Ruptured thoracic aortic aneurysm in young male patient: A rare case report

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Abstract
Thoracic aortic aneurysm (TAA) is the ballooning of the upper aspect of the aorta above the diaphragm. We herein present a rare case of a massive ruptured TAA in a young male patient. A 30-year-old male patient diagnosed with massive descending TAA with rupture Crawford Type I showing widening of the superior mediastinum with homogenous round opacity with regular smooth margins observed in left midzone silhouetting the aortic knuckle causing massive hemothorax with the passive collapse of the lung and mediastinal shift to the right. On color doppler it showed typical yin-yang sign. The patient underwent open chest repair with a graft. This case study demonstrated that TAA warrants an early diagnosis because the majority of patients exhibit no symptoms until the aorta ruptures or is dissected, which worsens patient morbidity and mortality. Furthermore, it prompts the necessity of early intervention like either endovascular stent placement or thoracotomy with grafting on the basis of CT angiography findings.

Keywords: TAA, AAA, descending thoracic aorta, CT angiography, chest graft

Introduction
The thoracic aorta comprises of the aortic root, ascending aorta, aortic arch, as well as descending aorta [1]. An aneurysm happens when the artery’s typical diameter rises by 50% [2]. It occurs due to the intrinsic weakness of the aortic wall. About 95% of the patients with Thoracic aortic aneurysms (TAA) are asymptomatic and rarely manifest with symptoms. [3,4] These aneurysms are known as “silent killers” as they may cause fatal complications including aortic dissection or rupture [5].

During an aneurysm complication, approximately 22% of the people will die before reaching the medical facility [6]. Most TAA develop in the ascending or root of the aorta, then in the descending aorta, and rarely in the arch [1]. The most commonly associated risk factors for TAA include male gender, advanced age, history of hypertension, smoking, chronic obstructive pulmonary disease (COPD), coronary artery disease, and past aortic dissection. [7] TAA's having an incidence which is estimated of at least 5–10 per 100,000 person per year, of which the majority of cases being discovered by chance [2]. The average growth rate of abdominal aortic aneurysms (AAAs) of sizes 30 to 55 mm ranges from 0.2-0.3 cm/year. Larger AAAs are linked with higher abdominal aortic aneurysm (AAA) growth rates. TAA’s that are genetically activated exhibit different behaviour from atherosclerotic aneurysms. TAA’s have a 0.1–0.2 cm/year growth rate and exhibit significant individual heterogeneity [2,8].

Aortic root or ascending aortic aneurysms are the most prevalent type of TAA (approximately 60 percent of cases), followed by descending aorta aneurysms (approximately 35 percent) and aortic arch aneurysms (less than ten percent). [2] The growth rate of descending aortic aneurysms is significantly higher (0.19 cm/year) than that of ascending aortic aneurysms (0.07 cm/year). Furthermore, bicuspid aortic valve (BAV) ascending aortic aneurysms grow more rapidly (0.19 cm/year) than tricuspid aortic valve (TAV) aneurysms (0.13 cm/year) [9].

In most individuals, unruptured aneurysms are asymptomatic. They are discovered accidentally during a clinical examination or population screening. TAA’s typical symptoms are usually related to localize mass effect, increasing aortic regurgitation, heart failure brought on by aortic root dilatation, or systemic embolization brought on by mural thrombus or atheroembolism.
Thoracic aortic rupture leads to sudden severe chest or back pain. Rupture into the pleural cavity (usually left) or into the mediastinum is associated with hypotension; rupture into the esophagus leads to hematemesis from an aortoesophageal fistula, and rupture into the bronchus or trachea results in hemoptysis. Infected TAAs are more commonly associated with fistulas. The acute aortic expansion contains rupture, and pseudoaneurysm can cause severe chest or back pain. Thoracic aortic dissection is more common than rupture. A contrast-enhanced Computed Tomography is the most commonly used imaging technique to diagnose TAAs. It is easily available and facilitates quick evaluation of the size, extent, as well as location of the aneurysm. Calcifications, dissections, and mural thrombus can also be clearly visualized. Herein, we aimed to describe and delineate the rare case of massive TAAs with rupture in a young male patient.

Case report
A 30-year-old male patient presented to the emergency department with a history of dry cough, hemoptysis associated with hypertensive (blood pressure 80/60 mmHg in both arms), and post nephrectomy. Based on the patient’s case history, radiological investigations viz. plain chest X-ray, transthoracic Ultrasoundonography, and CT angiography examination were done and diagnosed as massive descending TAA with rupture (Crawford type I; Stanford type B). CT aortogram showed widening of the superior mediastinum with homogenous round opacity with regular smooth margins noted in the left mid zone silhouetting the aortic knuckle causing massive hemorthorax with the passive collapse of the lung and mediastinal shift to the right. On color doppler it showed typical yin-yang sign (Figure 1). The patient underwent an emergency open chest repair with a graft (Figure 2).

3D Ultrasonography image features of aneurysms included fusiform dilatation type with a size of 92.5 x 86.9 x 116.6 mm (width anteroposterior craniocaudal view). Hyperacute crescent shape intraluminal thrombus, which confirms one of the earliest signs and most specific imaging manifestation of the rupture process. The posterior aortic wall was not identifiable and had a draping aorta sign of chronic contained aortic rupture (Figure 3A, 3B, 3C, 3D, 3E, 3F). Rupture of the aortic wall was surrounded by hematoma and massive hemorthorax with the passive collapse of the left lung (Figure 4A, 4B, 4C). 3D reformatted image of the aorta following aortic aneurysm protocol showed neck length angle of 130° with neck length measuring 12 mm, mural thrombus ratio of 0.255, confirmed tortuosity index was 51.121 with outer diameter of right and left iliac arteries measuring 8.5 mm and 9 mm respectively (Figure 5A and 5B).

Discussion
An aneurysm commonly described as the enlargement of a blood vessel to more than 50% of the expected diameter for gender, age, and weight. Though the true TAAs prevalence is unknown, it is estimated to have an incidence of at least 5–10 per 100,000 person-years. Atherosclerosis, along with its risk factors, connective tissue disease (e.g., Marfan syndrome), and abnormalities of the aortic valve (e.g., bicuspid aortic valve) are strongly linked with ascending aortic aneurysms development, but sporadic cases have also been reported.

Since the majority of TAAs are asymptomatic, diagnosing a patient with one is a real difficulty for medical professionals. TAA can be diagnosed only while doing routine radiological examination as an acute presentation with dissection or while screening the relative of a patient with a known aneurysm, or as a part of a known congenital cardiac defect. Some of the reported symptoms of thoracic aortic aneurysm are dysphagia, dyspnea, hoarseness of voice, cough, claudication, cerebrovascular events, as well as chest, abdominal, or back pain. Some of the uncommon presentations includes erosion of the overlying skin and sternum, rupture into the esophagus, compression and invasion of the pulmonary artery, superior vena cava syndrome, as well as recurrent laryngeal and phrenic nerve palsy.

Due to biliary ducts compression and obstruction, jaundice also has been described as a rare presentation of abdominal aortic aneurysm (not TAAs). Failure of right ventricle may precipitate liver congestion, as result hepatic blood flow will be reduced, arterial oxygen saturation will be decreased, and hepatic venous pressure will be increased. Abnormalities of laboratory parameters include elevated serum aminotransferase levels (2 to 3 times of normal reference range) and hyperbilirubinemia. In our case study, the patient was diagnosed as massive descending TAA with rupture (Crawford type I; Stanford type B) based on history and radiological investigations. Masked aneurysms risk rupture and dissection, which carry higher patient morbidity and mortality. Patient clinical history should be focused on the symptoms, family history, risk factors, and detailed physical examination for aneurysmal dilatation signs or its complications need to be evaluated to identify these aneurysms. Aortic imaging with echocardiography, magnetic resonance angiography and computed tomography angiography remain the cornerstone to diagnose aortic aneurysms. Transesophageal echocardiography screening is suitable to assess the aorta among populations at risk (e.g., bicuspid aortic valve, connective tissue disorders, or screening first-degree relatives of patients with TAA). However, screening of the general populace is not advised. The cornerstone of nonsurgical treatment of small, uncomplicated aneurysms involves strict controlling of blood pressure. The important factors for intervention are aneurysm size, expansion rate, and associated comorbidities. When the risk of rupture outweighs the risk of repair, aneurysms are repaired.

Based on the anatomical scope of the aneurysm, TAA is categorized into 5 types. Crawford Type I involves most of the descending thoracic aorta from the origin of the left subclavian to the suprarenal abdominal aorta. Crawford Type II is the most extensive, extending from the subclavian to the aortoiliac bifurcation. Crawford Type III involves the distal thoracic aorta to the aortoiliac bifurcation. Crawford Type IV TAAs are limited to the abdominal aorta below the diaphragm. The group modified this scheme by adding Type V, which extends from the distal thoracic aorta to the diaphragm. Safi’s group modified this scheme by adding Type V, which extends from the distal thoracic aorta to the diaphragm.
creation of true and false lumens. Patients may also exhibit with shortness of breath with a difference in blood pressure between the right as well as left arm, nausea, and hypotension [1]. A widening of the cardiac silhouette could be noticed on a chest X-ray. The intimal flap and the lumens in the aortic wall can be easily seen on a Computed Tomography. Surgery on emergency basis is needed to fix the defect.

Aortic rupture that is contained by the thin layer of adventitia is known as a thoracic aortic pseudoaneurysm. It appears after an injury or atherosclerotic ulcerated plaque erosion. Patients can present with chest pain or deficits due to embolization. This anomaly can be seen with angiography and CT scanning. To prevent aortic rupture, surgical repair is required. Aortic intramural hematoma is a hematoma that develops in the aorta's medial layer. Intimal tears are typically absent. They can occur because of a penetrating atherosclerotic ulcer. Hematoma is also a precursor for aortic dissection, hence surgical repair is necessary to restore the aorta wall’s architecture.

In conclusion Thoracic aortic aneurysm is localized or diffuse dilation of the thoracic aorta involving all layers of the aorta. Aneurysm development is multifactorial in nature with both genetic predisposition and environmental factors activating together to initiate a cascade of arterial wall degeneration. The most common affected is ascending aorta, and the most common cause is atherosclerosis. Aneurysm reaching 7cm in descending thoracic aorta carries a substantial risk of rupture, dissection, and death. This case study prompts the necessity of early intervention like either endovascular stent placement or thoracotomy with grafting on the basis of CT angiography findings.

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**Fig 1:** CT topogram showing widening of the superior mediastinum with homogenous round opacity with regular smooth margins noted in left midzone, massive hemothorax with mediastinal shift to the right.

**Fig 2:** Intraoperative picture demonstrating the repair of the aneurysmal dilatation of the thoracic aorta.
A. 3D VR anterior view showing fusiform dilation of descending thoracic aorta
B. Axial section and
C. Sagittal section demonstrating the large ruptured thoracic aortic aneurysm (92.5 × 86.9 × 116.6 mm)
D. and e. Sign of rupture-hyperacute crescent shape intraluminal thrombus, one of the earliest and most specific imaging manifestations of the rupture process
F. Draped aorta sign, posterior aortic wall not identifiable, sign of chronic contained aortic rupture

Fig 3: CT aortogram
A., B., and C., focal discontinuity of aortic wall (rupture) with surrounding hematoma and massive left hemothorax, hemo mediastinum with the passive collapse of the left lung.

**Fig 4:** CT aortogram

A. Neck angle: 130°

B. Neck length: 12 mm, mural thrombus ratio (thrombus 16.5 / lumen 64.6) =0.255, Tortuosity index (central line 337 mm/straight line 1223) x 100=51.121, outer diameter of right and left iliac arteries are 8.5 mm and 9 mm.

**Fig 5:** 3D reformatted image of the aorta with aortic aneurysm protocol

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