Case Report

Cardiac Amyloidosis Treated with an Implantable Cardioverter Defibrillator and Subcutaneous Array Lead System: Report of a Case and Literature Review

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ABSTRACT

Background: Preventing ventricular arrhythmias in patients with cardiac amyloidosis is challenging since the amyloid protein deposition in the myocardium may interfere with the normal cardiac electric excitation. Most of these patients succumb to either progressive congestive heart failure, or sudden cardiac death (SCD). Implantable Cardioverter Defibrillator (ICD) offers a near sure means of preventing SCD.

Hypothesis: Myocardial infiltration with amyloid results in elevated defibrillation threshold (DFT). Intraoperative strategies may fail to lower DFT during implantation.

Methods: We present a case of a 64-year-old female who had cardiac amyloidosis, and was successfully treated with an ICD and a subcutaneous array lead system.

Conclusion: A subcutaneous array lead system is useful in reducing the DFT, and can terminate ventricular tachycardia or fibrillation by allowing more energy delivery and efficient defibrillation.

Background

Cardiac involvement in amyloidosis is an important predictor of prognosis. Most of these patients succumb to either progressive congestive heart failure, or sudden cardiac death (SCD). An implantable cardioverter defibrillator (ICD) offers a near sure means of preventing SCD because of ventricular arrhythmias. Myocardial infiltration with amyloid often results in elevated defibrillation threshold (DFT). Intraoperative strategies may fail to lower DFT during implantation. In that case, placement of a subcutaneous electrode array on the left side of the chest can help to lower the elevated DFT in these patients.

Case

A 64-year-old African American woman with systemic amyloidosis (AL) with cardiac and renal involvement was admitted for an acute exacerbation of congestive heart failure. Her past history was significant for hypertension, and idiopathic dilated cardiomyopathy. Extensive workup including coronary angiography done a few months ago was unremarkable. Urinary monoclonal paraproteinemia raised the suspicion for amyloidosis. The AL amyloidosis was diagnosed 2 mo ago with a kidney biopsy. Subsequent noninvasive testing including electrocardiography, echocardiography, and nuclear scintigraphy suggested likely involvement of cardiac tissue. The patient refused endomyocardial biopsy and any chemotherapy for her condition at that time.

She gave a history of multiple episodes of dizziness and palpitation prior to this admission. Her medications included aspirin, atorvastatin, and carvedilol. Transthoracic echocardiography showed an ejection fraction of 15%–20%. Computed tomography of the chest showed severe cardiomegaly with marked left ventricular wall thickening (Figure 1). During her hospital stay, multiple episodes of nonsustained ventricular tachycardia were observed on telemetry. During electrophysiology study (EPS), sustained ventricular tachycardia (VT) was easily inducible.

In view of her low ejection fraction, history of presyncope, and easily inducible VT during EPS, we decided to implant an ICD. After securing right subclavian venous access, a dual chamber ICD (Guidant Vitality DS, Indianapolis, Ind., USA) was placed in the right infra-clavicular region due to the presence of an arteriovenous fistula in the left arm (for hemodialysis for end-stage kidney disease). Pacing mode was VVI, and rate limits were programmed between 40 and 120 beats/min. Lead parameters were checked, which showed sensing amplitude of 8.4, threshold of 0.8, pulse width of 0.5 msec, and impedance of 685 ohms. While testing the DFT of the device, it failed to terminate the induced episode of ventricular fibrillation (VF) at serially delivered internal shocks of 17 and 21 joules. External defibrillation with a 200 J biphasic shock immediately terminated the episode and restored sinus rhythm. Hence, a high-energy output ICD (Guidant Vitality HE) with capacity to deliver shocks at 31 and 41 joules was placed. Again, while testing the device, it failed to terminate the episode of induced VF at 31 and 41 joules and rescue defibrillation had to be performed with an external defibrillator for the second time.

After attempting reversal of lead polarity in an effort to reduce the DFT, it was decided to place a subcutaneous

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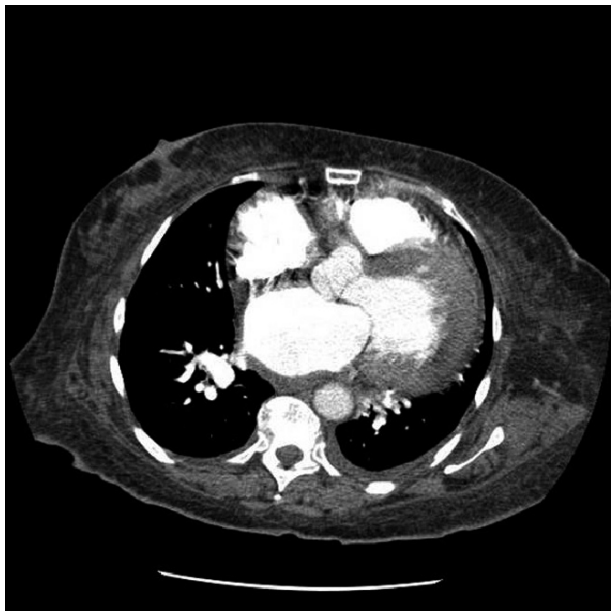

Figure 1. Biventricular enlargement due to massive deposition of Amyloid in heart on computed tomography.

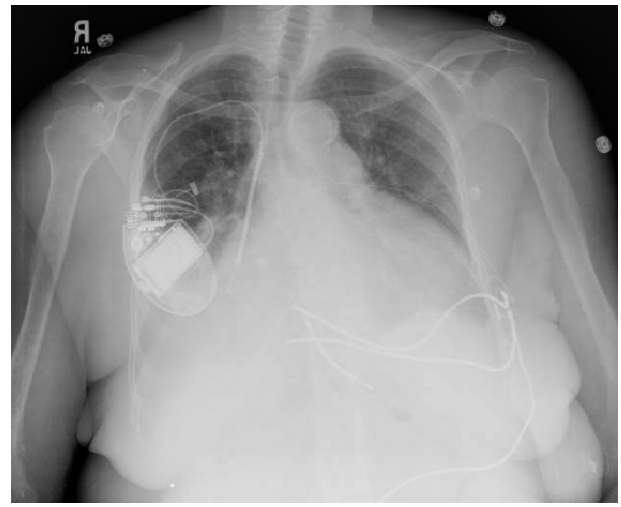

Figure 2. Chest X ray - Implantable cardioverter defibrillator with subcutaneous lead.

array lead system to lower the DFT. The 3 leads of the Phase Array Subcutaneous Tissue System were placed via a subcutaneous tunnel by the surgical team. The electrode array was then connected to the proximal port of the ICD (Figure 2). Upon rechecking the DFT, induced VF could be successfully terminated by the high output ICD by an internal shock of 31 J. This internal defibrillation ability of the device was verified by retesting. The device was finally programmed for VT as a monitoring zone of 175 to 220 bpm, and a therapy zone consisting of ATP followed by incremental 11, 21, 31 followed by 41 J shocks. For VF, therapy was programmed at 31 J followed by 41 J shocks.

There were no complications during the procedure. Incision sites healed in a timely manner. The patient was fairly regular in keeping up with the appointments for the next 2 y, and was then lost to follow-up. Contrary to the reported median survival period, our patient survived for a longer time. During that period, the ICD successfully delivered seven 31 J shocks in response to ventricular tachycardia.

Discussion

Amyloidosis is a systemic disorder characterized by extracellular deposition of insoluble homogeneous material (amyloid), throughout various tissues of the body. Infiltration of the heart by amyloid proteins results in biventricular wall thickening, resulting in restrictive cardiomyopathy manifesting late in the course with heart failure and conduction abnormalities.[1] Although sudden death is not uncommon in cardiac amyloidosis, ventricular arrhythmias are an uncommon presenting feature.[2] The reported incidence of SCD in primary amyloidosis is about 10% to 30%.[2,3] Sudden death is often due to electromechanical dissociation.[4] This precludes the use of ICDs in a majority of such patients.

Despite the low incidence, implantable defibrillators are a near sure means of preventing sudden cardiac arrest due to ventricular arrhythmias. There are very few case reports and series which mention the use of ICDs in patients with cardiac amyloidosis. Some patients with amyloidosis may have a high DFT as our patient had. The DFT is the amount of minimal shock needed to terminate VF with at least 1 failure with lower energy shock.[5] The reasons for high DFT in this patient were chronic renal failure and biventricular thickening due to amyloid protein deposition in the myocardium. Patients with high DFTs represent a small, but significant population among those with ICDs. Suggested management options include using a high output device, reversing polarity of the leads, removing SVC coil especially if impedance is $<40\ \Omega$, adding an additional coil, treating empirically with sotalol, or adding a subcutaneous (SQ) electrode array.[6] In our patient, after trying some of these options, a decision was made to implant an SQ electrode array system.

The SQ array is a common strategy used for lowering DFTs. Another strategy is to use a subcutaneous patch electrode. Implantation of an SQ array is shown to be more effective, and associated with fewer complications compared with a patch electrode.[7–10] The SQ array is usually placed through a tunnel made at the left lateral chest wall. In the event of inadequate lowering of DFT, a second SQ array can be placed inferior to the first and connected with a Y-connector.[6] With the help of the SQ array electrodes, the defibrillation field is significantly enlarged posteriorly. This

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helps encompass the septum and the left ventricular free wall in the defibrillation field, leading to more widespread current distribution.[11,12] Exact positioning of the SQ array is less critical than the positioning of a patch electrode.[11] In our patient, the ICD was placed on the right side of the chest because of the presence of a fistula in the left forearm.

The use of an SQ array to lower the DFT in patients with cardiac amyloidosis following ICD implantation has not been described before. This case illustrates the novel use of an already known strategy to lower elevated DFT. Our patient has subsequently done well at 2 y postprocedure. She did not experience any short-term or long-term complications from the procedure. We therefore suggest considering this modality of treatment in patients with cardiac amyloidosis and elevated defibrillation thresholds.

Conclusion

Preventing SCD in patients with cardiac amyloidosis is challenging since the amyloid protein deposition in the myocardium may interfere with the normal cardiac electric excitation. The ICDs remain the mainstay of therapy for preventing SCD in these patients, if not cardiac transplantation. Because of the massive deposition of amyloid protein, DFT may be elevated and a standard ICD may be unable to terminate the arrhythmias. A subcutaneous array lead system is useful in reducing the DFT, and can terminate ventricular tachycardia or fibrillation by allowing more energy delivery and efficient defibrillation.

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