Left ventricular (LV) noncompaction is a rare cardiomyopathy caused by an arrest of normal endomyocardial morphogenesis. Morphologically, it is characterized by prominent trabeculation with deep intertrabecular recesses and a thin compacted myocardial layer comprising less than 50% of the LV wall thickness. It is an uncommon finding, which may remain undiagnosed for years unless it presents as heart failure or severe arrhythmia, systemic embolic event, or sudden cardiac death. Echocardiography is usually the first-choice diagnostic approach. Apart from a typical 2-layered myocardial appearance, nonspecific echocardiographic findings include reduced global LV systolic function, LV diastolic dysfunction, and abnormal papillary muscle structure. In extremely rare cases, noncompaction can be accompanied by other congenital malformations such as ventricular septal defect, bicuspid aortic valve, or Epstein anomaly.

A 44-year-old man with newly diagnosed heart failure was referred to our cardiac outpatient clinic for an echocardiographic examination, which revealed LV enlargement (72/63 mm), global hypokinesis, and low LV ejection fraction (30%) along with prominent trabeculation and deep intertrabecular recesses with a thin compacted layer of the myocardium. In addition, a bicuspid aortic valve was reported (FIGURE 1A). No abnormalities were observed on coronary angiography. LV noncompaction was diagnosed and the patient received standard treatment including angiotensin-converting enzyme inhibitors, β-blockers, and diuretics. Following an episode of ventricular fibrillation, the patient received an implantable cardioverter-defibrillator (ICD). Cardiac magnetic resonance was scheduled to confirm LV noncompaction and to identify myocardial abnormalities.

Unfortunately, 4 days after ICD implantation, the patient died because of acute pulmonary edema and pulseless electrical activity. Marked global
dilation and hypertrophy of the heart (cor bovinum) with LV predominance were found on autopsy. A detailed study revealed a sponge-like appearance of the LV wall with multiple deep intertrabecular recesses communicating with the ventricular lumen (FIGURE 1B). A microscopic examination of the heart muscle confirmed the abnormalities observed on gross pathology and revealed massive myocardial hypertrophy and features of early ischemic injury. The morphological diagnosis was primary cardiomyopathy—LV noncompaction—which results from impaired prenatal development of the ventricle (FIGURE 1C).

LV noncompaction is a distinct congenital malformation, which develops early during embryogenesis. It may have a crucial effect on prognosis in young age because of major cardiovascular complications such as severe and rapidly progressing systolic dysfunction and death, as observed in our case.

REFERENCES