Left Ventricular Noncompaction (LVNC)

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Left ventricular noncompaction (LVNC), also called spongiform cardiomyopathy, is a rare congenital cardiomyopathy that results from interrupted cardiac embryogenesis. Pathologically, it is characterized by prominent trabeculations and deep intertrabecular recesses, creating a "spongy" myocardium. The cause is thought to be an arrest in the gradual compaction of the loosely interwoven meshwork of myocardial fibers during endomyocardial morphogenesis at 5–8 weeks of fetal life. The condition predominately affects the left ventricle (LV).

Engberding and Bender first described LVNC as an isolated condition in 1984 [1]. Subsequently LVNC has been associated with other congenital anomalies such as obstruction of the right or left ventricular outflow tracts, complex cyanotic congenital heart disease, coronary artery anomalies, hypertrophic and restrictive cardiomyopathies, and recently with ARVD.

Originally reported to be present in only 0.05 % of adults [2], it is more frequently recognized primarily as result of high-resolution images of the left ventricular apex afforded by magnetic resonance imaging (MR) and computed tomography (CT) as compared to echocardiography. More recently, LVNC was reported in 2 % of patients undergoing MDCT for assessment of coronary artery disease [3]. LVNC may be associated with development of systolic and diastolic LV dysfunction, arrhythmias, congestive heart failure, and thromboembolic events. The age of onset and degree of clinical symptoms depend on the extent of the noncompacted cardiac segments [4].

Traditionally, echocardiography has been used to establish the diagnosis of myocardial noncompaction, although CT and MRI provide better visualization of the trabeculations. CT findings of LVNC are prominent trabeculations and deep recesses in the myocardium usually affecting the apical and basal surfaces of the left ventricle. The RV apex also may be involved [5, 6]. Based on MRI criteria, a noncompacted to compacted myocardium ratio (NC/C ratio) of 2:1 at end systole (or 2.3:1 in end diastole in the long axis) supports the diagnosis of LVNC. A recent, small CT study suggests that a ratio of noncompacted to normal myocardium of 2.2:1 in more than one myocardial segment suggests the diagnosis of LVNC [7]. Left ventricular systolic dysfunction and restrictive filling may be seen on cine imaging. CT examples of LVNC are presented in Figs. 9.1 and 9.2.

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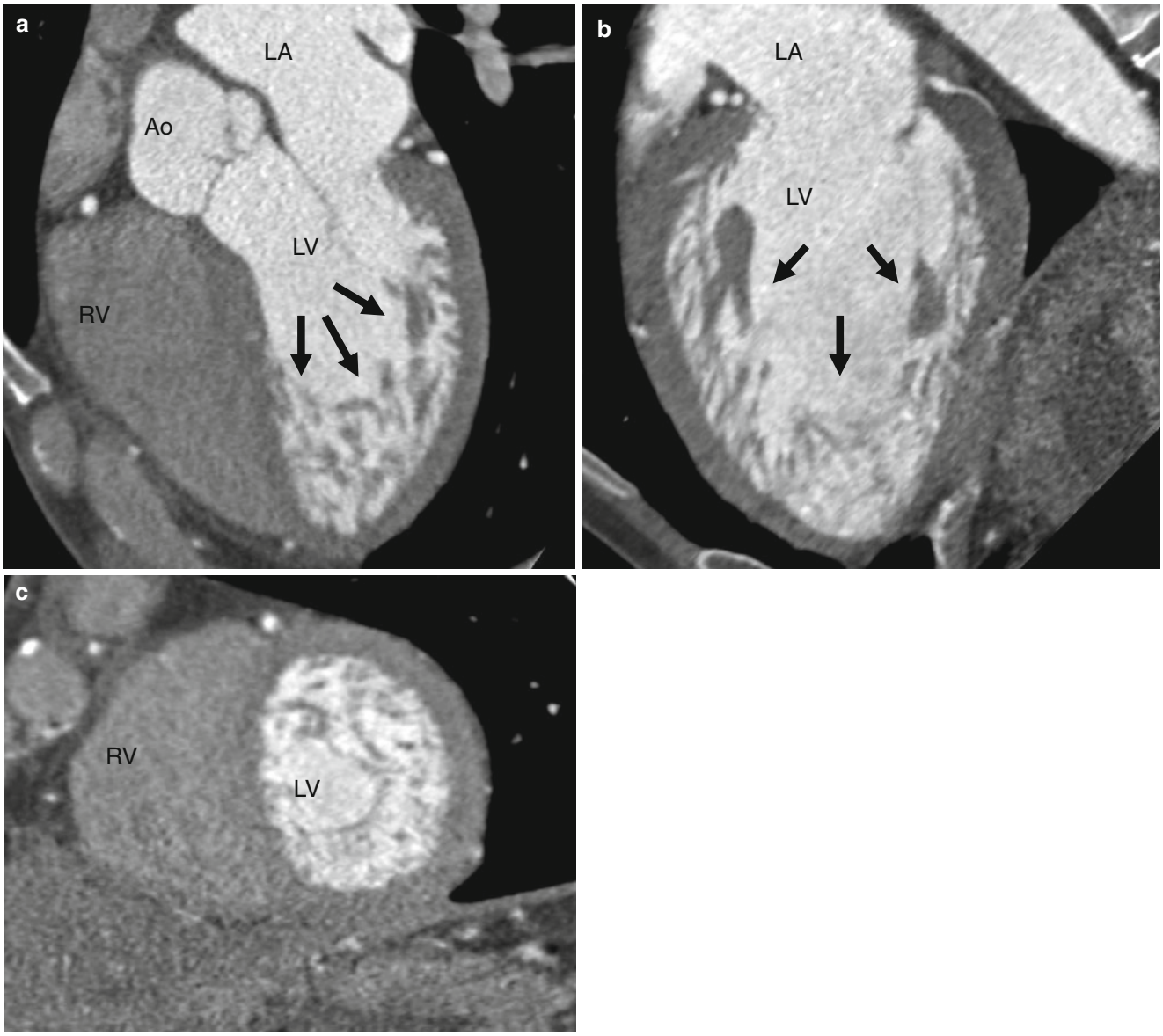

Fig. 9.1 Left ventricular noncompaction (LVNC) in an asymptomatic marathon runner. Panel (**a**) is a five-chamber view, panel (**b**) is a two-chamber orientation, and panel (**c**) is a short-axis cut. Note the dilated left ventricle and the fine trabeculations with deep intertrabecular recesses (*arrows*) in the myocardium on the apical regions of the left ventricle. *LA* left atrium, *LV* left ventricle, *RA* right atrium, *RV* right ventricle, *Ao* aorta

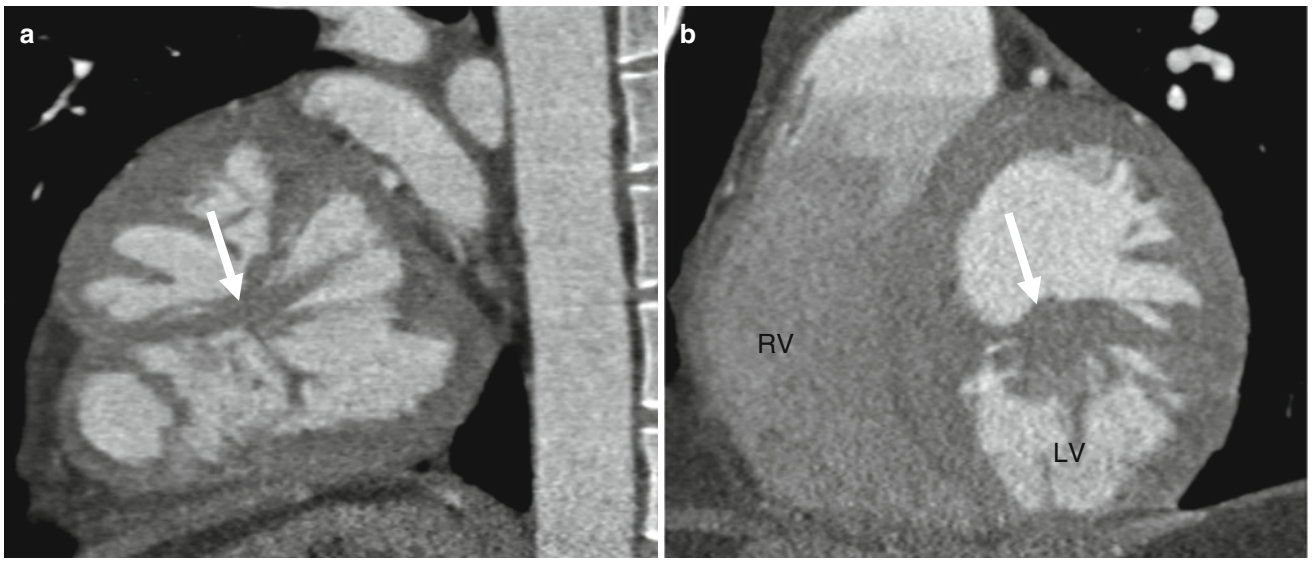

Fig. 9.2 Left ventricular noncompaction (LVNC) in a 22-year-old patient presenting with a new-onset heart failure. Panel (**a**) a long-axis two-chamber computed tomographic (CT) view. Panel (**b**) a short-axis CT image. Note the thickening of the left ventricular wall with very prominent trabeculations and absence of distinct papillary muscles. The hypertrabeculation in the mid-left ventricle (*white arrow*) nearly separates the ventricle into two chambers. Ventricular septation is best appreciated in the short axis (*arrow*, panel b). *LV* left ventricle, *RV* right ventricle

9.1 Differential Diagnoses of LVNC

The differential diagnoses include prominent normal LV trabeculations (normal variant), hypertrophic cardiomyopathy (described in the following section), apical left ventricular thrombus, and endocardial fibroelastosis. However, using the above diagnostic criteria, LVNC is quite distinguishable if a high index of suspicion is maintained in the appropriate clinical presentation.

9.1.1 Normal Variant LV Trabeculations

Prominent LV trabeculations related to the attachment of the papillary muscles to the LV myocardium is a normal variant on CT (see Chap. 3). These trabeculations are few in number and rarely located in the apical region. The scarcity of apical involvement helps to differentiate between LVNC and normal trabeculation.

9.1.2 LV Thrombus

LV thrombus appears as a filling defect in the LV chamber. Left ventricular thrombi occur in regions of ventricular dyskinesia or aneurysm formation, both of which usually result from prior myocardial infarction. Thrombi have homogeneous attenuation on CT. The CT attenuation value is less than that of the myocardial wall and of normal ventricular trabeculations (Fig. 9.3). In addition, thrombi do not significantly enhance after contrast administration, whereas the LV wall shows modest contrast enhancement.

9.1.3 Endocardial Fibroelastosis

Endocardial fibroelastosis refers to a pronounced, diffuse thickening of the ventricular endocardium and presents as unexplained heart failure in infants and children less than or equal to 2 years of age. It is not a disease of adults.

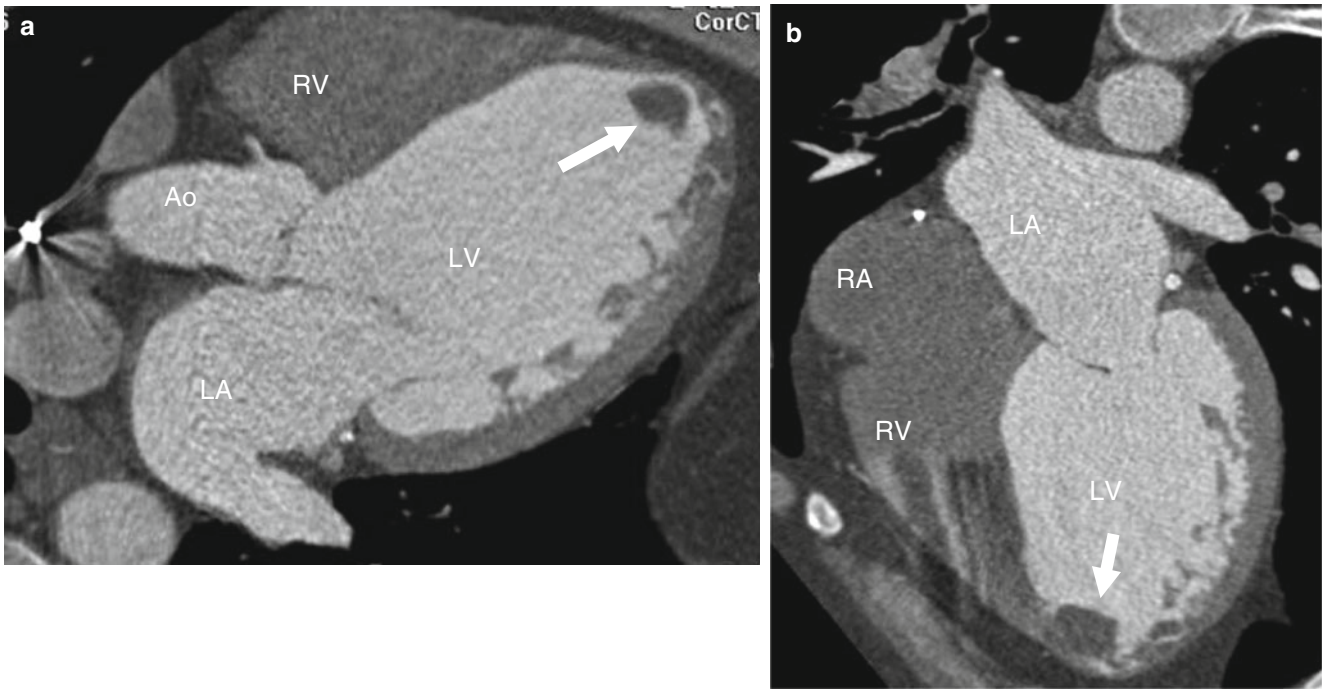

Fig. 9.3 Left ventricular thrombus in a 50-year-old woman who suffered a recent myocardial infarction. Panels (**a**) and (**b**) are two long-axis contrast-enhanced CT scans showing a homogeneous soft tissue mass (*arrow*) in the apex of a dilated left ventricle (*LV*). The thrombus does not enhance and the attenuation of the thrombus is less than that of the left ventricular trabeculations and myocardial wall distinguishing it from normal myocardial tissue. *LA* lft atrium, *RA* right atrium, *RV* Right ventricle, *Ao* Aorta

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