Our echocardiography laboratory was consulted to determine whether a patient’s echocardiogram would fulfill the criteria of left ventricular noncompaction cardiomyopathy (LVNC). Images of the left ventricle showed a 2-layer structure with a compacted, thin epicardial band and a much thicker noncompacted endocardial layer of trabecular meshwork with deep endomyocardial spaces. In addition, there was color Doppler evidence of deep perfusion of these intertrabecular recesses. The predominant localization of the pathology was lateral and apical, with a ratio of end-systolic noncompacted to compacted layers of 2.4. Thus, from an imaging point of view, everything seemed to fit the diagnosis of LVNC. However, comprehensive workup with medical information from the past revealed a different cause of cardiomyopathy.

In 2010, the patient had already been referred to our hospital because of dyspnea. A transthoracic ECG revealed a markedly reduced ejection fraction of only 7%, with a dilated left ventricle. In addition, it showed prominent apical and anterior trabeculation, but none of the Jenni criteria1,2 for an LVNC were fulfilled (Figure 1 and Movie I in the online-only Data Supplement). ECG showed a left ventricular branch block with ST elevations in V1 through V5 (Figure 2). A coronary angiogram showed no coronary stenoses, but ascertained the markedly reduced ejection fraction. Cardiac magnetic resonance tomography confirmed the echocardiographic findings (Figure 3) and revealed a late enhancement pattern as usually seen in myocarditis, with late enhancement speckled and distributed over the left ventricle (Figure 4A and 4B). Last, left ventricular biopsies proved acute myocarditis (Figures 5 and 6). A subsequent ECG showed ST elevations only in V3 and V4, and they were not very prominent (Figure 7).

In the recent (latest) echocardiogram, the ejection fraction had improved up to about 20%, but the echo pattern showed the previously described morphology with increased trabeculation, noncompaction-like layers, and intertrabecular recesses (Figure 8 and Movie II in the online-only Data Supplement).

This case highlights several problems related to the diagnosis of LVNC. First, there is no clear genetic or laboratory test for the diagnosis of LVNC. Second, over time, some well-defined cardiac diseases like myocarditis can remodel toward an echocardiographic pattern compatible with LVNC, as documented in our patient. Last, medical information from the past is of key importance; the documented myocarditis in the history excludes the diagnosis of LVNC, according to Jenni’s1,2 criterion number 1 (ie, absence of other cardiac abnormalities). Thus, the pure documentation of the typical echocardiographic criteria is insufficient to establish the diagnosis of LVNC. In summary, these factors facilitate overdiagnosis of LVNC, which is, in fact, a very rare disease.

Disclosures

None.

References

Figure 2. ECG at first presentation in 2010. Left bundle-branch block with subsequent ST elevations in V1 through V5 is seen, which fits a picture of myocarditis.

Figure 3. MRI 4-chamber view of the patient’s heart at baseline.

Figure 4. Cardiac magnetic resonance images taken at baseline to demonstrate late enhancement, which was speckled and distributed over the left ventricle. (A) Late enhancement, anterior medial. (B) Late enhancement, inferior basal.
Figure 5. Masson-Trichrome routine staining with small, speckled interstitial fibrosis and infiltration of mononuclear cells and inflammatory cells. Courtesy of Reinhard Kandolf, Institute of Pathology, University Tübingen, Germany.

Figure 6. Immunohistochemistry: major histocompatibility class II expression in activated macrophages, natural killer cells, and endothelium. Relatively fresh myocarditis with necrobiosis of myocytes is seen. Courtesy of Reinhard Kandolf, Institute of Pathology, University Tübingen, Germany.
Figure 7. ECG at follow-up. ST elevations are seen only in V3 and V4, and they are not very prominent.

Figure 8. Echocardiographic 4-chamber view of the patient’s heart at follow-up (taken from loop 2). Please note the noncompaction of the lateral wall.
Left Ventricular Noncompaction Cardiomyopathy: An Overdiagnosed Disease
Markus Niemann, Stefan Störk and Frank Weidemann

Circulation. 2012;126:e240-e243
doi: 10.1161/CIRCULATIONAHA.112.095059
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 2012 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/126/16/e240

Data Supplement (unedited) at:
http://circ.ahajournals.org/content/suppl/2012/10/12/126.16.e240.DC1

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Circulation can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to Circulation is online at:
http://circ.ahajournals.org//subscriptions/