Heart & Lung 42 (2013) 382–386

Contents lists available at SciVerse ScienceDirect

Heart & Lung

journal homepage: www.heartandlung.org

Two rare conditions in an Eisenmenger patient: Left main coronary artery compression and Ortner's syndrome due to pulmonary artery dilatation

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ARTICLE INFO

Article history:
Received 29 January 2013
Received in revised form
28 May 2013
Accepted 2 June 2013
Available online 5 July 2013

Keywords:
Eisenmenger syndrome
Pulmonary hypertension
Pulmonary dilatation
Left main compression
Ortner's syndrome

ABSTRACT

The left-main coronary artery extrinsic compression due to enlarged pulmonary artery has been described in several case series. Ortner's syndrome is also a rare condition in some cardiovascular disorders. There have been no reports about these two rare conditions in the same patient. Hence, we report a very rare case of an Eisenmenger patient with severe pulmonary hypertension and dilated pulmonary artery which has compressed the left main coronary artery, severely narrowing it, and the left laryngeal recurrent nerve with subsequent Ortner's syndrome and brief literature review.

Introduction

The extrinsic compression syndrome consisting of simultaneous severe narrowing of LMCA and hoarseness as a manifestation of recurrent laryngeal nerve paralysis due to compression by dilated pulmonary artery has been rarely reported. LMCA stenosis is most frequently seen in elderly as a consequence of atherosclerosis.[1,2] However, there have been reports of its extrinsic compression known as the left main coronary artery compression syndrome.[3] This can be due to anomalous origin of the left coronary artery and its course between the ascending aorta and pulmonary artery trunk or, due to compression by a dilated pulmonary artery trunk in various congenital heart diseases, pulmonary hypertension (Eisenmenger syndrome) or some inflammatory diseases (e.g., syphilis).[1,4] Ortner's or cardiovocal syndrome is a rare condition that can be seen in cardiac patients with the enlarged cardiac chambers or dilated large blood vessels. It is a consequence of compression of the recurrent laryngeal nerve and its paralysis.[2] This article presents a patient with Eisenmenger syndrome associated with rather rare complications.

Case report

In July 2012, a 37-year-old female patient was admitted to our Cardiology department because of shortness of breath and exertional fatigue. She was aware of having congenital heart disease since childhood but was not operated or regularly followed up. On admission, she was hypotensive (85/65 mm Hg), eupneic, cyanotic (oxygen saturation at rest, measured by pulse oximetry was 92%, and after 6 min walking distance of 389 m it decreased to 87%), with hoarseness of voice unexpected with her tiny stature (body height 150 cm and body weight 38 kg, body surface area 1.27 m^2 according to Mosteller R. calculator). Physical examination revealed a systolic 3/6 ejection murmur at the left parasternal border with fixed splitting of the second heart sound. ECG showed sinus rhythm, right bundle brunch block, downward ST depression in diaphragmal and precordial leads. Chest X-ray showed cardiomegaly and enlarged pulmonary artery. Transthoracic echocardiography revealed a 22 mm large ostium primum atrial septal defect (ASD); right ventricular enlargement (4.0 cm) and hypertrophy with dilated

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0147-9563/$ – see front matter © 2013 Elsevier Inc. All rights reserved.
http://dx.doi.org/10.1016/j.hrtlng.2013.06.001

pulmonary artery (diameter of 43 mm). See enclosed video clip. The right ventricular systolic pressure of about 150 mm Hg was calculated using the maximum velocity of tricuspid regurgitation obtained by continuous Doppler. The left ventricle was not dilated and had preserved systolic function. The pulmonary artery trunk: aortic root ratio was 1.77. Heart catheterization with angiography and oxygen pulmonary vascular resistance (PVR) testing showed high PVR (21.7 Wood Units) and irreversible pulmonary hypertension with over systemic pressure values in the right ventricle and pulmonary artery (116/0–9 mm Hg and 120/49/76 mm Hg respectively). Coronary angiography indicated significant ostial narrowing of the left main coronary artery (90%) free of atherosclerotic changes (Fig. 1). Suspicion of extrinsic compression was proven by cardiac 64-slice multidetector computed tomography (MDCT), which confirmed compression and displacement of the aortic root and left coronary artery by the dilated pulmonary artery (Figs. 2 and 5). Stress testing for ischemia documentation was not performed due to Eisenmenger phase of the congenital defect and previous poor effort tolerance. Coronary flow reserve (CFR) testing with maximal dose of adenosine failed to achieve adequate flow increase in the distal segment of the left anterior descending artery (CFR LAD 1.5 m/s). Anatomic and functional confirmation of the significance of LM stenosis was the basis for treatment. After giving informed consent, our patient underwent a percutaneous intervention of the left main stenosis with stenting, in order to protect the lumen from collapsing under the high-pressure extrinsic compression. Implantation of 4 × 15 mm XIENCE™ V Everolimus Eluting Coronary Stent System (Abbott Vascular, A subsidiary of Abbott Laboratories, Santa Clara, CA) in the left main coronary artery was performed under intravascular ultrasound (IVUS) guidance, which also confirmed adequate stent deployment. Intervention proceeded uneventfully. Coronary flow reserve improved to 2.0 m/s. Medicament treatment of pulmonary hypertension with sildenafil, as the only available drug in our country, was initiated carefully with low doses 2 × 12.5 mg to avoid further systemic arterial hypotension. Six months later, MDCT follow up imaging study was performed and confirmed stent patency (Figs. 3 and 4). Clinically, she is relived of symptoms.

Along with the cardiac investigations, she was examined by an otorhinolaryngologist who performed indirect laryngoscopy and diagnosed the left recurrent laryngeal nerve palsy, with the left vocal fold in paramedian position, narrowing the respiratory tract. We performed various exams (immunology tests and contrast enhanced CT of the neck) to exclude other possible causes for hoarseness of voice.

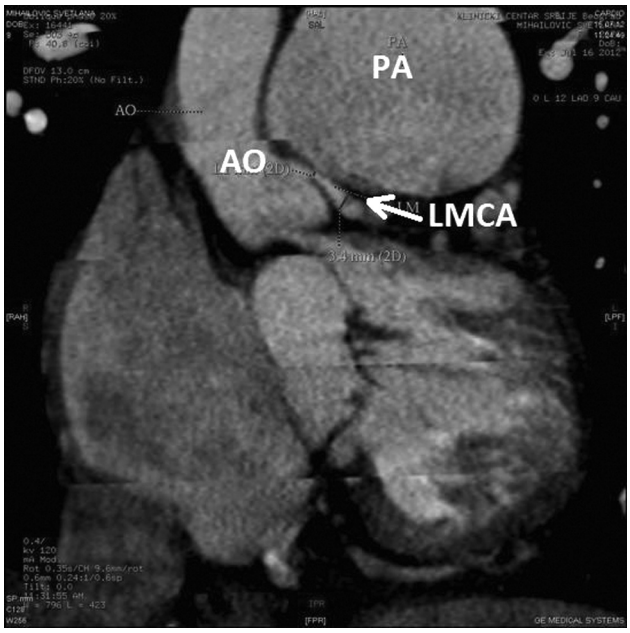

Fig. 2. External left main coronary artery (LMCA) compression by dilated pulmonary artery (PA) on cardiac 64-slice MDCT scan. AO = aorta.

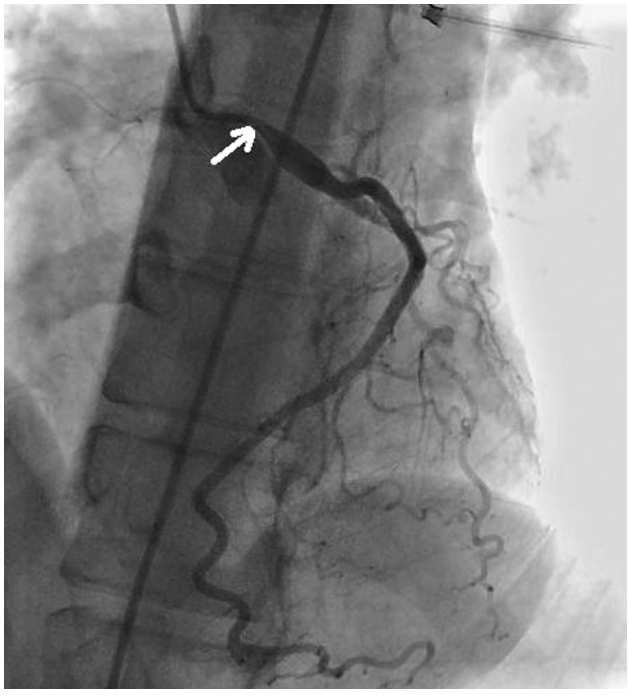

Fig. 1. Hemodynamic significant ostial left main stenosis (white arrow) on coronary angiography.

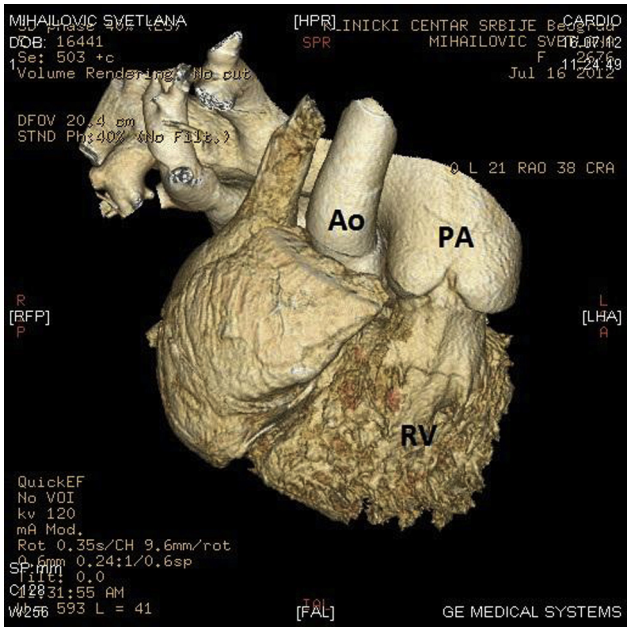

Fig. 3. Volume rendered (3 dimensional) MDCT image. AO = aorta, PA = pulmonary artery, RV = right ventricle.