Multiple coronary fistulae that drain into the left ventricle are rarely associated with apical hypertrophic cardiomyopathy. On the other hand, associations of left ventricular noncompaction (LVNC) and multiple coronary fistulae or hypertrophic cardiomyopathy have previously been reported in several cases. We report here a unique case of apical hypertrophy of the left ventricle, multiple coronary–left ventricular fistulae, and a morphological structure of the left ventricular myocardium mimicking LVNC.

Case Presentation
A 59-year-old man was admitted to our hospital for fever and rapidly progressing dyspnea that had persisted for 10 days. He had been diagnosed with apical hypertrophic cardiomyopathy in his 30s, although he had no regular medical checkups and was asymptomatic. On admission, the patient had mild wheezes audible in the midportion of the right lung and a diastolic murmur heard at the Erb and the cardiac apical area.

Chest x-ray and computed tomography revealed cardiac enlargement with increased pulmonary vasculature, granular shadows in the lower areas of both lungs, and increased density of the right middle lung area. These findings suggested failure of the left side of the heart with pneumonia. A 12-lead electrocardiogram (ECG) confirmed normal sinus rhythm with inverted T waves in leads II, III, aVF, and V3 through V6 (Figure 1). He recovered rapidly with antibiotics and standard therapy for heart failure.

Transthoracic 2-dimensional echocardiography showed diffuse hypertrophy of the mid to apical left ventricular walls (Figure 2A and Movie I in the online-only Data Supplement). Doppler echocardiography revealed multiple areas of mosaic color flow within the anterior, lateral, and apical myocardium connected to the left ventricular cavity, with a peak flow speed of 0.8 to 0.9 m/s in the diastolic phase (Figure 2B and Movie II in the online-only Data Supplement). Enlarged left and right coronary arteries and rapid diastolic flow with a peak speed of 1.5 m/s in the left anterior descending artery were also identified.

A markedly enlarged, tortuous coronary artery with multiple coronary artery–left ventricular fistulae was visible on cardiac computed tomography examination (Figure 3A). Coronary computed tomography angiography, diastolic-phase imaging, showed a 2-layer left ventricular myocardium mimicking LVNC, with a maximal ratio of noncompacted to compacted myocardium of 2.2 (Figure 3B and 3C).

Coronary angiography confirmed multiple fistulae between the left and right coronary arteries and the left ventricle (Figure 4A and 4B). Left ventricular angiography revealed a spade-shaped cavity indicative of apical hypertrophic cardiomyopathy.

Cine magnetic resonance images showed small fine trabeculae extending from the lower base to the apical anterior, lateral, and inferior endocardium in the diastolic phase. In the systolic phase, thickening of both layers of the myocardium (mimicking compacted and noncompacted myocardium) was present (Movies III and IV in the online-only Data Supplement).

Discussion
Coronary artery fistula has been reported in ≈0.2% of patients undergoing coronary angiography, and other cardiac abnormalities are associated in 20% of those cases. Coexistence of multiple coronary artery–left ventricular fistulae and apical hypertrophic cardiomyopathy is very rare, and most drain from the right and left coronary arteries to the left ventricle. This phenotype is considered to represent the partial persistent embryonic myocardial sinusoids that arise from the endothelial protrusion into the intertrabecular spaces.
The association of multiple coronary artery–left ventricular fistulae and LVNC was reported by Dias et al., with persistent myocardial sinusoids and compaction of the myocardium present in the same patient.

LVNC has gained increasing recognition in the last 30 years, and there has been some discussion of its terminology and definition. Our case fulfilled the morphological definition of LVNC, although there may be some suspicion that this structure really represents an arrest of the myocardial compaction process during the first trimester. Oechslin and Jenni described that multiple persistent sinusoids are generally present with pulmonary artery atresia and that these patients should not be included the category of LVNC. Unlike the Oechslin and Jenni report, the patient described here did not have pulmonary atresia. Coexistence of hypertrophic cardiomyopathy and LVNC in one family with overlapping phenotypes has been reported. A comprehensive diagnostic assessment, including multimodality imaging, systematic screening, and genetic assessment, may help elucidate the true cause of cases with multiple anomalies.

To the best of our knowledge, this is the first report describing a patient with apical hypertrophic cardiomyopathy, multiple coronary artery–left ventricular fistulas, and a morphological structure mimicking LVNC.

Acknowledgments

We thank Dr Naoto Fukuda for the computed tomography image acquisition and Professor Fukiko Ichida, Dr Takaaki Matsuyama, and Professor Katsu Takenaka for their kind academic suggestions.

Disclosures

None.

References


Figure 1. The 12-lead ECG identified a normal sinus rhythm with inverted T waves in leads II, III, aVF, and V5 through V6.

Figure 2. A and B. Apical short-axis view of 2-dimensional echocardiography and color doppler imaging at the same level. Multiple coronary fistulas (white arrow) that drained through thickened myocardium into the left ventricular cavity were visible as color doppler flow signals in the diastolic phase.
Figure 3. A, Computed tomography coronary angiography, diastolic phase, showed an enlarged and tortuous left coronary artery. B and C, Multiple coronary artery–left ventricular fistulas (white arrow) and hypertrabeculation of the endocardial side of the myocardium mimicking noncompaction of the left ventricle (black arrow) were clearly shown in diastolic synchronized contrast computed tomography imaging. Maximal ratio of noncompacted to compacted myocardium was 2.2 in the lower mid short-axis view.

Figure 4. A, The right coronary angiogram showed multiple fistulas (arrow) draining into the left ventricular cavity. B, The left coronary angiogram showed an enlarged and tortuous left coronary artery. Multiple fistulas (arrow) draining into the left ventricular cavity were also identified.
A Case With Apical Hypertrophic Cardiomyopathy, Multiple Coronary Artery–Left Ventricular Fistulae, and a Morphological Structure Mimicking Left Ventricular Noncompaction: Statue of Cerberus or Double-Headed Eagle?
Sakura Nagumo, Mio Ebato, Masaaki Kurata, Kohei Wakabayashi, Hisa Shimojima, Tokutada Sato, Yoshiro Hori and Hiroshi Suzuki

_Circulation_. 2015;131:2161-2163
doi: 10.1161/CIRCULATIONAHA.114.015008
_Circulation_ is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 2015 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/131/24/2161

Data Supplement (unedited) at:
http://circ.ahajournals.org/content/suppl/2015/06/18/CIRCULATIONAHA.114.015008.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/