Aortic Dissection Presenting with Secondary Pulmonary Hypertension Caused by Compression of the Pulmonary Artery by Dissecting Hematoma: A Case Report

The rupture of an acute dissection of the ascending aorta into the space surrounding the pulmonary artery is an uncommon occurrence. No previous cases of transient pulmonary hypertension caused by a hematoma surrounding the pulmonary artery have been documented in the literature. Herein, we report a case of acute aortic dissection presenting as secondary pulmonary hypertension.

A 64-year-old woman with a medical history consisting only of hypertension came to the emergency room (ER) with complaints of chest pain and shortness of breath for 4 days. On admission, she presented with vague discomfort in the right upper abdomen, anterior chest tightness and exertional dyspnea (class III according to the functional classification of the New York Heart Association). Physical examination revealed a blood pressure of 100/80 mmHg, a pulse of 88 beats/min, respiration of 34 breaths/min and a body temperature of 36.2°C. There were crackling sounds in both lower lung fields, but the cardiac sounds were normal. Various laboratory tests were performed on admission, including a blood chemistry profile, coagulation studies and a complete blood count, in which the cardiac enzymes revealed leukocytosis and elevated hepatic enzymes without evidence of myocardial infarction. A chest X-ray taken on admission showed the presence of cardiomegaly without mediastinal widening, patch infiltration in the right lower lobe and blunting of the bilateral costophrenic angles (Fig. 1A). The electrocardiogram disclosed sinus tachycardia of 100 beats/min with no S-T or T wave changes. The Doppler echocardiography revealed an ejection fraction (EF) of 56.4%, moderate to severe tricuspid regurgitation, scanty pericardial effusion and bilateral pleural effusions, hepatic congestion, and an increased systolic pulmonary arterial pressure (SPAP) of 75 mmHg (normal range; 15–30 mmHg), suggesting the presence of severe pulmonary hypertension. After a preliminary diagnosis of “idiopathic right heart failure”, the patient was admitted to the medical intensive care unit for treatment. On the 4th day following admission, atrial fibrillation and a sudden drop in systolic blood pressure to 70 mmHg appeared.

Index terms: Radiograph, aorta Computed tomography (CT), aorta Aorta, dissection Dissection, pulmonary hypertension

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Fig. 1. A 64-year-old woman with ascending aortic dissection presenting as secondary pulmonary hypertension.
A. An admission chest radiograph shows cardiomegaly with tortuous thoracic aorta, patch infiltration in right lower lobe, and blunting in both costophrenic angles. Mediastinal widening is not definite.
B. Preoperative transthoracic echocardiography shows an undulating intimal flap (arrow), dilated ascending aorta, and pericardial effusion (E).
C. Preoperative chest CT demonstrates intimal flap (arrow) of ascending aortic dissection (DeBakey type 2), pericardial effusion, and bilateral pleural effusions.
D. Chest CT image obtained 1.5 cm above C shows compression of the pulmonary artery by hematoma. Expanding dissection causes aneurysmal dilatation of ascending aorta (4 cm in diameter).
E. Schematic drawing in craniocaudal view with cross section of the ascending aorta and pulmonary artery. Right and left pulmonary arteries (rPA & lPA) are severely compressed by the hematoma (H), but coronary ostia are intact. The tear point is situated at the posterolateral wall (arrow) of the ascending aorta.
Aortic Dissection Presenting with Secondary Pulmonary Hypertension Caused by Compression of the Pulmonary Artery by Dissecting Hematoma

Acute aortic dissection is a life-threatening condition requiring prompt diagnosis and treatment. 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 (8, 9). In aortic dissection, the leakage of blood from the aorta into the periaortic space is not rare. Although compression of the pulmonary artery by a chronic aneurysm is a frequent occurrence, the presence of a hematoma surrounding the pulmonary artery causing pulmonary hypertension (right heart failure) accompanied by acute aortic dissection has not previously been reported. Rupture of the false sac into any of the neighboring structures is a possible occurrence, with a potentially fatal outcome. The dissecting hematoma may also expand and compress any contiguous structure. Displacement of the right ventricular outflow tract and 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 (2, 7). The ascending aorta and the pulmonary trunk are enclosed within a common tube of the visceral pericardium. The right pulmonary artery is horizontal and immediately dorsal to the ascending aorta. Similar compression of the pulmonary artery by an acute dissecting aneurysm has been described at autopsy by Buja et al. (3). They demonstrated the anatomic relation between the ascending aorta and the pulmonary outflow tract, which share the same adventitia layer, as well as the relation between the ascending aorta and the right pulmonary artery. Castaner et al. described a case of mediastinal hematoma dissecting the sheath of the pulmonary arteries (6). Rupture usually occurs in the posterior wall of the ascending aorta, adjacent to the right pulmonary artery. Blood seeping from the ruptured aortic false lumen can reach the interstitial space bordering the pulmonary arteries.

In our patient, the clinical manifestations, including the sudden onset of acute respiratory distress and hypotension, in spite of the history of hypertension, are suggestive of aortic dissection. However, cardiomegaly with patch infiltration mimicking pulmonary edema in the plain chest radiograph, and increased systolic pulmonary artery pressure with severe tricuspid regurgitation in the transthoracic echocardiograph, are indications of pulmonary hypertension and right heart failure. The initial chest CT scan clearly confirms the presence of dissection of the ascending aorta and compression of the right and left pulmonary arteries by the acute expanding hematoma. When dissection occurs at the posterior wall of the ascending aorta, as in our case, the hematoma can compress the right pulmonary artery which lies posterior to the ascending aorta. However, our case shows compression of both pulmonary arteries (Fig. 1E).

Although rare, obstruction of the pulmonary artery caused by a dissecting aneurysm or hemorrhage of the ascending aorta can mimic pulmonary thromboembolism (7). The radiographic changes resulting from pulmonary thromboembolism are normal, oligemia, hemorrhage and infarction. The CT scan can depict these findings. The distinction between these entities is important, since anticoagulation therapy for suspected embolism is contraindicated if the patient has aortic dissection. Evidence of acute mediastinal widening favors the diagnosis of dissection of the aorta. However, our case showed a congestive heart failure pattern in the plain chest radiograph.

Rupture of an aortic dissection can cause pulmonary artery compression presenting with secondary pulmonary hypertension or can mimic pulmonary embolism. Although acute pulmonary hypertension caused by aortic dissecting hematoma is rare, the potential risk must be borne in mind when treating patients with this condition.

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3. Buja LM, Ali N, Fletcher RD, Roberts WC. Stenosis of the right pulmonary ostia were intact. A graft suture was done in the ascending aorta. Postoperative CT showed re-expansion of the compressed pulmonary artery. The patient’s postoperative systolic pulmonary arterial pressure was 20 mmHg and she made an uneventful recovery without residual sequelae.

DISCUSSION

Acute aortic dissection is a life-threatening condition requiring prompt diagnosis and treatment. 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 (8, 9). In aortic dissection, the leakage of blood from the aorta into the periaortic space is not rare. Although compression of the pulmonary artery by a chronic aneurysm is a frequent occurrence, the presence of a hematoma surrounding the pulmonary artery causing pulmonary hypertension (right heart failure) accompanied by acute aortic dissection has not previously been reported. Rupture of the false sac into any of the neighboring structures is a possible occurrence, with a potentially fatal outcome. The dissecting hematoma may also expand and compress any contiguous structure. Displacement of the right ventricular outflow tract and 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 (2, 7). The ascending aorta and the pulmonary trunk are enclosed within a common tube of the visceral pericardium. The right pulmonary artery is horizontal and immediately dorsal to the ascending aorta. Similar compression of the pulmonary artery by an acute dissecting aneurysm has been described at autopsy by Buja et al. (3). They demonstrated the anatomic relation between the ascending aorta and the pulmonary outflow tract, which share the same adventitia layer, as well as the relation between the ascending aorta and the right pulmonary artery. Castaner et al. described a case of mediastinal hematoma dissecting the sheath of the pulmonary arteries (6). Rupture usually occurs in the posterior wall of the ascending aorta, adjacent to the right pulmonary artery. Blood seeping from the ruptured aortic false lumen can reach the interstitial space bordering the pulmonary arteries.

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