Radiofrequency Ablation of Post-Incisional Atrial Flutter and High-Output Heart Failure in a Patient with Interrupted Inferior Vena Cava and Hereditary Hemorrhagic Telangiectasia

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A 61-year-old female with a history of secundum atrial septal defect repair and hereditary hemorrhagic telangiectasia presented with epistaxis. She was found to have atypical atrial flutter with 2:1 atrioventricular conduction. Radiofrequency ablation was planned, but inferior vena cava interruption precluded right atrial (RA) access. The RA was then accessed through both subclavian veins, and activation mapping revealed a dense atriotomy scar in the posterolateral inferior RA. Wavefront propagation proceeded caudally through an area of slow conduction confined by the atriotomy scar. Atypical atrial flutter terminated during a second radiofrequency application to an isthmus confined by 2 regions of dense scar. The arrhythmia did not recur, although the patient later experienced typical atrial flutter and atrial fibrillation. High-output heart failure due to systemic arteriovenous shunt was confirmed by cardiac catheterization and improved markedly with bevacizumab therapy.

Case presentation
A 61-year-old female with a history of secundum atrial septal defect repair and HHT presented with epistaxis lasting for more than an hour. In the emergency room, she was noted to exhibit a narrow-complex tachycardia with a heart rate of 214 bpm. Treatment with intravenous diltiazem gradually decreased her heart rate to 108 bpm.

The heart rhythm was initially interpreted as atrial fibrillation with rapid ventricular response and subsequent conversion to sinus rhythm. The patient was admitted and an electrophysiology consult was obtained the next day. The patient was found to be in atypical atrial flutter with 2:1 atrioventricular (AV) conduction.

Interruption of the inferior vena cava (IVC) due to congenital or acquired causes can occasionally prevent catheter access to the right atrium (RA) via the femoral vein approach. Hereditary hemorrhagic telangiectasia (HHT) is a rare autosomal dominant vascular disorder with multiple arteriovenous malformations (AVMs). Herein, we present a patient with HHT and post incisional atrial flutter, in whom IVC interruption necessitated a bilateral subclavian approach for radiofrequency ablation (RFA). HHT patients with large AVMs may suffer from high-output heart failure due to systemic shunts; this patient experienced a dramatic response to bevacizumab treatment.
(Figure 1) rather than sinus rhythm; this persisted for >2 days after admission. This patient had undergone an uneventful repair of a secundum atrial septal defect 35 years before. She had recurrent severe epistaxis due to HHT. She was noted to have an elevated heart rate at rest.

**Figure 1.** A. 12-lead ECG of post-incisional atrial flutter with 2:1 AV conduction recorded in the emergency department after administration of IV diltiazem. This was initially misinterpreted as sinus tachycardia. B. Sinus rhythm after cardioversion (slightly different lead placement) is shown for comparison. Incomplete right bundle branch block and left axis deviation is consistent with a secundum atrial septal defect.
approximately 5 months earlier and was undergoing evaluation for presumed sinus tachycardia elsewhere.

The patient opted for RFA. The transesophageal echocardiogram excluded an intracardiac thrombus and confirmed normal left ventricular function. Both atria were dilated (left atrial diameter 51 mm). In the electrophysiology (EP) laboratory, neither a deflectable EP catheter nor a 0.035” J-tipped wire could be advanced from the right femoral vein into the RA. Contrast dye injection revealed a large hepatic AVM and IVC interruption (Figure 2). RFA of the atrial arrhythmia was performed a few weeks later. A 7 Fr vascular sheath was placed in each subclavian vein. A deflectable decapolar catheter was introduced to the coronary sinus from the left subclavian vein. A 4 mm curve F Navistar ( Biosense-Webster) ablation catheter was introduced to the RA from the right subclavian vein and was used for mapping and ablation. The patient was initially in normal sinus rhythm, with normal A-H and H-V intervals. A stable, well-tolerated atrial tachycardia with a cycle length of 335 ms and 2:1 AV conduction was easily induced with RA burst pacing. Electroanatomic mapping of the RA was performed during tachycardia using the CARTO ( Biosense-Webster) mapping system. This revealed a relatively small, dense atriotomy scar in the postero-lateral inferior RA, and a large area of low-amplitude, fractionated potentials, extending anteriorly to the crista terminalis and superior to the superior vena cava. The activation map indicated that the propagation wavefront proceeded caudally through an area of slow conduction confined by the atriotomy scar (Figure 3A), and subsequently propagated faster towards the interatrial septum along the RA floor. The coronary sinus activation proceeded in a proximal to distal direction. Twelve RFA applications were delivered to the presumed isthmus connecting the two areas of dense scar. The atrial tachycardia terminated during the second RFA delivery (Figure 3C). Conduction block across the isthmus was confirmed by differential pacing. Subsequent attempts to reinduce the initial atrial tachycardia by burst pacing from the RA and coronary sinus failed and no stable monomorphic atrial tachycardia could be induced. Interestingly, it was quite easy to manipulate the ablation catheter and maintain its stable position during RF energy delivery in the area of interest; the catheter formed a natural curve between the right subclavian vein access site and the lateral RA wall isthmus.

The patient was discharged the next day without any antiarrhythmic medication. She was not a candidate and the lateral RA wall isthmus.

Discussion

Catheter access to cardiac chambers for RFA of cardiac arrhythmias is obtained via the femoral vein and IVC in most cases. Interruption or obstruction due to one of several acquired or congenital causes occurs infrequently, but can occasionally hinder catheter access during EP procedures. This seems to occur most frequently in the setting of congenital IVC interruption with azygos continuation. RFA can still be successfully accomplished using either the subclavian or internal jugular vein to access the RA, or using the femoral vein–azygos vein route.

Successful RF ablation despite lack of IVC access to the heart has been reported for accessory pathways, typical AV nodal reentrant tachycardia, junctional tachycardia related to the presence of two AV nodes, typical atrial flutter, focal ectopic atrial tachycardia, and for pulmonary vein isolation to treat atrial fibrillation. Here, we report successful RF ablation of post-incisional atrial flutter due to prior open heart surgery, through a subclavian vein approach in a patient with IVC interruption. To the best of our knowledge, such a case has not been reported before, although RF ablation of reentrant left atrial tachycardia due to prior pulmonary vein isolation and procedure-related IVC thrombus has been described.

In our patient, the cause of the IVC interruption was most likely congenital, although the large hepatic AVM related to HHT could possibly have caused IVC occlusion by external compression. This patient did not have a well developed azygos continuation of the IVC, and subclavian (or internal jugular) access
was necessary. From a technical perspective, the catheter manipulation along the lateral wall of the RA was very easy. This approach appears to be well suited to target the lateral RA wall.

Figure 2. A. The venous portion of a large hepatic arteriovenous malformation (AVM) is back-filled by a selective contrast injection through a pigtail catheter advanced to the inferior vena cava (IVC). B. Contrast injection to the femoral vein sheath fills the IVC and extensive hepatic collaterals, which bypass the hepatic IVC segment and empty into the right atrium (RA) (arrow). The IVC itself is interrupted (arrowhead) above the AVM. C-E. coronal reconstruction of a contrast CT scan showing the intrahepatic venous collaterals (arrowhead) opening into the RA and the tortuous vessels of the arterial portion of the AVM (thin arrow). The portal vein (thick arrow) is labeled in E. Note the large bowel malrotation.
Regarding the clinical presentation, it appears very likely that the presumed “sinus tachycardia” diagnosed several months earlier actually represented 2:1 conduction of post-incisional flutter. Increased sympathetic tone during epistaxis was likely responsible for the transient 1:1 AV conduction in the emergency department.

Figure 3. A. Activation map of atrial tachycardia (AT); right lateral view of the right atrium (RA). B. Left anterior oblique view of the catheters used for the electrophysiological study and mapping. C. Clean termination of post-incisional flutter with the second radiofrequency (RF) lesion (red arrows – site of successful RF application; black arrow – onset of ablation). Grey tag – scar; pink tag – double potentials; cyan tag – fractionated signals.
HHT is a rare autosomal dominant vascular disorder with prevalence of 1:5000-8000. Endoglin gene (ENG) mutation is responsible for HHT1, activin receptor-like kinase (ACVRL1) is responsible for HHT2, and SMAD4 gene mutation is responsible for the rare syndrome combining HHT with juvenile polyposis (JPHT). Mutations in these three genes are responsible for almost all HHT cases. HHT may present with heart failure or pulmonary hypertension. Heart failure in HHT is commonly a high-output heart failure due to systemic shunt related to an AVM. Management options include embolization of the AVM, hepatic transplantation, and bevacizumab. Bevacizumab, a monoclonal antibody inhibiting the activity of vascular endothelial growth factor, was originally approved for the treatment of metastatic colon cancer. In a recent French study, 20/24 HHT patients with a large hepatic AVM and high cardiac output showed either complete or partial normalization of the cardiac index. At a dose of 5 mg/kg body weight every two weeks for a total of six doses, bevacizumab has resulted in a decrease in cardiac index to completely normal values in ~21% of cases, and a decreased cardiac index in ~65% of cases, while it failed to be effective in ~13% of cases after six months. It also decreased New York Heart Association dyspnea and pulmonary hypertension.

Tranexamic acid upregulates activin receptor-like kinase 1 expression and the endoglin pathway. It has shown efficacy in epistaxis secondary to HHT. Its role in the management of HF, either as single agent or in combination with bevacizumab, has not been evaluated.

This case report describes an unusual combination of post-incisional atrial flutter and IVC abnormality, which required non-standard vascular access for RF ablation. This patient’s response to bevacizumab treatment supports the role of this medication in HHT-associated high-output heart failure.

References