Unsolved Issue in Left Ventricular Noncompaction: Is the Strange Form of Myocardium Congenital or Acquired?

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Left ventricular noncompaction (LVNC) is morphologically characterized by a thinning of the compacted layer, excess reticulated trabeculae, and deep gaps in the noncompacted layer [1]. The noncompacted layer is also found in the early normal fetal stage, and the rough myocardium is supplied with blood flow via the gaps and sinusoids in communication with the ventricular cavity. In the developmental process, the myocardium gradually acquires a compacted structure, and eventually the blood supply from the coronary artery becomes its main supply. LVNC is considered to be caused by a failure of the sponge-like embryonic myocardium due to the impaired process of compaction in the fetal myocardium [2]. In other words, abnormal myocardium already exists from birth. However, in recent years, many asymptomatic cases have been reported with the development of imaging modalities, in which LVNC-like findings are identified incidentally in a medical checkup [3]. According to a survey in Japan, around 40% of the patients with LVNC are suspected to be affected by genetic factors, and genetic diversity such as autosomal inheritance and mitochondrial gene mutation in addition to X-linkage has been pointed out [4]. Furthermore, some reports on adult cases have confirmed over time the sudden expression and improvement of this strange structure. Thus, many re-

searchers are interested in whether LVNC is a congenital cardiomyopathy or some acquired syndrome.

Loria et al. [5] report an adult case of LVNC identified unexpectedly, along with a literature review. In their report, a 57-year-old female with a history of anthracycline treatment for breast cancer showed prominent myocardial trabeculations. Despite having no family history of cardiomyopathy and not having been noted for LVNC-like findings on echocardiography in the past, LVNC-like structural changes and deep depression with perfusion of the interventricular space were suddenly and incidentally observed with color Doppler. Furthermore, these findings were also confirmed by cardiac magnetic resonance imaging (MRI). It seems as if the LVNC-like findings suddenly developed.

We have two major points to consider when discussing LVNC etiology. One aspect is the kind of diagnostic modality. Echocardiographic diagnosis classically reported may not be suitable for observing apical abnormalities commonly found in LVNCs of adult patients. The prevalence of LVNC based on echocardiography is approximately 0.13% [6]. On the other hand, in cardiac MRI, which is a more accurate diagnostic modality, LVNC was detected at a frequency of 6% in a large population [7]. The other aspect is genetic abnormalities. The abnormal-

ities in adult LVNC have been reported to differ from those in children. In adult LVNC, we can often detect the sarcomere gene mutations reported in dilated or hypertrophic cardiomyopathy. However, genetic abnormalities such as those in *TAZ* or *LDB3*, which are found in pediatric LVNC, are not common [8]. This difference in genetic abnormalities indicates that adult LVNCs are rather similar to sarcomeric abnormalities, such as dilated or hypertrophic cardiomyopathy, and may help us understand the difference in the onset mechanism of LVNC-like appearance in adults and children. Presumably, congenital cardiomyopathy is unlikely to co-exist for a long time because the compacted myocardium in LVNC is approximately 30% thinner than the normal myocardium. Moreover, additional genetic mutations or epigenetic mechanisms are expected to result in LVNC-like changes.

Reports on pregnant women and athletes have alluded the claim that the LVNC-like appearance may reverse, and the strange structure disappears when the etiology is removed [9]. In addition, Stöllberger et al. [10] report a case in which the trabecular structure became less prom-

inent after cardiac resynchronization therapy. This phenomenon may occur because the LVNC-like appearance becomes more visible on echocardiography in the presence of acquired and transient LV dilation. Alternatively, the trabeculae may have emerged as a compensation to increase endocardial surface area to optimize the stroke volume in failing hearts [11], and when cardiac contraction improves by cardiac resynchronization therapy, the trabecular structure may become inconspicuous and no longer appear [10]. On the other hand, cardiac MRI with high precision may clearly identify LVNC structural features. In cases with poor echocardiographic imaging, more accurate imaging should be done using high-precision modalities such as MRI to avoid missing LVNC-like appearance [12].

Disclosure Statement

The authors have no conflicts of interest to disclose.

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