Ventricular Tachycardia in a Patient with Mitral Valve Prolapse

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3.1 Case Summary

The patient is a 58-year-old man with mitral valve prolapse (MVP) and a non-ischemic cardiomyopathy with a left ventricular ejection fraction of 45–50%. A single chamber defibrillator (ICD) was implanted 8 years before because of frequent multiform ventricular ectopy and inducible polymorphic ventricular tachycardia (VT). Cardiac magnetic resonance imaging at that time showed no evidence of myocardial scar. One year after implantation, he had an appropriate ICD discharge for ventricular fibrillation and he started amiodarone. Six years later, while taking amiodarone, he developed recurrent episodes of monomorphic VT, including episodes of VT storm treated with antitachycardia pacing and high voltage shocks. He was found to have severe mitral regurgitation with prolapse of the posterior leaflet. He underwent robotic mitral valve repair and concomitant bilateral sympathetic denervation. He continued to have VT after surgery, and he presented for a catheter ablation. What is the pathogenesis of ventricular arrhythmias in this case, and what structures are expected targets for ablation?

3.2 Case Discussion

MVP is associated with complex ventricular arrhythmias in a subset of patients, who may develop frequent ventricular ectopy and polymorphic or monomorphic VT. MVP is sometimes documented in young patients who have otherwise unexplained sudden death and is implicated as a cause of cardiac arrest. Risk factors for malignant ventricular arrhythmias in MVP syndrome have been identified, including bileaflet prolapse, female gender, repolarization abnormalities, and myocardial fibrosis. Imaging and post-mortem studies in MVP patients with malignant arrhythmias have recently identified fibrosis in the papillary muscles or the subvalvular myocardium, particularly the inferobasal wall, adjacent to the papillary muscles [1–4].

In this case, the patient originally had polymorphic VT but he developed recurrent episodes of monomorphic VT after being treated for several years with amiodarone. This change in VT morphology could be attributed to the effects of amiodarone in slowing conduction and prolonging refractoriness of the myocardium, and thus promoting a stable reentrant circuit. Alternatively, the patient may have developed new substrate in the form of myocardial fibrosis.

Before ablation, a transesophageal echocardiogram identified thinning and dyskinesis of the basal anterolateral wall below the mitral annulus, adjacent to the anterolateral papillary muscle (Video 3.1). Electroanatomical mapping identified patchy areas of low voltage in this region (Fig. 3.1). Abnormal electrograms during sinus rhythm, characterized by fragmentation and late potentials, were also identified in this area (Fig. 3.2). The predominant inducible VT demonstrated a cycle length of 480 ms, right bundle branch (RBBB) pattern, and right inferior axis with positive precordial concordance (Fig. 3.3). Mapping of this tachycardia revealed diastolic electrograms between the anterolateral papillary muscle

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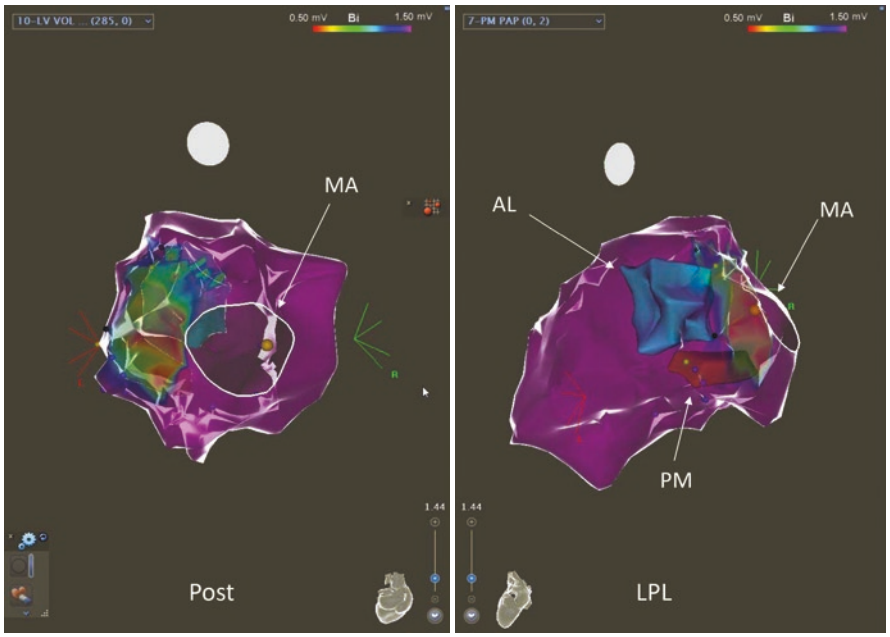

Fig. 3.1 Electroanatomical voltage map of the left ventricle constructed with fast anatomical mapping (FAM®) and superimposed on CartoSound® contours of the papillary muscles. The map shows an area of low voltage in the basal lateral left ventricle. The anterolateral papillary muscle (AL) is indicated in blue and the posteromedial (PM) papillary muscle in orange. *MA* indicates mitral annulus, *Post* posterior, *LPL* left posterolateral

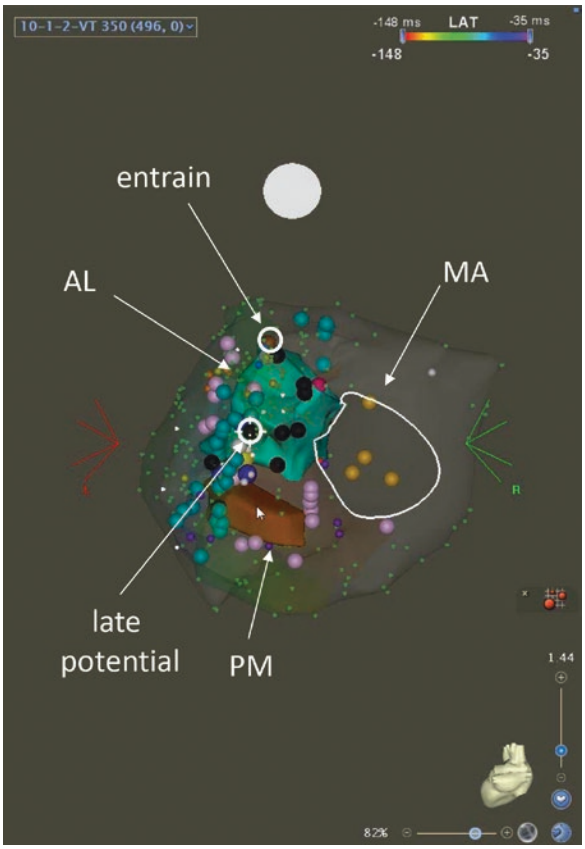

Fig. 3.2 Electroanatomical voltage map in a posterior projection shows the anterolateral (AL) and posteromedial (PM) papillary muscles. Back tags indicate sites with local abnormal ventricular activities (LAVAs) in sinus rhythm. Pink tags indicate sites with fragmented diastolic potentials during ventricular tachycardia (VT) and blue tags are sites with double potentials during VT. Brown tags are pacing sites, either entrainment during VT or pace map sites. A site of concealed entrainment consistent with an entrance site is indicated superior to the AL papillary muscle ("entrain"), as shown in Fig. 3.3. An entrance to a protected channel, which was identified by late potentials in sinus rhythm, is indicated between the AL and PM papillary muscles

and the mitral annulus. Figure 3.3 shows concealed entrainment from a basal site adjacent to the anterolateral papillary muscle, demonstrating a long stimulus to QRS interval (65% of the tachycardia cycle length) and post-pacing interval nearly equivalent to the tachycardia cycle length, consistent with a proximal isthmus site within the tachycardia circuit.

Two other VT morphologies were inducible (RBBB, indeterminate axis, cycle length 310 ms; RBBB, indeterminate axis, cycle length 510 ms, which terminated spontaneously). Ablation was performed in sinus rhythm to target components of the circuit identified with entrainment mapping and also electrograms within the abnormal region which showed local abnormal ventricular activities. Figure 3.4a, b shows disappearance of a late potential on the proximal electrodes of the ablation catheter during radiofrequency application, a response consistent with elimination of the entrance to a protected channel. After ablation at the base of the anterolateral papillary muscle, between the anterolateral and posteromedial papillary muscles, and the basal anterolateral wall, the clinical VTs were no longer inducible.

This case illustrates the development of fibrotic substrate in the basal LV in proximity to the papillary muscles and the mitral annulus, which became a substrate for monomorphic VT. In MVP, arrhythmogenicity may develop beyond the papillary muscles per se, as replacement fibrosis involves neighboring myocardial tissue. In patients with otherwise unexplained ventricular arrhythmias, the presence of MVP should focus attention on these specific areas prone to develop fibrosis. Conventional techniques, such as entrainment mapping and substrate modification targeting locally abnormal ventricular electrograms and late potentials, are useful in treating VT in this substrate.

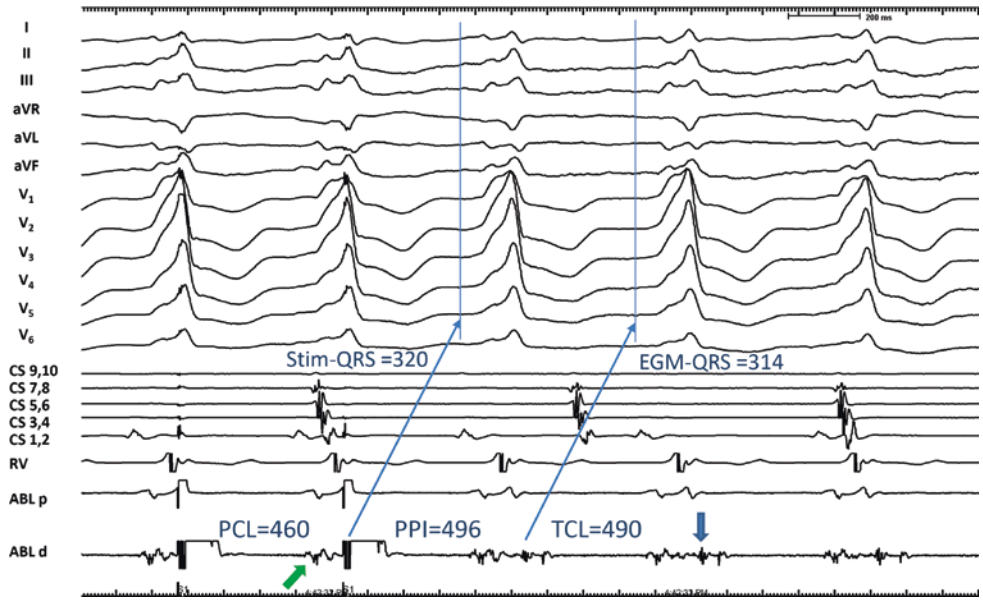

Fig. 3.3 Ventricular tachycardia with RBBB, positive concordance, and inferior axis. Entrainment with concealed fusion is demonstrated from a site superior to the anterolateral papillary muscle (location indicated in Fig. 3.2). The local electrogram is highly fragmented with presystolic components. Of the multiple components of the fragmented electrogram, the deflection indicated with a vertical blue arrow is locally captured by pacing and is used to measure post pacing interval (PPI) and electrogram (EGM) to QRS. The presystolic components of the complex electrogram are activated orthodromically. These far-field potentials are not captured by the pacing stimuli (green arrow). The stimulus to QRS interval is 65% of the tachycardia cycle length and nearly identical to the local EGM to QRS interval. The PPI is also nearly identical to the tachycardia cycle length (TCL). These features are consistent with a proximal isthmus site within the tachycardia circuit. *ABL* indicates ablation, *CS* coronary sinus, *PCL* paced cycle length, *RV* right ventricle, *Stim* stimulus

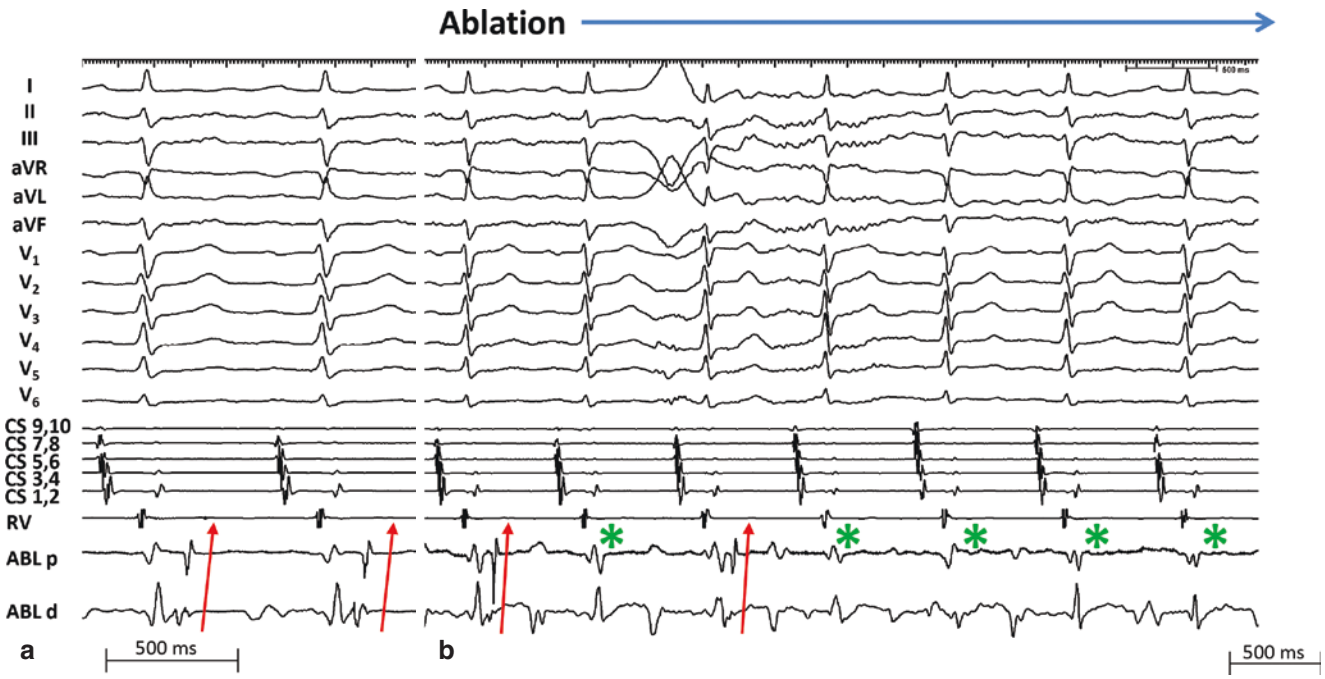

Fig. 3.4 Ablation of entrance to channel between papillary muscles. (**a**) Ablation catheter records late potentials on distal and proximal poles, with later activation on the proximal poles, consistent with activation of a channel from distal to proximal (red arrows). (**b**) During ablation, the proximal late potential (which is identified with arrows) is abolished on beats indicated with the green asterisks

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