Non-sustained Ventricular Tachycardia in a Patient With Non-compaction of Left Ventricle: Natural History or Associated Condition?

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ABSTRACT. Left ventricular non-compaction (LVNC) is frequently associated with ventricular arrhythmias. However, there have been few reports of radiofrequency ablation therapy in patients with this condition. We describe a 58-year-old man with symptoms of congestive heart failure for several months. Echocardiogram revealed the presence of LVNC in the lateral wall of the left ventricle, which was confirmed by magnetic resonance imaging. The ejection fraction (EF) was calculated at 28%. Further evaluation revealed frequent premature ventricular contractions (PVCs) on Holter (23% of the total heart beats recorded, 3,410 ventricular couplets, 653 triplets, and 676 episodes of non-sustained ventricular tachycardia (VT)). The patient underwent electrophysiology study (EPS), and the PVCs were mapped. The earliest site of activation was identified at the left ventricular outflow tract (LVOT). Radiofrequency pulses were applied in the area, resulting in dramatic reduction in the frequency of PVCs in a long-term follow-up, confirmed by Holter monitoring. The echocardiographic evaluation showed improvement in EF, from 28% to 57%. This case report describes the association of LVNC and ventricular arrhythmia and severe left ventricular (LV) dysfunction. The focus of arrhythmia in this patient was not necessarily related to LVNC, indicating the possibility of concomitant "idiopathic" arrhythmias in this population. The presence of LV dysfunction was mostly a presentation of tachycardia-induced cardiomyopathy. Several reports have shown a high prevalence of non-sustained VT among LVNC patients. We should increase our awareness about the possibility of a component of tachycardia (PVC)-induced cardiomyopathy in these patients, which is a potentially reversible cause of systolic LV dysfunction.

KEYWORDS. non-compaction, radiofrequency ablation, ventricular arrhythmia.

Introduction

Left ventricular non-compaction (LVNC) was first described in 1984,1 and it has been associated with congestive heart failure, ventricular ectopy (premature ventricular contactions (PVCs)), ventricular tachycardia (VT), and embolic events. Despite the high prevalence of VT in this population, there have been few reports of radiofrequency ablation therapy in patients with LVNC. We describe a case of left ventricular outflow tract (LVOT) tachycardia in the setting of LVNC with severe ventricular dysfunction that was successfully treated with radiofrequency ablation, with subsequent significant improvement in ventricular function.

Case presentation

A 58-year-old man presented for outpatient evaluation of symptoms of progressive dyspnea and palpitations for several months. His past medical history was significant for a ventricular septal defect (VSD) detected during childhood, which closed spontaneously leaving a residual aneurysm in the membranous septum. He had no history of syncope or neuromuscular disease. His family history was unremarkable for cardiac disease or sudden cardiac death. The physical examination revealed a healthy looking
middle-aged man. The blood pressure was normal in both upper extremities. His pulses were irregular, but full and symmetric. His lungs and cardiac auscultation were normal. Initial testing included an electrocardiogram showing sinus bradycardia, left anterior fascicular block, and voltage criteria for left ventricular hypertrophy. A 24-h Holter test showed frequent ventricular arrhythmias, including 21,993 PVCs, representing 23% of the total heartbeats recorded, 3,410 ventricular couplets, 653 triplets, and 676 episodes of non-sustained VT. Invasive and non-invasive studies ruled out the presence of significant coronary atherosclerosis, myocardial ischemia, or myocardial infarction. Echocardiographic evaluation revealed the presence of LVNC with a reduced ejection fraction (EF) in the range 28–32%. This finding was confirmed by gated magnetic resonance imaging, showing moderate non-compaction of the lateral wall of the left ventricle extending to the apex (Figure 1).

After 5 months of beta-blocker and angiotensin-converting enzyme inhibitor therapy the EF mildly improved to 46%, but the patient remained symptomatic with a large burden of ventricular arrhythmias. An electrophysiological study (EPS) was performed, demonstrating frequent spontaneous monomorphic PVCs exhibiting a left bundle inferior axis morphology. Mapping of the LVOT showed a good pace match (9 leads out of 12), and an activation map (Ensite Navx, St Jude Medical, St. Paul, MN) showed that the earliest site of activation was located just inferior to the aortic valve, and adjacent to the mitral-aortic intervalvular fibrosa (Figure 2). During PVCs, the local electrogram was 30 ms earlier than the onset of the surface QRS deflection, and a sharp presystolic potential was identified in this area (Figure 2a). Using a 4-mm-tip ablation catheter, three pulses of radiofrequency were applied, resulting in complete suppression of the PVCs (Figure 3). Two additional 60-s “insurance” pulses were

Figure 1: Non-compaction visualized on lateral wall of left ventricle, extending to the apex. Transthoracic echocardiogram images from (a) apical long axis and (b) parasternal short axis views; (c, d) MRI images demonstrating non-compaction myocardium in lateral wall (arrows).
applied in the region. All radiofrequency applications were delivered for 60 s with a power of 50 watts. The mean and maximum temperatures achieved with ablation were 55°C and 75°C, respectively. The mean and maximum power achieved was 27 and 50 watts, respectively. There were no complications after ablation.

At follow up, the patient improved clinically, with complete resolution of symptoms. Repeat echocardiographic evaluation performed 1 year after ablation showed an increase in EF to 57%, and a repeat 24-h Holter monitoring only showed a trivial number of isolated PVCs.

Discussion

Non-compaction of the left ventricle is a congenital anomaly caused by intrauterine arrest of compaction of myocardial fibers and meshwork, resulting in multiple trabeculations in the left ventricular myocardium. The prevalence of LVNC in one large series of adult patients was 0.014%. However, since this study was performed in a tertiary referral center, this may represent an over-estimation of the real prevalence of this condition. The most common clinical presentations of LVNC are congestive heart failure, thromboembolic events and
ventricular arrhythmias. Oeschslin et al. described an incidence of VT of 41% (14 patients out of 34 individuals with LVNC), mostly with non-sustained VT. Only 3 out of 14 patients had sustained episodes, and received an implantable cardioverter-defibrillator. None of these patients underwent radiofrequency ablation. Lofiego et al. also followed a large series of patients with LVNC, and they reported an incidence of sustained VT in 6% of the population. The authors did not mention the incidence of non-sustained VT, and that most likely explains the discrepancy between these two large cohorts of patients with LVNC. Murphy et al. identified 45 patients with LVNC in a referral center for cardiomyopathy, and non-sustained VT was detected on Holter monitoring in 22% of their population. The authors did not mention any event of sustained VT in this cohort. Fazio et al. followed 238 patients with isolated LVNC for about 4 years, and only 11 patients had VT documented on Holter (only 2 of them with sustained VT). Based on this very low prevalence of ventricular arrhythmias, Fazio, in his discussion, questions the general concept of high risk of arrhythmias associated with this condition, described as high as 50% of patients having fatal events.

These limited studies that reported non-sustained VT in patients with LVNC did not mention further invasive and non-invasive studies for determination of their type or origin. Our patient underwent an EPS secondary to his drug-refractory symptoms of palpitations and because of a significant drop in LVEF associated with a large PVC and VT burden. This invasive study revealed a LVOT focus, which was successfully ablated.

There are limited data in the current literature about mapping and ablation of PVCs or VT in the presence of LVNC. Fiala et al. described a successful ablation of VT originating in the interventricular septum. Derval et al. reported two symptomatic patients who had ventricular arrhythmias with subtle evidence of LVNC, one of whom had successful radiofrequency ablation of a monomorphic VT in the basolateral aspect of the left ventricle. Finally, Lim et al. also described a successful epicardial ablation of a monomorphic VT in a patient with LVNC, localized in the epicardial surface of the anterolateral wall.

Currently, the mechanism underlying these arrhythmias is not yet completely understood. There is evidence of perfusion defects in the areas of non-compaction, as previously demonstrated by MRI and positron emission testing. Paparella et al. reconstructed a ventricular electroanatomical mapping in a patient with LVNC and monomorphic VT. Despite detailed mapping, no areas of low voltages were identified, likely excluding the presence of scar-based tissue re-entry as a possible substrate. Steffel et al. reviewed the electrophysiological data of 24 patients with LVNC. The indications for EPS in this series were very diverse, with only two patients exhibiting documented non-sustained VT and only two patients with sustained VT. Nine patients had inducible VT, two of them were monomorphic and two had induced ventricular fibrillation. Right ventricular outflow tract tachycardia was induced in one of these patients. None of these patients had LVOT VT.

On the basis of the echocardiographic and MRI findings, our patient had the origin of his ventricular arrhythmia in the LVOT, in an area not directly affected by the non-compaction of the endocardial layer. One could argue that this patient also had a residual ventricular septal aneurysm after spontaneously closing a ventricular septal defect, which could function as an arrhythmogenic substrate. However, the site of ablation was close to the mitro-aortic intervalvular fibrosa and not related to the residual aneurysm.

To the best of our knowledge, this is the first description of an association of LVNC and LVOT tachycardia. As mentioned previously, the prevalence of non-sustained VT is significantly high in patients with LVNC, although no prior studies have provided a more detailed description of these arrhythmic events. Therefore, it is possible that the association described here is not as rare as intuitively considered. The mechanism of these two conditions is not well understood. LVNC is most likely secondary to an arrest of the normal compaction of the cardiac muscle, which should be completed between the fifth and eighth week of gestation. There have been suggestions that LVNC may be associated with different gene mutations, including taffazin, b-dystrobrevin, cipher/ZASP, lamin A/C, SCN5A, MYH7, and MYBPC3. Despite advanced knowledge about the genetic mutations related to LVNC, several questions still remain, including the differences in prevalence and severity of this condition between pediatric and adult populations, and the occurrence of both familial and sporadic cases of this condition. The origin of LVOT VT is also not well understood. It is well known that during embryogenesis neural crest cells migrate to LVOT, and these cells play an important signaling role in the development of the conduction system. Abnormalities during this phase of embryology could potentially affect the electrophysiologic substrate in that area. We could speculate that these two embryologic abnormalities may be related to each other, but further studies will be necessary to better determine the real prevalence of this association reported here and elucidate the possible mechanism involved in these cases.

Interestingly, it has been recognized that several forms of congenital structural heart disease, more commonly ventricular septal defect and left ventricular outflow obstruction can accompany LVNC. Other congenital anomalies have also been identified in association with LVNC, including pulmonary atresia, tricuspid atresia, and anomalous left coronary artery. The term non-isolated left ventricular non-compaction (ni-LVNC) is used when at least one of these congenital abnormalities is present. ni-LVNC is more commonly seen in a pediatric population and it most likely has a different type of genetic mutation. Our patient had a ventricular septal defect, which categorizes him as ni-LVNC.

Left ventricular systolic dysfunction is very common among patients with LVNC. Our patient presented initially with EF between 28% and 32%. It was surprising, however, that his left ventricular function improved dramatically after the ablation procedure.
His improvement in cardiac function made us conclude that his left ventricular dysfunction was mostly PVC-related cardiomyopathy, and not necessarily related to LVNC. It is also possible that the underlying LVNC makes one more susceptible to develop this type of ventricular arrhythmia-induced cardiomyopathy.

In summary, we present a case where the diagnosis of LVNC was made in association with ventricular arrhythmia and severe left ventricular dysfunction. Electrophysiologic study and ablation could be performed safely and successfully in the presence of LVNC. The focus of arrhythmia in this patient was not necessarily related to LVNC, indicating the possibility of concomitant “idiopathic” arrhythmias in this population. The presence of left ventricular dysfunction was mostly secondary to frequent episodes of non-sustained VT, and not related to LVNC. Several reports have shown a high prevalence of non-sustained VT among LVNC patients. We should increase our awareness about the possibility of a component of (PVC)-induced cardiomyopathy in these patients, which is a potentially reversible cause of systolic left ventricular dysfunction.

References