Acquired Aorto-Pulmonary Fistula: a Case of Ruptured Aneurysm of the Thoracic Aorta

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Introduction

Aortopulmonary fistula is an uncommon but highly lethal condition. It is usually the result of a late complication of an aortic aneurysm. The most common cause is erosion of a false aneurysm of the descending thoracic aorta into the pulmonary artery. This complication results in the development of an acute left-to-right shunt, new murmur, and rapid right heart failure. We report on a 68-year-old man who presented with hoarseness and dyspnea with acquired aortopulmonary fistula resulting in a significant left-to-right shunt and a pulmonary embolism with review of the literature.

Case Report

A 68-year-old man, smoker, with a medical history of hypertension, hypercholesterolemia was admitted to a local hospital with 4-month history of hoarseness and shortness of breath. A grade 2/6 continuous murmur was heard over the third left sternal border. No pulse deficit or asymmetry of blood pressure was found. Electrocardiogram showed normal sinus rhythm, right ventricular systolic strain and no evidence of ischemia or previous infarction. An admission chest x-ray study revealed cardiomegaly, mild mediastinal widening, dilated main pulmonary artery and the right pulmonary artery and the presence of inhomogeneous opacity in the middle and lower lungs lobes especially in the right side (Fig. 1). Laryngeal examination revealed paralysis of the left vocal cord. The remainder of the larynx, hypopharynx, and oropharynx was normal. After a short period later from hospitalization, sudden severe onset of hemoptysis developed. On physical examination, the patient was in apparent distress with a blood pressure of 160/80mmHg in both arms and there were signs of heart failure. Repeated chest X-ray showed progression of the bilateral infiltrates and bilateral pleural effusion. The patient subsequently underwent Doppler echocardiography that revealed shunt flow, a finding suggesting the presence of patent ductus arteriosus. Mild mitral regurgitation with normal left ventricular function was also documented. Computer tomography studies of the neck and chest were performed to identify the cause of vocal cord paralysis. As a result of study, it was believed that the enlarged and upwardly displaced saccular aortic aneurysm originating from distal part of arcus aorta was responsible for the compression of the left recurrent laryngeal nerve and the left vocal cord paralysis (Fig. 2). The patient subsequently underwent cardiac catheterization. The angiogram revealed a normal left ventricular ejection fraction and nonsignificant coronary artery disease except for nondominant right coronary artery. Right heart catheterization revealed a severe right-sided pressure elevation: right atrium 3-4 mm Hg, right ventricle 50/0/5mmHg, pulmonary artery, 50/20 mm Hg, pulmonary artery capillary wedge pressure, 25mmHg; and shunt ratio, 4.1:1. During angiography, contrast dye was found to transfer into the pulmonary artery but shunt level could not be identified. To determine the shunt level transesophageal echocardiography study was performed. Transesophageal echocardiography demonstrated an aortopulmonary fistula from the descending aorta to the main pulmonary artery with a tearing point, a large clot adherent to the aortic aneurysm wall, an irregularity, a protruding thrombus particles at full length of descending aorta and Doppler study revealed shunt flow (Fig. 3,4). Since, findings have suggested that saccular aortic aneurysm ruptured into the pulmonary artery, emergent operation was planned. The risks of surgery were believed to be prohibitive, and the patient was then...
transferred to the experienced cardiothoracic center with cold upper and lower extremities, and poor urine output. Surgery was performed but the patient could not be weaned from respiratory support and died on the seventh postoperative day.

**Discussion**

Aortic aneurysms and aortic dissection are the most important pathologic processes that can develop along the aorta. Aortic aneurysms are commonly related to the atherosclerotic disease, with subsequent weakening of the media and expansion of the involved area. In the thoracic aorta, the complication of atherosclerotic aneurysm include acute aortic regurgitation, rupture into the mediastinum, pleural cavity and bronchi, as well as proximal and distal arterial dissection (1). Rupture and distal propagation with renal artery compromise may result in a death. In rare instances, fistula formation between the aorta and pulmonary artery may complicate dissection. A chronic disease states of the aortic wall, such as a giant cell aortitis, syphilitic or mycotic disease, or an endocarditis are the other causes of acquired aortopulmonary fistulas.

**Figure 1.** Posteroanterior chest roentgenogram reveals cardiomegaly, vascular redistribution, markedly dilated main pulmonary artery, inhomogeneous density in both lungs, especially in the right lung.

**Figure 2.** The CT scans at the level of thoracic aorta shows presence of inhomogeneous opacity in the right lung (white arrow), dilated pulmonary trunk, and thoracic saccular aneurysm containing thrombus (black arrow) in the inner surface and thrombus particles in the descending aorta (black arrowheads). But the aortopulmonary communication remains undiagnosed. The aneurysm arises just at the site of origin of the left subclavian artery.

DA: descending aorta, SAA: saccular aortic aneurysm

**Figure 3.** Transesophageal echocardiography view shows enlarged saccular aneurysm in the thoracic aorta and communication between the aorta and the pulmonary artery (arrow)(tearing site). Aneurysm contains massive thrombus in the inner surface (arrowheads).

PA: pulmonary artery, SAA: saccular aortic aneurysm

**Figure 4.** Doppler image shows shunt flow passing from the aorta into the pulmonary artery (arrow).
In 1924, Boyd (2) reviewed 4000 autopsy reports of thoracic aortic aneurysms, finding 1197 cases of rupture with a 4% occurrence of aortopulmonary fistula.

Transverse arch aneurysms may produce a variety of signs and symptoms due to compression of adjacent structures. More than 60% of patients with thoracic aortic aneurysms are asymptomatic, and the aneurysm usually is detected on chest radiography obtained for another reason. Among symptomatic patients, chest pain and back pain secondary to impingement of associated structures by the enlarging aneurysm are the most common complaints. In aortic dissection, chest pain is the most common symptom and occurs in 90% to 95% of patients (3). Less common physical findings in aortic dissection include hoarseness secondary to vocal cord paralysis from compression of the recurrent laryngeal nerve, and Horner’s syndrome (unilateral ptosis, miosis, and anhydrosis) due to compression of the superior cervical sympathetic chain. In patients with aortopulmonary fistula, the most common symptoms are chest pain and hemoptysis, shortness of breath, fever, or other respiratory symptoms (4). The hemoptysis, which is characteristically intermittent or recurrent, occurs when developed hematoma “leaks” into the bronchopulmonary tree. Due to the unusual clinical presentation in this case, it is not clear whether the rupture was acute or chronic. This interesting case of an acquired aortopulmonary fistula demonstrates the possibility of atypical clinical presentation in patients with aortic aneurysm leading to dissection.

Four imaging modalities, which are a complementary, can be used to make the definitive diagnosis of aortic aneurysm and its complications. In the past, fistulas were most frequently diagnosed by aortography and cardiac catheterization, if they were discovered before the patient’s death. Many more, however, were diagnosed post mortem. The use of radiological contrast and the attendant risk of anaphylaxis and contrast-induced nephropathy are the disadvantages of angiography. Echocardiography is a commonly used imaging technique that has become an important tool in the diagnosis of aortic dissection. Two-dimensional echocardiography has the capacity to visualize aortic rupture very well. It also has the advantage of being able to evaluate left ventricular function and to determine whether there is aortic insufficiency (5). If on physical examination one suspects the presence of a fistula, Doppler echocardiography can be used to further visualize the flow from one compartment to another. Veerbeek and associates (6) used transthoracic Doppler echocardiography in the diagnosis of an aortopulmonary fistula. However, the major disadvantage of transthoracic echocardiography is the inability to identify distal dissections. Transesophageal echocardiography, in which the transducer is at the end of a flexible gastroscopy, is a useful technique for detecting distal aortic dissection. Transesophageal echocardiography can be performed safely even in critically ill patients. The sensitivity of TEE for detecting both proximal and distal aortic dissection is 100% (7). Transesophageal echocardiography must become the diagnostic study of choice for suspected dissection because it is rapid, can be performed at the bedside, and is only minimally invasive. Computerized tomographic scanning and magnetic resonance imaging are the less invasive and highly accurate procedures, but they necessitate moving the patient from the emergency department to the radiology department.

In this case, all diagnostic tests were used for the definitive diagnosis, but only TEE was able to demonstrate aortopulmonary fistula clearly. Echocardiography has been shown to be an excellent technique for visualizing complications. Cardiac catheterization and aortography did reveal the presence of left-to-right shunting, but the findings were not satisfactory for the correct diagnosis of a ruptured aortic aneurysm and the exact identification of an aneurysm tearing site.

Early and accurate diagnosis is essential for appropriate treatment and the management of enlarging or leaking aneurysm (4). To date, only few cases have been published describing successful surgical management (8-10), as the observed mortality rate for surgical correction of aortic aneurysm with acute aorto-pulmonary fistula is very high.

In summary, aortopulmonary fistula resulting from rupture of an aortic aneurysm into the pulmonary artery should be kept in the differential diagnosis whenever patients with hypertension present with symptoms of pulmonary embolism and left-to-right shunt. One must use all the information and imaging techniques at hand to ensure a timely diagnosis. Although it is a highly lethal condition, early and accurate diagnosis is essential for appropriate treatment and life saving.
References