Spontaneous Rupture of the Ascending Thoracic Aorta in Young Man

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Abstract

Spontaneous rupture of the aorta without previous history of trauma, hypertension or apparent aortic pathology is rare. Delayed or nonoperative repair of this condition is usually lethal. We report spontaneous rupture of the ascending thoracic aorta in young man. A 20-year-old male who had complained of sudden onset of severe chest pain and dispnea was admitted to our hospital. Initially, acute type A closing aortic dissection was suspected but computed tomography (CT) showed no intimal flap, false lumen or aortic aneurysm.

Key Words: Spontaneous, rupture, aorta

(Rec.Date: Jun 30, 2014 Accept Date: Jul 19, 2014)

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Introduction

Spontaneous rupture of the ascending thoracic aorta without history of trauma, hypertension or apparent aortic pathology is rare but catastrophic disorder. In this case, repair of a spontaneous Ascending Thoracic Aorta rupture is described in young man.

Case Report

A 20-year-old male who had complained of sudden onset of severe chest pain and dyspnea was admitted to our hospital. On admission, he was unconscious and in a state of shock with a systolic blood pressure of 70 mmHg. His blood pressure improved after treatment by intravenous drip combined with dopamine. Electrocardiogram (ECG) no showed myocardial infarct. Chest radiography revealed an enlarged mediastinal and cardiac silhouette. Transesophageal echocardiogram (TEE) showed a low attenuation echogenic space around the aortic arch corresponding to the mediastinal hematoma but no intraaortic hematoma. The aortic root and aortic valve were normal. Initially, acute type A aortic dissection was suspected; but CT scanning did not show a false channel, aortic dissection or intramural hematoma. CT scan of the chest revealed a mediastinal hematoma (Fig. 1). His blood pressure fell to 70 mmHg and so an emergency operation was performed. A median sternotomy was performed. Surgery revealed a hematoma over the main pulmonary artery the ascending aorta and pericardia. The patient was placed on cardiopulmonary bypass with femoral artery access, after a rectal temperature of 24°C was achieved. The arcus aorta was normal. Thus arcus aorta was clamped before the innominate artery. Myocardial preservation was achieved by retrograde administration of intermittent hyperkalemic cold blood cardioplegia and additional topical cooling with ice slush. Before the aortic cross-clamp was removed, an additional bolus of warm blood cardioplegia (hot shot) was infused. Alpha-stat management of acid-base status was used.

We were find 15x20 mm a rupture in the anteromedial aspect of the ascending aorta approximately 40-50 mm from the aortic valve. The rupture site showed no evidence of dissection, cystic medial necrosis and atherosclerosis in the intima but pseudoaneurysm had produced a hematoma around the aortic wall without medial dissection. Furthermore, the adventitia of the pseudoaneurysm at the site of the rupture was walled off by organizing granulation tissue with hemorrhaging (Fig. 2,3). The rupture was replaced with a dacron patch and polypropylene running

suture (Fig. 4). Histopathology revealed normal aortic wall. Atherosclerosis, medial wall degeneration and cystic media necrosis did not reveal.

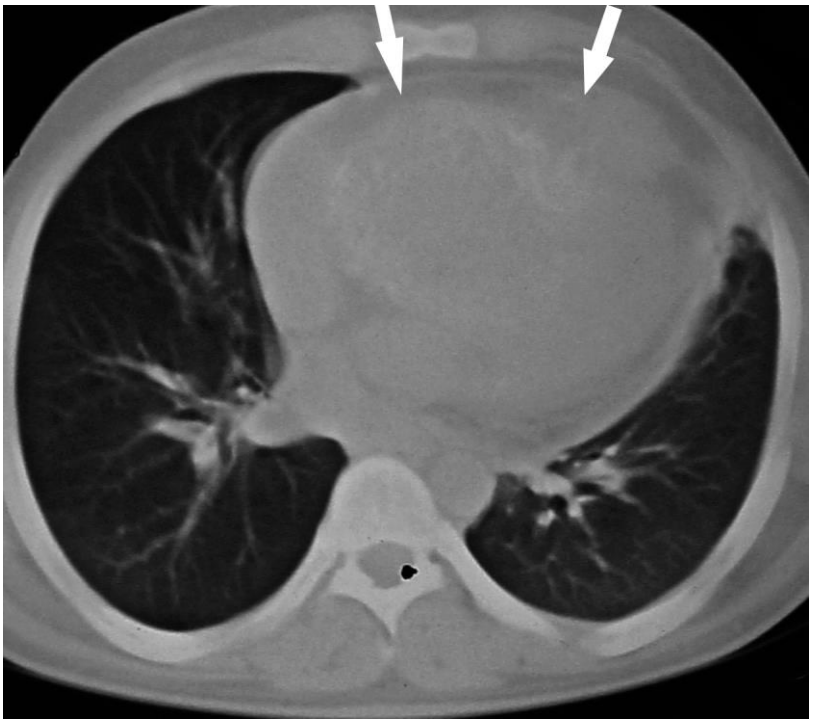

Figure 1. Computed tomography (CT) showed a large hematoma.

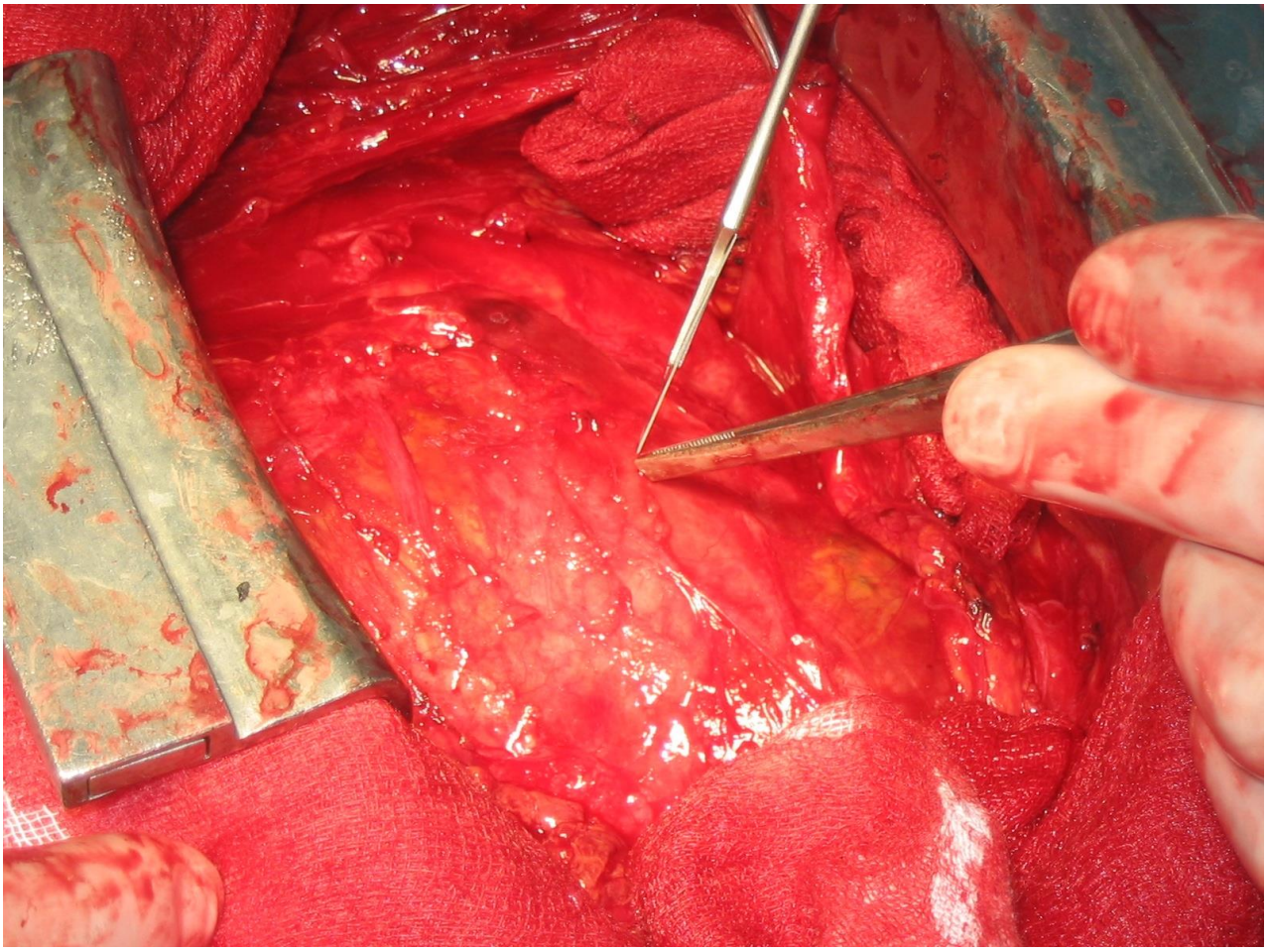

Figure 2. Intraoperative findings revealed pseudoaneurysm sack.

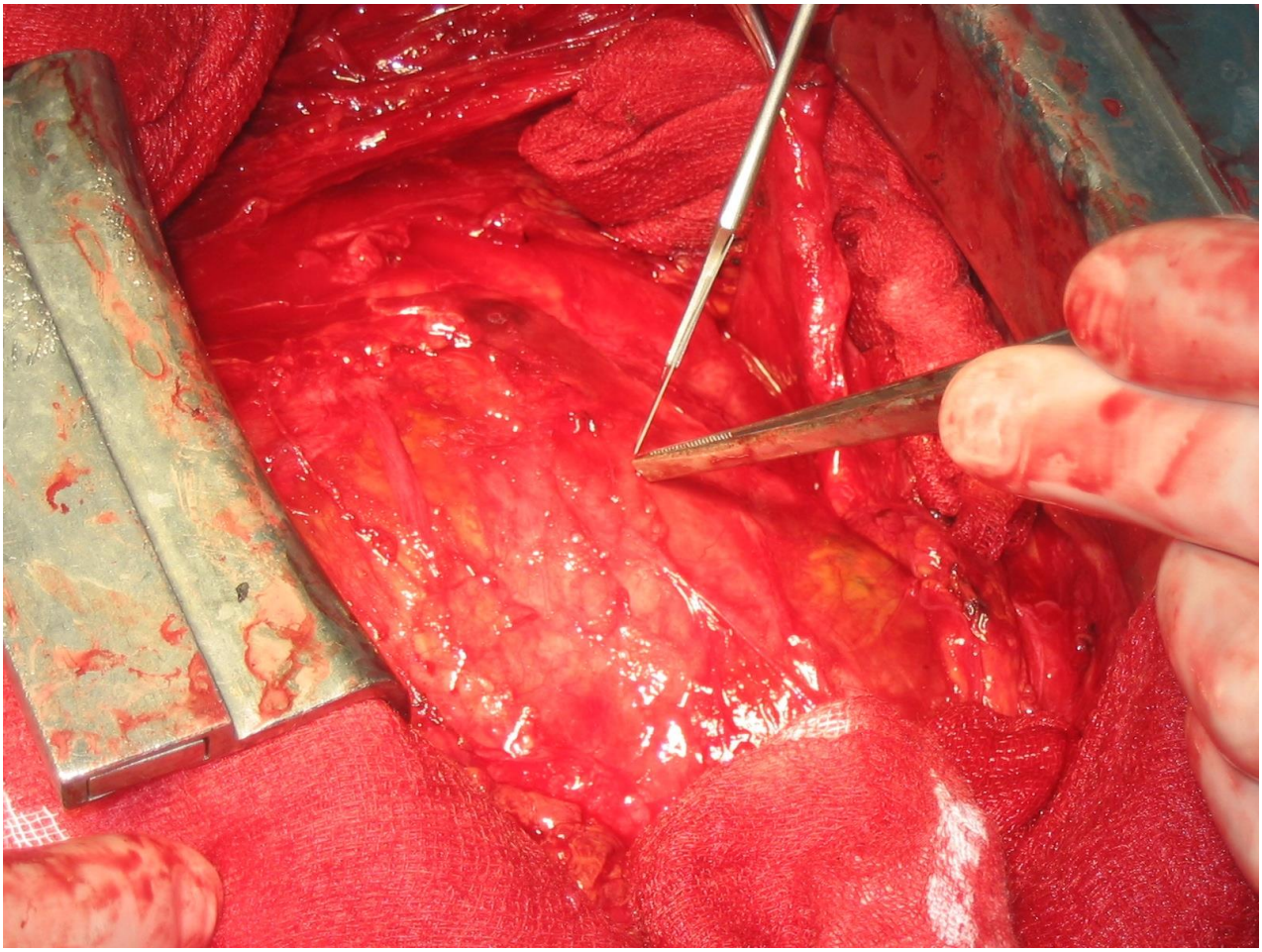

Figure 3. Intraoperative 15x20 mm a rupture in the anteromedial aspect of the ascending aorta.

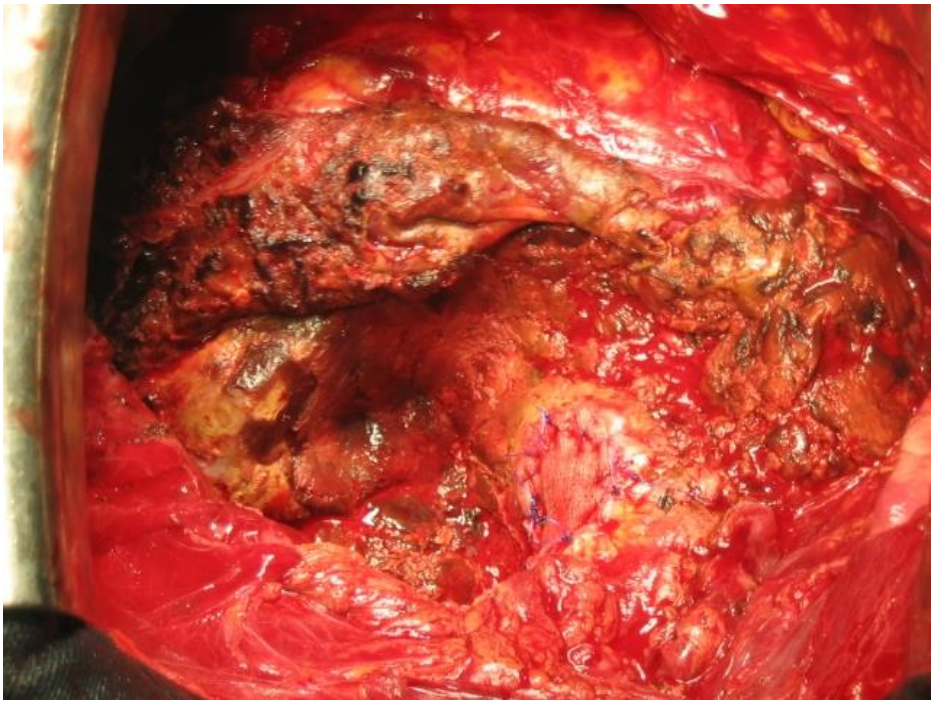

Figure 4. Intraoperatively, the rupture was replaced with a dacron patch.

Discussion

Spontaneous rupture of the aorta can be described as a sudden event not associated with aortic aneurysm, dissection or trauma, inflammation of the aortic wall or erosion from a neoplastic mass [1]. Aortic rupture is a rare seen and that resuscitation in these patients is almost always futile. Even if return of spontaneous circulation is achieved, only few patients survive to discharge from hospital. Clinical experience shows that many patients are diagnosed only post-mortem. Kuisma and Alaspää [2] report on 276 patients with out-of-hospital cardiac arrest of noncardiac origin. Non-traumatic bleeding due to aortic rupture or gastrointestinal hemorrhage was present in 13% of the study population, and 60% of these remained unrecognised out-of-hospitals. In another study, Marston et al. [3] reported that 30% of the patients with ruptured aortic aneurysm were initially misdiagnosed. History taking and diagnostic procedures were difficult and in some cases impossible. In our case, history taking was difficult too. Even in stable patients clinical signs and symptoms of ruptured aortic aneurysm can be misleading, as the leading symptom prior to collapse in thoracic aortic aneurysm is pain, which may be quite unspecific. The symptoms most commonly found were abdominal, back or flank pain, which are compatible with musculo-skeletal

pain, urinary or biliary tract or gastrointestinal disorder. Kefer et al. [4] reported that aortic rupture was the cause of unexpected death after discharge in three of nine patients who were dismissed from the emergency department under the erroneous diagnosis of unspecific back or abdominal pain.

Patients were typically elderly men and had significant co-morbidities, most frequently arterial hypertension. Our case was young man and no arterial hypertension and family history. In about 70% of the cases the index event was the first clinical presentation of aortic disease. Pulmonary embolism and subarachnoid bleeding however are also known to be significantly associated with pulseless electrical activity (PEA) [5,6].

Transthoracic and particularly transesophageal echocardiography allows differential diagnosis between myocardial infarction, cardiac tamponade or pulmonary embolism. In a recent study on patients with suspected cardiac cause of out-of-hospital cardiac arrest, Lederer et al. [7] reported that in 9 of 91 patients ruptured aortic aneurysm or pericardiac tamponade was found by autopsy. Four of them had been subjected to systemic thrombolysis. Chest-X-ray, which is ordered routinely in patients with chest pain, showed mediastinal widening in seven of nine patients. However, it will not be ordered in patients without a spontaneous circulation. CT is known to be both highly sensitive and specific for the identification aortic aneurysms, dissection and rupture. Kvilekval et al. [8] found that the delay imposed by obtaining a preoperative CT scan in patients with possible ruptured aneurysm did not adversely affect patient outcome. Recently, Stanson et al. Reported as a new clinical entity that penetrating atherosclerotic ulcers (PAUs) can be regarded as a cause of aortic rupture by atherosclerosis [8]. PAU of the aorta is ulceration of an atherosclerotic plaque that penetrates the intima, and it results in aortic intramural hematoma, adventitial pseudoaneurysm formation, or aortic rupture [9]. The aortic rupture through a small intramural hematoma with ruptured arteriosclerosis plaque could have been interpreted clinically as a spontaneous aortic rupture. The common clinical picture of this fatal disorder is that of a middle-aged patient presenting with acute chest pain, collapse, and a history of hypertension. A high degree of suspicion for this catastrophic disorder must be maintained for such a patient presenting with acute hemorrhage around the thoracic aorta without any evidence of aortic aneurysm, dissection, penetrating arteriosclerotic ulcer, or intramural hematoma.

Acute aortic syndrome includes aortic dissection, intramural hematoma and symptomatic aortic ulcer. Acute aortic dissection requires a tear in the aortic intima that commonly is preceded by

medial wall degeneration or cystic media necrosis [10]. Blood passes through the tear separating the intima from the media or adventitia, creating a false lumen. Propagation of the dissection can proceed in anterograde or retrograde fashion from the initial tear involving side branches and causing complications such as malperfusion syndromes, tamponade, or aortic valve insufficiency [10]. Both acquired and genetic conditions share a common pathway leading to the breakdown in the integrity of the intima. All mechanisms that weaken the media layers of the aorta will eventually lead to higher wall stress, which can induce aortic dilatation and aneurysm formation, eventually resulting in intramural hemorrhage, aortic dissection, or rupture. The factors culminating in a clinical dissection are quite diverse. The most common risk condition for aortic dissection is hypertension, with chronic exposure of the aorta to high pressures leading to intimal thickening, fibrosis, calcification, and extracellular fatty acid deposition. Furthermore, the extracellular matrix may undergo accelerated degradation, apoptosis, and elastolysis with eventual intimal disruption, most often at the edges of plaques [10].

Marfan's syndrome, vascular Ehlers-Danlos syndrome, annuloaortic ectasia, bicuspid aortic valve, and familial aortic dissection are genetic conditions that often cause acute aortic syndromes. These different genetic disorders are similar pathophysiology that includes a dedifferentiation of vascular smooth muscle cells and enhanced elastolysis of aortic wall components, leading to a compromised intima and aortic dissection [10]. Given this genetic predisposition, a detailed family history in patients diagnosed with acute aortic syndromes or sudden death is particularly important in assessing the need for family screening.

Conclusion

Although our case was young man, this was not associated with aortic aneurysm, dissection or trauma, inflammation of the aortic wall or erosion from a neoplastic mass and normal histopathology. Suspicion of spontaneous aortic rupture and aggressive exploration of the pericardium, mediastinum, or left thorax with appropriate aortic repair seems to provide the only chance for the patient's survival.

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