Clinical Scenarios: My Best Case, My Worst Case

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Introduction

In upcoming issues of the Journal, a new series entitled “Best and Worst Cases” will showcase clinical cases that represent successful outcomes and cases that may be associated with complication or poorer outcomes. This unique series will illustrate the highs and lows that occur when caring for complex patients, and demonstrate optimal clinical decision making strategies that enable us to turn our “worst case” into our “best case”.

To begin the series, I will present a case that represents both such scenarios in a single patient.

Presentation of a case

A 50-year-old Caucasian female was referred to me for atrial fibrillation (AF). She had been previously healthy, and had been a competitive figure skater as a young adult. She had a several-year history of drug-refractory symptomatic persistent AF. She had a history of acquired atrioventricular (AV) block for which a dual-chamber pacemaker had been implanted several years before. Her pacemaker interrogations had been demonstrating increasing AF burden corresponding to increasing symptoms of fatigue and exercise intolerance. She otherwise denied chest pain, orthopnea, pedal edema, syncope, or near syncope. She was taking propafenone and warfarin. Her family history was remarkable as her father suffered sudden cardiac death (SCD) in his 50s. Her social history was unremarkable. She works as a schoolteacher, and denied smoking or excessive alcohol consumption.

On physical examination, she was a well-developed Caucasian female in no acute distress. Her examination was unremarkable except for a well-healed pacemaker in the left infraclavicular region. Her laboratories were unremarkable, and her international normalized ratio (INR) was therapeutic at 2.3. Her electrocardiogram (ECG) revealed AF with ventricular pacing. A chest X-ray revealed a dual-chamber pacemaker with right ventricular lead in the midseptum and atrial lead in the right atrial appendage. An echocardiogram revealed a left ventricular ejection fraction (LVEF) of 45% with normal left ventricular size with global mild hypokinesis and mild left atrial enlargement. A nuclear stress test was performed without any evidence of reversible ischemia.

My best case

The patient was referred for catheter ablation for AF. She underwent ablation using a WACA (wide area circumferential ablation) approach with pulmonary antral encircling, roof line, mitral valve isthmus line, and cavotricuspid isthmus line (Figure 1). The patient converted to atrial tachycardia and then to sinus rhythm during ablation. The patient remained in sinus rhythm. Afterwards, the patient underwent pacemaker upgrade to cardiac resynchronization therapy pacemaker (CRT-P) device (Figure 2) without incident. The patient did well postoperatively and was discharged home on postoperative day (POD) 1.

My worst case

The patient returned in a follow-up visit with complaints of palpitations almost 3 years after her ablation for AF. She also reported an increase in dyspnea on exertion.
Device interrogation revealed AV sequential pacing with frequent premature ventricular contractions (PVCs). A repeat echocardiogram showed again mild hypokinesis with a LVEF of 45%. Her symptoms increased and she was admitted for chest pain and dyspnea. An angiogram was performed and revealed a short left main and normal epicardial coronary arteries. Her chest pain correlated with PVCs and salvos of non-sustained ventricular tachycardia (NSVT) (longest 4 beats). She was discharged home on heart failure therapy including carvedilol and enalapril. She was again admitted with worsening chest pain and seen to have increasing salvos of NSVT suggestive of a left ventricular outflow tract (LVOT) focus. She underwent electrophysiology study (EPS) with planned ablation. The procedure was done under conscious sedation, and three-dimensional (3D) mapping was performed of the right and left ventricular outflow tracts. A retrograde transaortic approach was utilized for left ventricular access. Mapping revealed the earliest activation at the anterior subvalvular LVOT and ablation was performed with a closed-loop irrigated catheter with suppression of the PVC focus. However, after several minutes, a unifocal PVC of a slightly different morphology was seen. A repeat map was to be performed. During an attempt to prolapse the catheter across the aortic valve, the catheter was seen to enter the left coronary artery deeply. The catheter was removed, and mapping was continued. However, several minutes later the patient started complaining of chest pain. Acute ECG changes were seen with hyperacute T-wave changes and QRS widening and the patient suffered a ventricular fibrillation (VF) arrest.

My best case

The patient was defibrillated multiple times and CPR was initiated. Acute angiography revealed left main coronary distal closure at the bifurcation. As no interventionalist was in the catheterization laboratory, both the left anterior descending (LAD) and left circumflex (LCx) arteries were rapidly wired and acute angioplasty was performed in the left main-LAD and left main-LCx arteries. This resulted in prompt cessation of VF and complete resolution of the ST-T wave changes. However, continued observation revealed recoil at the bifurcation of both vessels despite intracoronary nitroglycerin and repeated angioplasty. Interventional cardiology support was obtained and recommended stenting of the left main-LAD, which was performed. The patient remained stable after intervention. Serial troponin measurements were normal, and echocardiography revealed no wall motion abnormalities (Figure 3).

Figure 1: Anteroposterior (AP) (a) and posteroanterior (PA) (b) of three-dimensional ablation map for atrial fibrillation.

Figure 2: Fluoroscopy of cardiac resynchronization therapy pacemaker system after device upgrade. The patient was followed closely as an outpatient. Pacemaker interrogation revealed no further atrial fibrillation (AF), and propafenone was discontinued after 1 month. The patient remained free of AF at 3 months, and a repeat echocardiogram revealed improvement in left ventricular ejection fraction to 55%. The patient reported improved exercise tolerance and energy. The patient continued to do well on quarterly follow-up with no recurrent AF for approximately 2 years.

Figure 3:
My worst case

After the procedure, the patient’s family (mother and brother) were informed about the events/complication. The patient’s brother conveyed that their family had an extensive cardiac history. A detailed family history was obtained which revealed a familial pattern of SCD, cardiomyopathy, AF, and pacemaker implantation (Figure 4). The patient’s family had been characterized as an unknown familial cardiomyopathy in the 1980s. However, no underlying mechanism was elucidated at that time.

As the patient exhibited AV block, AF, and cardiomyopathy, genetic testing was recommended. A genetic familial cardiomyopathy panel revealed a lamin A/C mutation.

As the patient was seen to have a familial cardiomyopathy and high risk for sudden death, the patient’s pacemaker was upgraded to a biventricular implantable cardioverter-defibrillator (ICD). She did well after discharge until presenting again with now longer salvos of NSVT. She underwent repeat EPS and ablation, with 3D mapping revealing more extensive scarring in the posterior LVOT. Ablation in this region resulted in elimination of PVCs and no inducible ventricular tachycardia (VT) (Figure 5).

My best case

The patient remained stable clinically after VT ablation and device upgrade without evidence of further VT. The patient continued to be followed over the next several years. However, the patient eventually was seen to develop atrial standstill and progressive right heart failure with declining exercise tolerance and persistent systolic dysfunction with some progressive left ventricular dilatation. The LVEF at this time was 40%. Surveillance of any active ischemia was performed regularly, and repeat angiography revealed normal coronary blood flow and widely patent LM-LAD stent with good flow in the LCx artery. She was maintained on both oral warfarin and clopidogrel.

The patient was referred to an advanced heart failure program over this time period. With progression of her symptoms and declining ventricular function, the patient was listed for cardiac transplantation. The patient recently underwent successful transplantation and has recovered well.

Discussion

This case hopefully illustrates the “ups and downs” of caring for a complex patient, exhibiting both periods of triumph and of utter devastation. I hope that the reader understands the importance of the commitment we make in caring for our patients. The bond associated with the patient–doctor relationship is a sacred one, and the need
for giving our absolute best during that commitment should not be taken for granted. Moreover, our patients will continue to look to us for guidance and treatment even after a devastating complication. As a clinician, we should meet our “worst cases” head-on. As a caring physician, we should never abandon a patient. Our “worst case” can turn into our “best case.” This case certainly taught me the importance of not trivializing a common reason for consultation. This patient presented first with simply AF and two plausible explanations for a mild cardiomyopathy—persistent AF and chronic ventricular pacing. Resolution of the patient’s cardiomyopathy after correction of AF and initiation of biventricular pacing supported this initial diagnosis. Common things might be common, but uncommon diseases will usually present with typical manifestations of the disease and should not be overlooked. A constellation of findings should prompt the physician to consider a more esoteric diagnosis.

A former mentor once conveyed to me that as an interventional electrophysiologist, we are often characterized not from our routine uneventful procedures but from how we handle our rare (hopefully) complications. Every one of us will experience the sinking feeling of complication occurring. Honest physicians recognize their own complications. How we as proceduralists handle these complications often speaks more to our own resolve and experience than a routine case. Moreover, a healthy respect for anything invasive is an important attribute of every invasive electrophysiologist. Sadly, I clearly fell short in my initial evaluation of this patient by not obtaining a complete family history. This patient’s management may or may not have been different knowing about a familial cardiomyopathy, but certainly better knowledge of an underlying mechanism lends to a better understanding of a long-term prognosis and may affect therapeutic options. This is critically important in clinical electrophysiology, as better understanding of genetically inherited disorders may lead to eventual improvements in how we manage two disease entities at the very core of a clinical electrophysiology practice—AF and SCD.

Coronary artery occlusion has been described as a catastrophic complication associated with catheter ablation of the LVOT. It has most often been associated with inadvertent ablation in the left coronary artery, but has also been described from ablation in the endocardium. Recently, a review of 100 patients with computed tomography angiography revealed close proximity (5 mm or less) of the left main artery to a variety of areas important for ablation including the anterior left atrium, left atrial appendage, right ventricular outflow tract (RVOT), pulmonary artery, and left aortic sinus of Valsalva. Emergent stenting of the left main coronary artery in this situation has also been described. The laminopathies are a family of disorders associated with deficiencies of the nuclear lamina, and have multiple manifestations. The Lamin A/C or LMNA gene is found on chromosome 1q21-q21.3 and is the cause of 0.5–5% of dilated cardiomyopathy. Like our patient, the earliest manifestation is often conduction disturbances, with one series reporting greater than 90% incidence of conduction disturbances with 44% requiring pacemaker implantation in patients over 30 years of age. There is a high incidence of AF and thromboembolic risk. There is a high incidence (60%) of congestive heart failure (CHF) by age 50 years. In addition, these patients are seen to have a high incidence of SCD. In data from an Italian registry, NYHA Class III/IV, history of competitive sports, and mutation type were associated with risk of SCD or ICD discharges. More recently, a series of ICD usage in these patients showed high utilization of life-saving therapy despite...
preserved left ventricular function. Thus, making the diagnosis is critical to employ effective therapy. Given the family history, genetic testing is clearly warranted.

References


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