A young patient with left ventricular hypertrophy and accidentally discovered aortic dissection: hypertensive heart disease or hypertrophic cardiomyopathy?

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A 36-year-old man with a family history of hypertension and sudden cardiac death, who was admitted a month earlier to a local hospital due to acute cardiogenic pulmonary edema and newly diagnosed hypertension, was referred to our institution with suspicion of hypertrophic cardiomyopathy (HCM).

On admission, the patient was asymptomatic and did not complain of fatigue, chest pain, or syncope. On physical examination, his blood pressure (BP) was significantly elevated (180/100 mm Hg). The lungs were clear on auscultation. Pulse in the radial and femoral arteries was preserved. Neither edema of lower extremities nor heart murmur were detected. Abdominal auscultation revealed vascular murmur in the umbilical region.

The baseline level of N-terminal pro–B-type natriuretic peptide was 811.4 pg/ml (normal range, 0–125 pg/ml), high-sensitivity troponin T, 20.2 ng/l (normal range, 0–14 ng/l), and creatinine, 1.4 mg/dl (normal range, 0.7–1.2 mg/dl). The standard 12-lead electrocardiogram showed sinus rhythm, left atrial (LA) enlargement, and left ventricular (LV) hypertrophy (FIGURE 1A). No significant pathology was present on chest X-ray.

Transthoracic echocardiography revealed concentric LV hypertrophy (maximally 19 mm at the interventricular septum) with preserved LV ejection fraction (70%), moderately decreased global longitudinal strain (–13.7%), and nonsignificant LV outflow tract gradient (10 mm Hg at rest and 14 mm Hg after the Valsalva test). There was moderate LA enlargement (LA volume index, 37.2 ml/m^2), impaired LV relaxation (E/A, 0.88; E/e', 6.6; E-wave deceleration time, 297 ms), and a normal ascending aorta diameter (FIGURE 1B).

Cardiovascular magnetic resonance imaging confirmed concentric LV hypertrophy (maximally 18 mm) and increased myocardial mass (LV mass index, 124 ml/m^2; normal range, 59–92 ml/m^2) (FIGURE 1C).

Due to vascular murmur in the abdomen, ultrasound imaging was performed and revealed abdominal aortic dissection (FIGURE 1D). A computed tomography scan confirmed aortic dissection originating below renal arteries and involving common iliac arteries (Stanford B; FIGURE 1E and 1F). The presence of thrombi formed at the site of the aortic dissection suggested a chronic presentation.

The patient was managed conservatively with strict BP control, intensified antihypertensive treatment (metoprolol, telmisartan, torasemide, spironolactone, amlodipine, clonidine), and close follow-up.[1] Secondary causes of hypertension, including renal artery stenosis, abnormal kidney and adrenal glands, hyperaldosteronism, hyper- and hypothyroidism, were excluded.

Genetic analysis did not reveal any potentially disease-causing mutation. Next-generation sequencing was conducted with the TruSight

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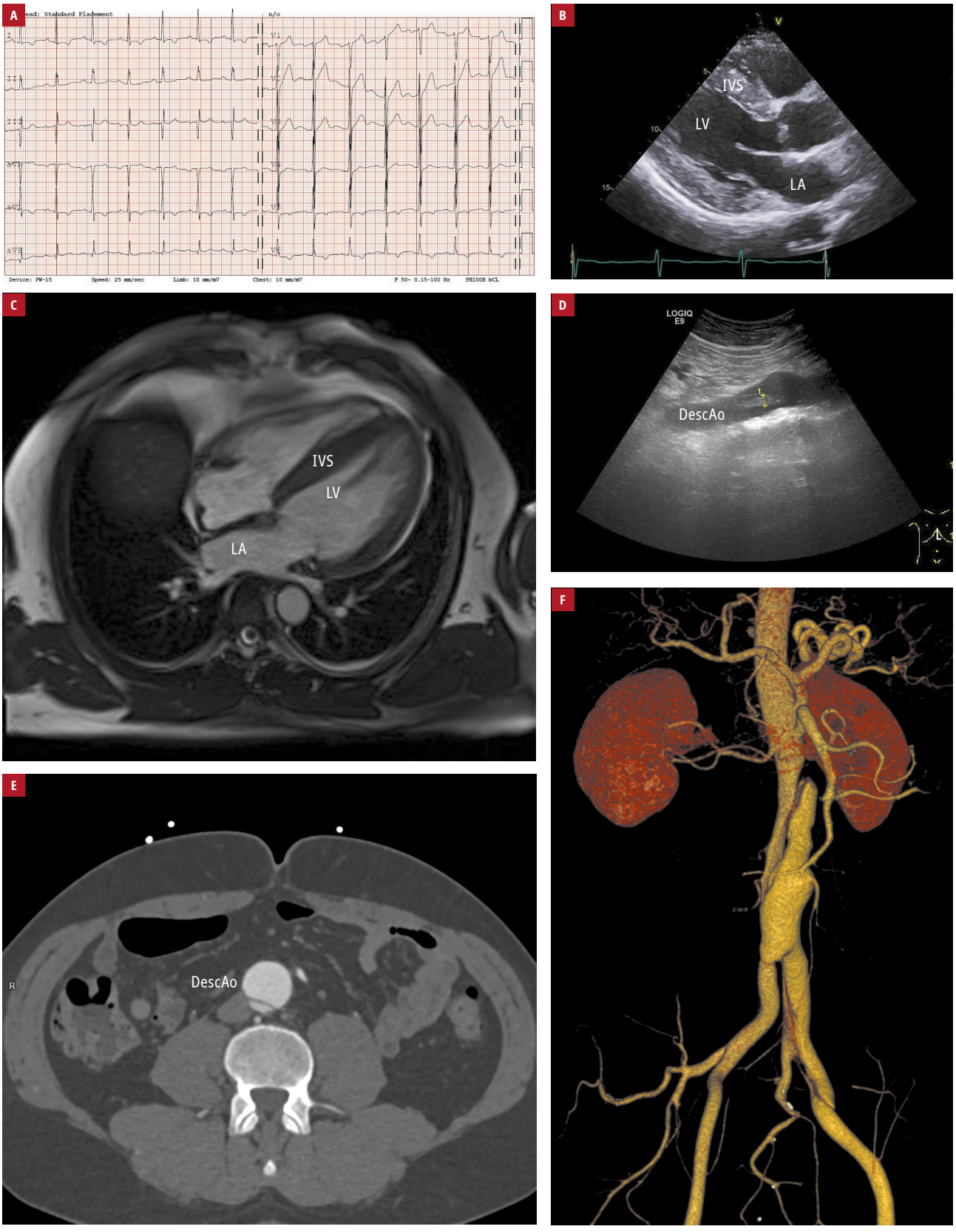

FIGURE 1 **A** – a standard 12-lead electrocardiogram with sinus rhythm, left atrial enlargement, and left ventricular hypertrophy; **B** – transthoracic echocardiography (parasternal long-axis view) showing left ventricular hypertrophy; **C** – cardiovascular magnetic resonance scan: (cine 4-chamber view) showing left ventricular hypertrophy; **D** – ultrasound imaging showing abdominal aortic dissection and thrombi within the aortic lumen; **E** – a computed tomography scan showing dissection of the descending aorta; **F** – computed tomography showing 3-dimensional reconstruction of the descending aorta
Abbreviations: DescAo, descending aorta; IVS, interventricular septum; LA, left atrium; LV, left ventricle

Cardio Sequencing Panel (Illumina, San Diego, California, United States) providing a comprehensive coverage of 174 genes related to inherited cardiac conditions (ie, cardiomyopathies, aortopathies, and arrhythmias), with a special emphasis on genes associated with HCM (*MYH7*, *MYBPC3*, *TNNT2*, *TNNI3*, *MYL3*, and *MYL2*).[2]

In summary, our case underlines the importance of accurate physical examination, which guides further diagnostic workup. Aortic dissection typically presents with tearing chest pain and severe hemodynamic compromise. Painless dissection, as in this case, occurs relatively rarely. Our patient had one symptom—vascular murmur in the umbilical region—and at least one risk factor for aortic dissection—history of elevated BP of unknown duration, treated only for a month. However, vascular murmur in the umbilical region in a patient with hypertension and preserved pulse in the femoral arteries might be more probably a sign of renal artery stenosis.

Secondly, the differential diagnosis between hypertensive heart disease and HCM is crucial because it directly affects treatment and both diseases carry different risk of adverse cardiovascular events. Due to the fact that in this particular case no mutations in the genes associated with cardiomyopathies or aortopathies were found, and the patient suffered from hypertension with such complications as pulmonary edema, aortic dissection, and chronic kidney disease stage 2, our final diagnosis was hypertensive heart disease.

ARTICLE INFORMATION

CONFLICT OF INTEREST None declared.

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