Apical hypertrophic cardiomyopathy associated with multiple coronary artery–left ventricular fistulae: a report of a case and review of the literature

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We present a rare case of multiple coronary artery–left ventricular (LV) fistulae, associated with apical hypertrophic cardiomyopathy in an 83-year-old woman with electrocardiographic abnormalities and a history of arterial hypertension and paroxysmal atrial fibrillation. In order to evaluate the clinical significance and obtain further insights into this unusual disease, the patient has undergone coronary angiography, left ventriculography, and magnetic resonance imaging which better substantiated the structural abnormalities of the LV and the coronary network.

Keywords
Coronary artery–left ventricular fistulae • Coronary steal • Apical hypertrophic cardiomyopathy

Case presentation

We report the case of an 83-year-old woman admitted to our outpatient clinic for routine evaluation due to electrocardiographic (ECG) abnormalities. She had a history of arterial hypertension and paroxysmal atrial fibrillation treated with nebivolol. She also reported a recent history of abrupt episode of dysarthria lasting 2 h associated with symptoms of numbness of her inferior extremities. Her family history was unremarkable from the cardiovascular point of view. On physical examination, the patient appeared generally well. Her blood pressure was 120/80 mmHg and her pulse was regular; 65 bpm. Electrocardiography revealed sinus rhythm, symmetrical, deep T-wave inversions in leads I, AVL, and V2–V6, and biphasic T-waves in leads II, III, and AVF. Chest X-ray was normal. Biochemical blood parameters were also within the normal range. Due to her recent neurological symptoms, she underwent brain computed tomography scan without, however, evidence of cerebral stroke.

The patient was further subjected to two-dimensional transthoracic echocardiography that revealed apical hypertrophy complicated with a small-sized apical aneurism. Transthoracic colour Doppler echocardiography, using a high-frequency transducer and a low Nyquist limit revealed multiple linear colour-flow signals perpendicular to the epicardium draining in the left ventricular (LV) chamber. Close inspection of the colour signal in association with pulse-wave Doppler revealed a diastolic flow pattern arising from the left anterior descending (LAD) coronary artery, suggestive of multiple coronary artery–LV microfistulae (Figure 1; see Supplementary data online, Video 1). Multiple coronary artery–LV microfistulae were better visualized utilizing a second-generation contrast agent (SonoVue®) (Figure 2; see Supplementary data online, Video 2). Continuous wave Doppler was finally used to quantify the inter-ventricular pressure gradient which turned out insignificant, from the haemodynamic point of view.

After the completion of the non-invasive diagnostic approach, the patient underwent cardiac catheterization in order to exclude concomitant coronary artery disease and the presence of other co-existing congenital coronary anomalies. Coronary angiography showed no significant lesions of epicardial coronary arteries. However, left coronary angiography reveals multiple microfistulae originating from the LAD artery and emptying in LV cavity at the level of the apex. Left ventriculography revealed a spade-like LV—typical of apical hypertrophic cardiomyopathy (HCM)—with LV obstruction during systole and aneurismal-like...
bulging in the apical segment (Figure 3). In accordance to echocardiographic findings, a haemodynamically insignificant pressure gradient between the basal and apical region of LV was demonstrated.

In order to obtain further insights about this rare condition, a cardiac magnetic resonance imaging was performed. The latter technique substantiated the diagnosis of apical HCM (Figure 4), excluded the diagnosis of apical myocardial non-compaction but did not reveal coronary microfistulae (Figure 4).

The patient was discharged 4 days after the completion of the clinical investigation under medical therapy including nebivolol and vitamin K antagonists.

**Discussion**

Fistulae between coronary arteries and cardiac chambers are rare. They are reported in ~0.2% of the patients undergoing cardiac catheterization.\(^1\) They usually arise from the right coronary artery and drain into the right heart chambers (right atrium, right ventricular, and pulmonary artery). In 20% of the cases, coronary artery fistulae are associated with other cardiac anomalies including aortic and pulmonary atresia and patent ductus arteriosus.\(^1\)

Fistulae draining into the LV are uncommon and their co-existence with apical HCM has been rarely reported. Morphological studies provide a possible explanation for the development of coronary artery–cardiac chamber fistulae. They might represent a partial persistence of embryonic myocardial sinusoids that arise from endothelial protrusions into the intertrabecular spaces.\(^2,3\) Foetal regression of this structure results in the formation of the Thebesian vessels of the adult heart. Thus, interference with developmental changes might produce an abnormally prominent Thebesian system with morphological appearance of multiple coronary microfistulae.\(^2,3\)

Reviewing the literature, we identified six more reports of coronary artery–heart chamber fistulae associated with HCM.\(^2,7\) All the available clinical and anatomical characteristics of the reported cases are summarized in Table 1. In 9 of the 10 patients, HCM was
associated with coronary–LV fistulae. Angina was the most common presenting symptom. Notably, the majority of the cases were reported in South Europe (Mediterranean territory). All authors concluded that aetiology is obscure. Hypertrophic cardiomyopathy and coronary artery–cardiac chamber fistulae could simply co-exist or there could be a causative relationship between the two conditions. Even if a causative relationship exists, the pathophysiological background remains unclear. The apical hypertrophy could be the result of chronic LV volume overload through the coronary artery–LV shunt or could be the cause of multiple coronary microfistulae, possibly due to disarray of myocardial cells.

The clinical diagnosis of coronary artery–LV fistulae is difficult because clinical presentation and ECG manifestations are non-specific. Most patients with coronary artery fistulae and HCM present with typical angina pectoris without angiographic evidence of co-existing coronary artery disease. Nevertheless, HCM can also cause ischaemia by altering the oxygen demand/supply balance to the hypertrophic myocardium. Since both coronary artery fistulae and HCM can produce myocardial ischaemia and angina, it is also plausible that the co-existence of the two conditions could aggravate symptoms. In the presented case, multiple coronary artery to LV microfistulae were identified by transthoracic colour Doppler echocardiography using high-frequency transducer and a low Nyquist limit. Multiple linear colour flow signals perpendicular to the epicardial surface were visualized. Pulse-wave Doppler further identified a diastolic flow pattern towards the LV cavity. The presence of an extensive microfistulae network interconnecting the left coronary artery with the apical part of LV was finally substantiated by coronary angiography. Angiography also excluded concomitant coronary artery disease and the presence of other co-existing congenital coronary anomalies. Ventriculography further demonstrated the spade-like LV morphology which is representative of apical HCM.

Contrast echocardiography also provided impressive images revealing flow within the apical myocardium. Close inspection of the flow pattern could identify the diastolic flow towards the left cavity; however, contrast echocardiography did not provide any additional information to colour Doppler images and created the initial false impression of apical non-compaction myocardium.

Finally magnetic resonance imaging revealed the apical hypertrophy, excluded the diagnosis of apical myocardial non-compaction but provided no evidence of the coronary microfistulae.

There is no experience in the management of patients with HCM related to coronary–LV fistulae. Treatment of isolated HCM and coronary fistulae is essentially medical. Thus, conservative management with continued follow-up seems appropriate when these conditions co-exist. Our patient was treated with nebivolol and a vitamin K antagonist and was discharged after the investigation was completed. She remained asymptomatic and no adverse events were recorded in the next 5-month follow-up.

In conclusion, we reported a rare case of left coronary–LV microfistulae associated with apical HCM. We applied a number of imaging techniques to substantiate the presence of this rare condition; however, it seems that colour Doppler imaging in the hands of an experienced echocardiographer is enough to set the diagnosis.

**Table 1** Clinical and epidemiological characteristics of patients with apical hypertrophic cardiomyopathy and coronary–heart chamber microfistulae

<table>
<thead>
<tr>
<th>Gender</th>
<th>Age</th>
<th>Geographical origin</th>
<th>Anatomy of fistulae</th>
<th>Clinical presentation</th>
<th>Diagnosis</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>This study</td>
<td>Female</td>
<td>82</td>
<td>Greece</td>
<td>LCA–LV</td>
<td>ECG abnormalities</td>
<td>ECHO, CA, MRI</td>
</tr>
<tr>
<td>Alyan et al.</td>
<td>Male</td>
<td>63</td>
<td>Turkey</td>
<td>LCA–LV</td>
<td>Exertion angina</td>
<td>ECHO, CA</td>
</tr>
<tr>
<td>Hong et al.</td>
<td>Female</td>
<td>67</td>
<td>Korea</td>
<td>LCA–LV</td>
<td>Dyspnoea</td>
<td>ECHO, CA</td>
</tr>
<tr>
<td>Caputo et al.</td>
<td>Male</td>
<td>6</td>
<td>Italy</td>
<td>RCA–RV</td>
<td>Cardiac arrest</td>
<td>ECHO, CA</td>
</tr>
<tr>
<td>Monmeneu et al.</td>
<td>Male</td>
<td>NA</td>
<td>Spain</td>
<td>LCA–LV</td>
<td>Exertion angina</td>
<td>ECHO, CA</td>
</tr>
<tr>
<td>Delarche et al.</td>
<td>Male</td>
<td>NA</td>
<td>France</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>Female</td>
<td>Female</td>
<td>53</td>
<td>Japan</td>
<td>RCA–LV</td>
<td>Angina</td>
<td>ECHO, CA</td>
</tr>
<tr>
<td>Kiyokawa et al.</td>
<td>Female</td>
<td>46</td>
<td>Japan</td>
<td>LCA–LV</td>
<td>ECG abnormalities</td>
<td>ECHO, CA</td>
</tr>
</tbody>
</table>

LCA, left coronary artery; RCA, right coronary artery; LV, left ventricular; RV, right ventricular; TIA, transient ischaemic attack; PA, pulmonary artery; ECHO, echocardiography; CA, coronary angiography; MRI, magnetic resonance imaging; ECG, electrocardiogram; ICD, implantable cardiac defibrillator.

**Supplementary data**

Supplementary data are available at European Journal of Echocardiography online.
References


