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Case Report

Severe pulmonary hypertension secondary to giant chronic dissecting aortic aneurysm presenting as chronic right heart failure

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ABSTRACT

We report on an 81-year-old woman who presented to hospital with effort dyspnea and leg edema. Transthoracic echocardiography revealed normal left ventricular function but marked dilated ascending aorta with an intimal flap in the proximal ascending aorta, severe tricuspid regurgitation, and severe pulmonary hypertension. Contrast enhanced multi-slice computerized tomography of the chest was performed for detailed evaluation of aortic dissection and this also showed the intimal flap of the ascending aortic dissection (DeBakey type II) and giant ascending aorta (10.5 cm) compressing the main and right pulmonary artery. This is the first case where chronic right heart failure is caused by chronic aortic dissection.

<**Learning objective:** Although dissecting aortic aneurysms generally present acutely, they may have chronic presentation. Obstruction of the pulmonary artery caused by a dissecting aortic aneurysm, which mimics acute pulmonary thromboembolism, is one of the rare acute presentations. In this paper, we suggest a giant dissecting aortic aneurysm compressing pulmonary artery substantially may also lead severe pulmonary hypertension and symptoms of chronic right heart failure.>

Introduction

Acute aortic dissection is a life-threatening condition and giant aneurysms of the ascending aorta may cause symptoms by compressing adjacent structures. Although rare, obstruction of the pulmonary artery caused by a dissecting aneurysm or hemorrhage of the ascending aorta can mimic pulmonary thromboembolism, but chronic right heart failure due to chronic dissecting aortic aneurysm has not been reported. Here we describe a patient presenting with leg edema and exertional dyspnea having a chronic giant aortic dissection that compressed the pulmonary trunk and led to severe pulmonary hypertension.

Case report

An 81-year-old woman presented to hospital with effort dyspnea and leg edema. She had undergone cardiac operation under cardiopulmonary bypass due to myxoma that had been diagnosed following a cerebrovascular accident eight years previously. Her preoperative examination showed normal left ventricular systolic function, normal pulmonary artery pressure, and mild coronary artery disease. Her operation and postoperative period had been uneventful. While she exhibited no symptoms of hypertension, she started to experience effort dyspnea which increased progressively, and moderate leg edema in the previous month. Physical examination revealed a blood pressure of 130/80 mmHg, a pulse of 92 beats/min, and respiration of 28 breaths/min. S2 was accentuated with a 3/6 systolic murmur on the mesocardiac area and her lungs were clear on auscultation. Laboratory examination revealed mildly elevated blood urea nitrogen (33 mg/dl, normal range < 20 mg/dl) levels. Other biochemical tests including D-dimmer and complete blood count were all within the normal range. Brain natriuretic peptide could not be measured. Sinus rhythm with nonspecific ST-T changes was observed on electrocardiography. Chest radiography showed severe enlargement of the mediastinum with pronounced bulging to the right hiler area (Fig. 1). 2D and Doppler transthoracic echocardiography revealed marked dilated ascending aorta (10 cm) with an intimal flap in the proximal ascending aorta (Fig. 2), severe tricuspid regurgitation, and severe pulmonary hypertension (95 mmHg, estimated from tricuspid regurgitation and inferior vena cava) (Fig. 3), dilated right atrium, mildly hypertrophic normal left and right ventricular function, pseudonormal left ventricular diastolic

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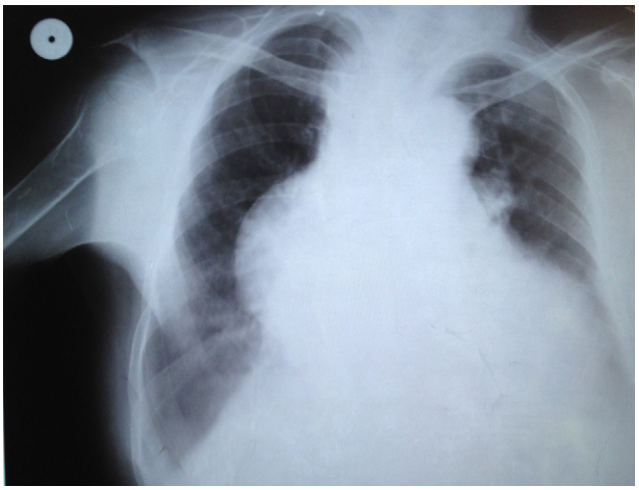

Fig. 1. Chest X-ray showed severe enlargement of the mediastinum.

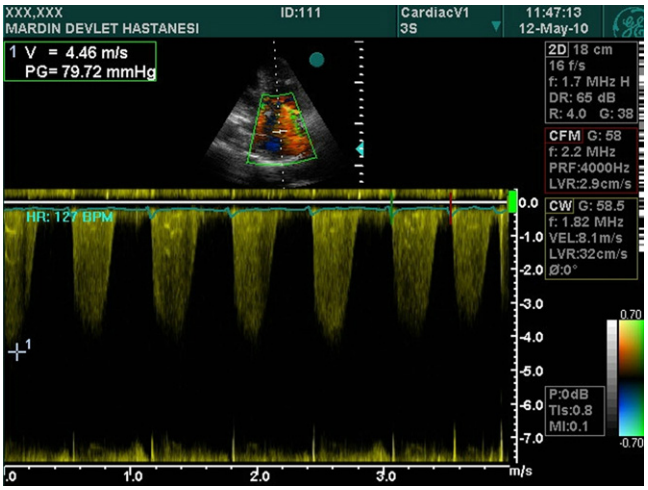

Fig. 3. Severe pulmonary hypertension estimated from tricuspid regurgitation.

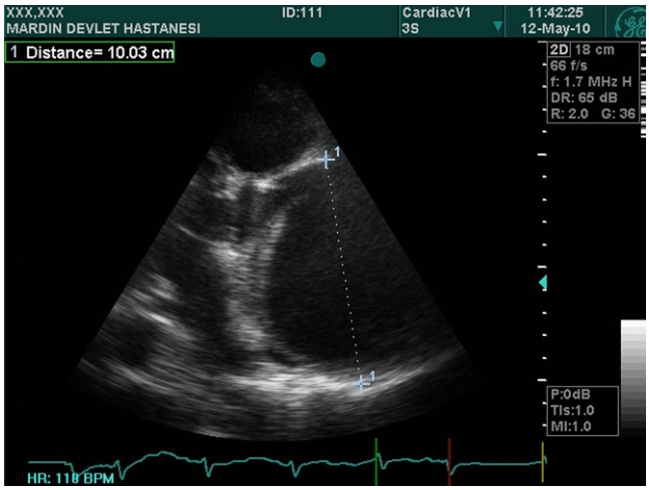

Fig. 2. Transthoracic echocardiography revealed marked dilated ascending aorta (10 cm) with an intimal flap in the proximal ascending aorta.

pattern, mildly degenerative aortic valve, and mild to moderate aortic regurgitation. Contrast-enhanced multi-slice computerized tomography (CT) of the chest was performed for detailed evaluation of aortic dissection and this also showed the intimal flap of the ascending aortic dissection (DeBakey type II) and giant ascending aorta (10.5 cm) compressing the main and right pulmonary artery (Fig. 4). Lung parenchymas were normal and there was no evidence of pulmonary thromboembolic disease on thorax CT. A respiratory function test was performed to exclude severe pulmonary disease and the test revealed a mildly restrictive pattern. She had neither any additional systemic disease nor any drug addiction.

According to this finding, we diagnosed pulmonary hypertension and clinical right heart failure as a consequence of pulmonary artery compression. We proposed surgical intervention to the patient but the patient refused. The patient was discharged but had furosemide prescribed in addition to her previous medical therapy of candesartan and metoprolol.

Discussion

Chronic, non-dissecting, thoracic aortic aneurysms are considered as possible causes of acquired pulmonary artery obstruction, which is not surprising in view of the close anatomical relationship between the ascending thoracic aorta and the pulmonary artery. Conversely, acute aortic dissection infrequently causes pulmonary artery obstruction [1], partly because of the nature of this acute condition. Only a few cases have been reported in the literature [2–8].

Aortic dissection is characterized by a laceration of the aortic intima and the inner layer of the aortic media that allows blood to course through a false lumen in the outer third of the media [9]. The leakage of blood from the aorta into the periaortic space can be seen in aortic dissection and the presence of a hematoma surrounding the pulmonary artery is the main mechanism responsible for pulmonary artery obstruction [3,4,8]. Compression of the main and right pulmonary arteries by the large dissecting hematoma can easily be explained by the proximity of the ascending aorta to the right of the pulmonary valve [3,6].

Obstruction of the pulmonary artery caused by a dissecting aneurysm or hemorrhage of the ascending aorta can often be catastrophic and mimic acute pulmonary thromboembolism [2,5–8].

However, this patient presented to hospital with effort dyspnea and leg edema and our diagnostic investigations demonstrated giant dissecting aneurysm of the ascending aorta which compressed the right and main pulmonary arteries and led to severe pulmonary hypertension. We thought that aortic dissection may be due to previous cardiac surgery and aortic cannulation. Dissection and compression of the pulmonary artery was probably recent because echocardiography and electrocardiography did not show the sign of long-standing pulmonary hypertension. Pseudonormal left ventricular diastolic pattern was probably secondary to systemic hypertension and mild left ventricular hypertrophy. Although we could not document anatomically because of the patient's refusal of surgery, evidence from contrast enhanced multi-slice CT and chronic presentation of the patient suggested that the observed compression of the pulmonary arteries was due to aortic dilatation rather than rupture. According to the literature, this is the first case where chronic right heart failure is caused by chronic aortic dissection and it is among the largest (10.5 cm) chronic dissecting aneurysm of the ascending aorta reported, second only to the case of Ryomoto et al. in which an aortic diameter of 14 cm is reported [10].

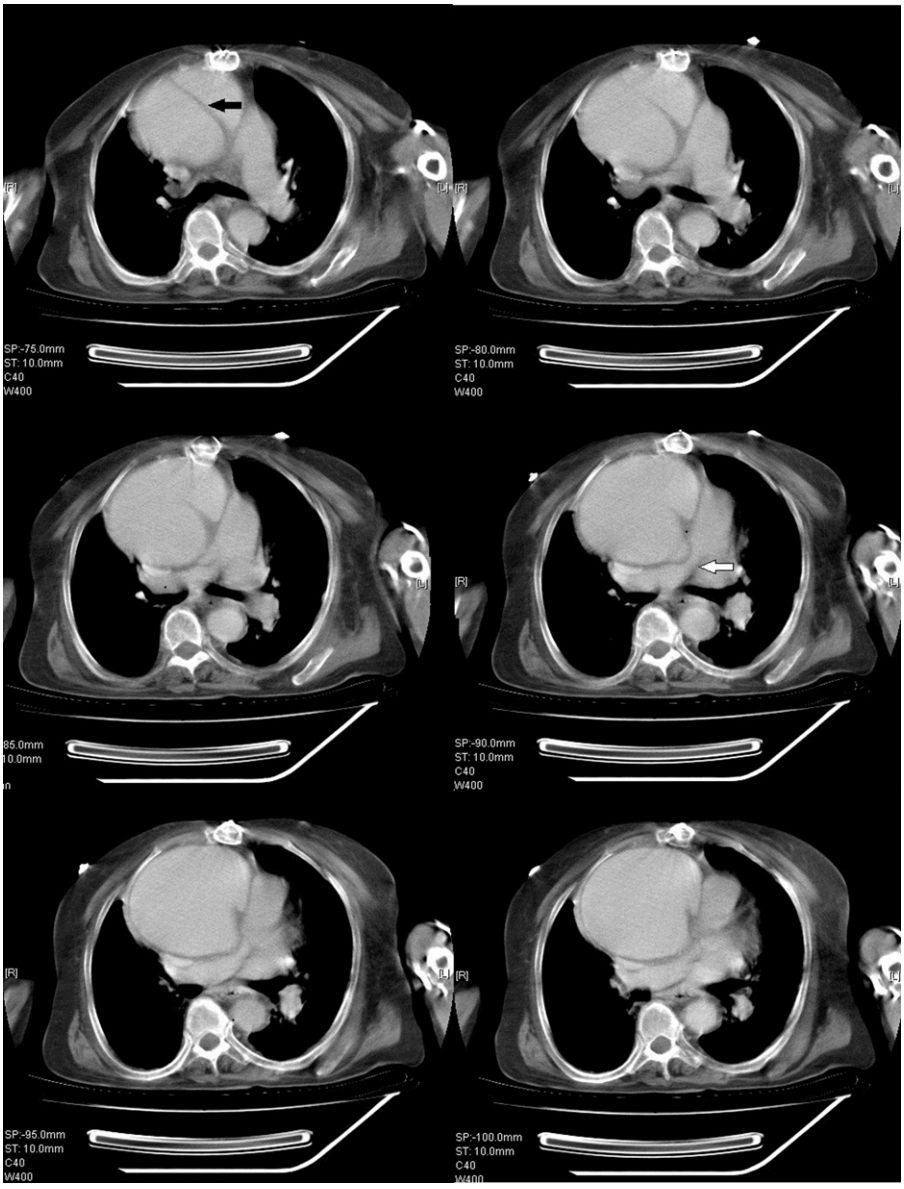

Fig. 4. Contrast enhanced computerized tomography of the chest showed the intimal flap (black arrow) of the ascending aortic dissection and giant ascending aorta compressing the main and right pulmonary artery (white arrow).

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