with an anomalous pulmonary vein in whom the anomalous vein had apparent electrical connection to the right atrium and served as a trigger for atrial fibrillation. Furthermore, we present the potential utility of epicardial access to improve the safety of ablation in patients in whom anatomic proximity to the phrenic nerve otherwise obviates safe endocardial atrial ablation. These findings have potential importance to pre-ablation considerations when determining pulmonary vein anatomy, in planning the ablation approach in patients with anomalous venous connections and in identifying ways of limiting the risk of phrenic injury when endocardial atrial ablation is required in close proximity to the nerve.

References