Chapter 21
AS and HCM

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Abstract LVH (left ventricular hypertrophy) is one of the most common presentations in aortic stenosis and sometimes it would be missed to evaluate for other causes of the LVH in the presence of AS. Combination of AS and hypertrophic CMP is not common too. In this case of such combination, HCM was missed until presentation with VT and apical aneurysm.

A 36-year-old man with history of aortic valve replacement (AVR) 8 years ago because of severe aortic stenosis (AS) and bicuspid aortic valve (BAV) presented with palpitation and cold sweating with hemodynamically compromised state and wide QRS tachycardia to the Emergency room (ER). Electrocardiography showed a wide QRS tachycardia with RBBB pattern and transition in V2, and superior axis, considering VT with left ventricular (LV) origin (Fig. 21.1, ER ECG was inaccessible so we used the figure of the 12 lead ECG of the induced VT in EP lab).

QRS Synchronized electrical cardioversion was done in the ER with 100 J and resulted in the termination of arrhythmia 0.12 lead ECG revealed LVH in normal sinus rhythm state (NSR).

Echocardiography was done by cardiology assistant and showed LVH with normal functioning prosthetic valve without significant valvular gradient. CMR was recommended because of the discrepancy between LVH and normal function of the prosthetic valve without significant size mismatch. CMR was done with 1.5 Tesla Aventus Siemens device and showed LVH with apical aneurysm and diffuse LGE with mid-cavity gradient, all findings infavour of HCM (Fig. 21.2a, b).

Because of history of sustained monomorphic VT (SMMVT), electrophysiological study (EPS) and mapping was done, using transseptal approach because of the prosthetic aortic valve. Substrate mapping during NSR revealed large low voltage

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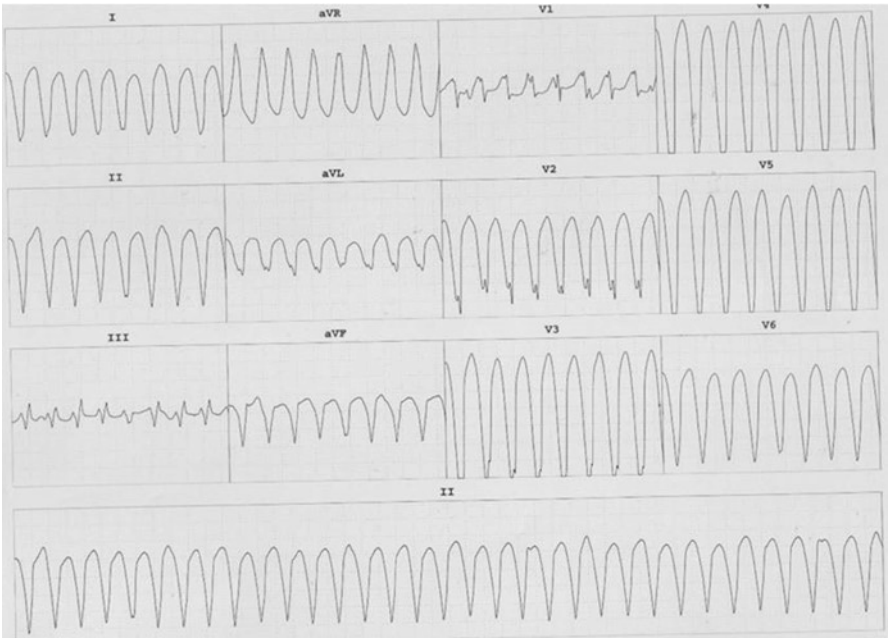

Fig. 21.1 VT with RBBB pattern, transition in V2, superior axis

apical area and activation mapping after VT induction showed reentrant VT around the apical aneurysm.

Radiofrequency ablation (RFA), using a cooled tip catheter with 30 W and 43 °C was done and the apical aneurysm was isolated (Fig. 21.3).

At the end of the procedure, VT was non-inducible. Then because of high-risk features of the patient, ICD was inserted.

Discussion

The coincidence of BAV and HCM was already described by Brown in 1990 in four adult patients [1–5]. The coincidence of HCM with other cardiomyopathies such as left ventricular noncompaction (LVNC) has been previously reported in some studies [6]. Also echocardiography remains now as a gold standard in examination of patients with HCM heart morphology, weak points of echocardiography are the anterolateral segments of the LV, Papillary muscles, Some Portions of the right ventricle (RV) and apex [7, 8]. CMR using gadolinium can reveal areas of fibrosis and also better clarify missing points of echocardiography. However, necrosis can be found in half of the patients suffering from hypertrophy of LV as a consequence of aortic stenosis or arterial hypertension but diffuse fibrosis with an apical aneurysm are seldom findings of BAV and AS, and strongly suggest HCM [9].

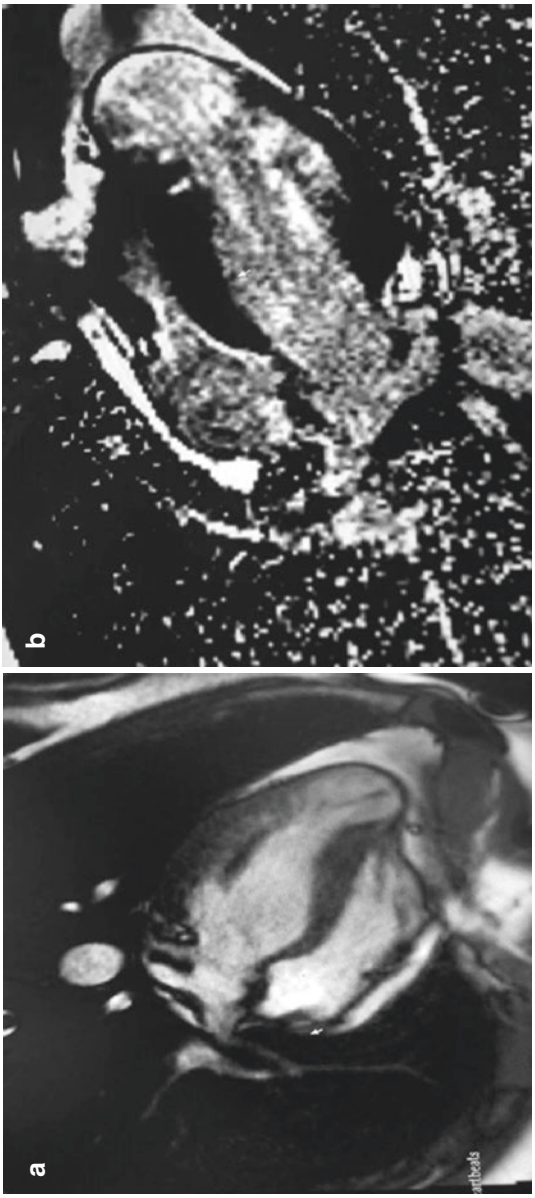

Fig. 21.2 (**a**, **b**) LV apical aneurysm with LGE

Fig. 21.3 Apical aneurysm core isolation using 3D Navx system

Late gadolinium enhancement in CMR in many studies has been suggested to be a high-risk predicting factor [9].

LV apical aneurysm is also a high-risk factor for arrhythmia.

In this patient despite arrhythmia ablation, ICD was implanted, because of high-risk features for sudden cardiac death [10].

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