Many disorders of one heart - accidentally discovered aortic dissection, bicuspid aortic valve and hypertrophic cardiomyopathy in young patient with hypertension

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A 29-year-old male with history of untreated hypertension, hospitalized for few days in local hospital due to pneumonia, was referred to our institution for further evaluation with suspicion of myocarditis and hypertrophic cardiomyopathy. On admission patient suffered from dyspnea and cough. He denied syncope or any episode of chest pain. On physical examination the patient was normotensive, with normal body temperature. Auscultation revealed bilateral rales. The liver was not enlarged, jugular veins were normal, there was no oedema of lower extremities. The baseline level of CRP was 4.4 (range 0 - 0.5) mg/dl, high-sensitivity cardiac troponin T was 324 (range 0-14) ng/L, and NT-proBNP was 3285 (range 0-125) pg/mL. The standard 12-lead electrocardiogram demonstrated sinus rhythm, left atrial enlargement and left ventricular (LV) hypertrophy with non-specific ST segment and T-wave abnormalities (Fig. 1A). The chest X-ray showed bilateral lung consolidation and pulmonary congestion (Fig. 1B). Transthoracic echocardiography (TTE) revealed significant asymmetric LV hypertrophy with preserved LV ejection fraction. Moreover bicuspid aortic valve (BAV) with moderate regurgitation and mild dilatation of the ascending aorta (41 mm) were found. Due to suspicion of myocarditis, cardiac magnetic resonance (CMR) imaging was performed. CMR confirmed hypertrophic cardiomyopathy (HCM) with the maximal LV wall thickness of 32 mm and increased myocardial mass (LV mass index 149 ml/m², range 68–103). Systolic function of LV and right ventricle (RV) were mildly decreased (LVEF 54%, range 57-74; RVEF 45%, range 48-74%). Moreover ascending aortic dissection was detected (Fig. 1C, 1D). Patient was transferred to the computed tomography (CT) unit to confirm the diagnosis. Aortic dissection originated in aortic root and involving descending aorta was detected (DeBakey Type I). The maximal diameter of the ascending aorta was 44 mm with no evidence of coronary arteries involvement (Fig. 1E, 1F). Patient underwent Bentall procedure. He recovered well from the surgery and was discharged home with close follow up arranged.

Aortic dissection typically presents with tearing chest pain and severe hemodynamic compromise. Painless dissection, like in this case, is relatively rare. The case underscores relatively low sensitivity of TTE in detection of aortic dissection. Other imaging modalities, for the search of aortic pathology, should always be considered, especially in patients with risk factors. Our patient had at least two risk factors for aortic dissection: BAV and history of hypertension. Negative TTE for aortic dissection and diagnosis of another disorder (in this case HCM) should never let our guard down.

Figure 1. A- The standard 12-lead electrocardiogram; B- Chest X-ray; C- Cardiac magnetic resonance scans showing hypertrophic cardiomyopathy and D- aortic dissection; E- Computed tomography scans showing bicuspid aortic valve and F- aortic dissection.
A 29-year-old male with history of untreated hypertension, hospitalized for few days in local hospital due to pneumonia, was referred to our institution for further evaluation with suspicion of myocarditis and hypertrophic cardiomyopathy. On admission patient suffered from dyspnea and cough. He denied syncope or any episode of chest pain. On physical examination the patient was normotensive, with normal body temperature. Auscultation revealed bilateral rales. The liver was not enlarged, jugular veins were normal, there was no oedema of lower extremities. The baseline level of CRP was 4.4 (range 0–0.5) mg/dl, high-sensitivity cardiac troponin T was 324 (range 0–14) ng/L, and NT-proBNP was 3285 (range 0–125) pg/mL. The standard 12-lead electrocardiogram demonstrated sinus rhythm, left atrial enlargement and left ventricular (LV) hypertrophy with non-specific ST segment and T-wave abnormalities (Fig. 1A). The chest X-ray showed bilateral lung consolidation and pulmonary congestion (Fig. 1B). Transthoracic echocardiography (TTE) revealed significant asymmetric LV hypertrophy with preserved LV ejection fraction. Moreover bicuspid aortic valve (BAV) with moderate regurgitation and mild dilatation of the ascending aorta (41 mm) were found. Due to suspicion of myocarditis, cardiac magnetic resonance (CMR) imaging was performed. CMR confirmed hypertrophic cardiomyopathy (HCM) with the maximal LV wall thickness of 32 mm and increased myocardial mass (LV mass index 149 ml/m², range 68–103). Systolic function of LV and right ventricle (RV) were mildly decreased (LVEF 54%, range 57–74%; RVEF 45%, range 48–74%). Moreover ascending aortic dissection was detected (Fig. 1C, 1D). Patient was transferred to the computed tomography (CT) unit to confirm the diagnosis. Aortic dissection originated in aortic root and involving descending aorta was detected (DeBakey Type I). The maximal diameter of the ascending aorta was 44 mm with no evidence of coronary arteries involvement (Fig. 1E, 1F). Patient underwent Bentall procedure. He recovered well from the surgery and was discharged home with close follow up arranged.

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