Unexpected transesophageal echocardiography tee finding: mediastinal lipomatosis fakes an aortic intramural haematoma

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Mediastinal lipomatosis (ML) is a benign condition characterized by accumulation of fatty tissue. This condition is usually related to spontaneous or iatrogenic corticosteroid-induced Cushing syndrome or to obesity (1). Although it is an asymptomatic disorder, it can rarely result in compression of adjacent organs with thoracic pain, dyspnea, cough, dysphonia, and dysphagia. In this report we describe a case of fatty infiltration around descending aortic wall in a patient with autoimmune disorders, dilated cardiomyopathy and mitral insufficiency, simulating an intramural hematoma (IMH). A 49-year-old obese woman patient presented to our Heart Failure Outpatient Clinic for evaluation of progressive shortness of breath and sharp intermittent chest pain episodes. She was affected by dilated cardiomyopathy, systemic lupus erythematosus, rheumatoid arthritis on treatment with glucocorticoids, hyperlipidemia on treatment with statins and moderate renal dysfunction. On physical examination, her heart rate was 60 beats/min and regular, blood pressure was 130/70 mmHg bilaterally, carotid and peripheral pulses were normal bilaterally. A grade III/IV systolic murmur was heard at cardiac apex. Lung auscultation did not reveal any dry inspiratory crackles, as well as expiratory squeaks. On examination of extremities, neither digital clubbing or cyanosis or peripheral edema were noted. Trans-thoracic echocardiography showed mild left ventricular dilatation [left ventricular diastolic diameter (LVDD) 6 cm; LVDD/Body Surface Area (BSA) 3.3 cm/m²] with moderate reduction of global systolic function (ejection fraction 40%) and severe-moderate mitral regurgitation [effective regurgitant orifice area (EROA) was 4 cm² in site of an eccentric jet directed toward the side wall of the left

atrium) without any pericardial or pleural effusions. We decided to perform trans-esophageal echocardiogram (TEE) to screen the patient for percutaneous mitral valve repair (MVR) using the MitraClip system. Multiplane TEE showed moderate functional mitral regurgitation (3+/4+) associated with an unexpected remarkable new detail not consistent with previous diagnosis of precedent acute aortic syndrome. Indeed, withdrawing the TEE probe to visualize the descending aorta in short-axis (SAX) (0°) and long-axis (90°) views from the coeliac trunk to the left subclavian artery, as we use to perform to complete the exam, we recognized an hypoechoic, well-defined, crescent-shaped thickening of anterior aortic wall (>10 mm), mimicking IMH (Figure 1A). The descending aorta was no- dilated (diameter 2×2.2 cm²) and TEE assessment did disclose neither intimal flap nor laminar flow. The aortic lumen's shape was preserved and the luminal wall was curvilinear and smooth. Color Doppler analysis detected a signal of demodulated systolic flow inside the crescent-shaped thickening of aortic wall, as intercostal vessels or aortic vasa-vasorum (Figure 1B). Therefore, in order to make an accurate differential diagnosis among aortic syndromes, a contrast-enhanced multidetector computed tomography (MDCT) was practiced, showing ML around descending aorta wall, without any sign of aortic dissection, and around the free wall of right ventricle.

Discussion

ML is a benign condition, related to obesity, iatrogenic o spontaneous Cushing syndrome. Rarely is a symptomatic condition, needing surgical decompression or reduction of



Figure 1 Mid esophageal descending aorta short-axis (SAX) view (0°) by multiplane trans-esophageal echocardiogram (TEE) showed the transverse axis of the descending aorta with hypoechoic, well-defined, crescent-shaped thickening of anterior aortic wall (>10 mm), mimicking intramural hematoma (IMH). Color Doppler analysis detected a signal of demodulated systolic flow inside the crescent-shaped thickening of aortic wall, as intercostal vessels or aortic vasa vasorum.



Figure 2 CT angiography (CTA): axial scan clearly demonstrates visualization of descending aorta without flap of dissection, with homogeneous high density inside. Near the aorta and the free wall of right ventricle, focal bulging with negative density is revealed (ROI: –88 HU) with a CT finding relates with absolute certainty to fat accumulation [mediastinal lipomatosis (ML)].

the dosage of corticosteroids in the less severe forms (2). Few cases of ML are described in literature. Bulakci *et al.* reported a case of ML compressing the right ventricular outflow tract in a patient with Bechet disease, diagnosed with cardiac magnetic resonance imaging (MRI) (3). In addition, Sorhage *et al.* described an intrathoracic

obstruction related to ML of the upper airway leading to symptoms of bronchial asthma (4). Finally Peek et al. reported a rare case of ML and severe aortic stenosis, mimicking an enormous cardiomegaly in which a plain chest X-ray showed bilateral widening of the mediastinal silhouette (5). In our case, TEE showed a crescentshaped thickening of the anterior wall of descending aorta, mimicking an IMH. Of note, IMH can be difficult to distinguish from a thrombosed false lumen of classic aortic dissection because these can be both appear as a crescentshape thickening of the aortic wall. The clinical scenario of our patient pushed us to require an urgent total body-MDCT to make the differential diagnosis and to exclude an aortic dissection. MDCT has greatly improved imaging of acute thoracic aortic (6-11) disease with excellent contrast and spatial resolution after administration of intravenous contrast medium. In our reported case MDCT allowed to distinguish the lumen of descending aorta (with perfect opacification in the arterial phase without signs of dissection) from the excess of adipose tissue surrounded the thoracic aorta. A region of interest (ROI) positioned in the context of component closely adjacent to the aorta, showing negative value, excluded the presence of aortic dissection supporting the evidence of ML along the descending aorta. Moreover redundant adipose tissue, with the same density of the tissue around the aorta, surrounded the right ventricle (Figures 2,3). We have scheduled a new MDCT scan for follow-up in next six months. To our knowledge, this is the first reported case describing ML around aortic wall simulating an IMH.



Figure 3 CT angiography (CTA): multi planar reconstruction (MPR) coronal reconstruction shows clearly the extent of the accumulation of fat along the descending aorta (green arrows).

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Footnote

Conflicts of Interest: The authors have no conflicts of interest to declare.

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