A 64-year-old woman was admitted to hospital because of acute exacerbation of congestive heart failure. She had been treated for hypertension and cardiomyopathy of unknown cause over an extended period. She had no history of myocardial infarction and no familial history of cardiovascular diseases or sudden cardiac death. Echocardiography revealed severely reduced left ventricular (LV) contraction, severe mitral regurgitation, thickened myocardium with prominent trabeculations, and deep intertrabecular recesses in the LV walls. Cardiac cine-magnetic resonance imaging (MRI) demonstrated a severely dilated LV with an end-diastolic volume of 463 ml, and severely depressed LV contractility with an ejection fraction of 12%.

Multiple prominent ventricular trabeculations were observed in the LV wall. The ratio of the noncompacted to compacted myocardium was >2.3 in diastole (Figure 1). These findings defined a rare case of isolated noncompaction of the LV myocardium (INVM) without any associated cardiac anomaly.1,2 Interestingly, late gadolinium enhancement (LGE), suggesting myocardial fibrosis on LGE-MRI, was observed not only in the endocardial side of the compacted myocardium, but also in the mid-layer of the compacted myocardium. Although there are a few previous case reports of subendocardial and trabecular myocardial fibrosis, the finding of “mid-layer fibrosis” on LGE-MRI in a patient with INVM has not been reported.3,4 Furthermore, this is the first case of

Figure 1. Cine images showing unusual muscle trabeculations extending into the left ventricular cavity in the vertical long-axis (left) and short-axis (right) views.
combined subendocardial myocardial fibrosis and mid-layer fibrosis on LGE-MRI in a patient with INVM. Previous INVM autopsy case reports suggest that either a mismatch in blood supply and demand from the numerous prominent trabeculations or coronary microcirculatory dysfunction is the mechanism of the progressive ventricular failure and myocardial fibrosis in the endocardial side of the compacted myocardium. In addition, the mechanism for the mid-layer fibrosis of the compacted myocardium is thought to be a combination of factors including genetic predisposition, exposure to toxins and pathogens, microvascular ischemia, and abnormal modulation of immune and metabolic responses, such as overactivity of the renin–angiotensin–aldosterone system\(^5\)\(^6\) (Figure 2).

In conclusion, the combination of subendocardial myocardial fibrosis and mid-layer fibrosis by LGE-MRI in a patient with INVM is a very rare finding and this is the first case report of such a specific fibrosis.

**Disclosure**

There is no potential conflict of interest.

**References**