Huge Dissected Ascending Aorta Associated with Pseudo Aneurysm and Aortic Coarctation

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Abstract- We report a unique case of chronic dissection of the ascending aorta complicated with huge and thrombotic pseudoaneurysm in a patient with coarctation of descending aorta. Preoperative investigations such as transesophageal echocardiography (TEE) confirmed the diagnosis of dissection. Intraoperative findings included a 12 cm eccentric bulge of the right lateral side of dilated the ascending aorta filled with the clot and a circular shaped intimal tear communicating with an extended hematoma and dissection of the media layer. The rarity of the report is an association of the chronic dissection with huge pseudoaneurysm and coarctation. The patient underwent staged repair of an aneurysm and coarctation and had an uneventful postoperative recovery period.

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Introduction

Tesler revealed 90% of unrepaired isolated aortic coarctation cases are died up to the age of 50 and showed that aortic coarctation is a serious pathology required surgical treatment (1). The 25% of reason for death from uncorrected aortic coarctation is an acute rupture of the aorta with normal diameter or dilated aorta due to aortic aneurysmal dilatation or dissected dilated aorta (2).

Chronic dissected ascending aorta complicated with huge pseudoaneurysm together with aortic coarctation is a unique occurrence, and surgical decision and treatment is difficult. Aortic coarctation as a congenital aortic disease can cause complications such as myocardial infarction, congestive heart failure, infective endocarditis, aortic aneurysm, but aortic dissection with pseudoaneurysm as a result of present resistant hypertension in adult age is an exceeding rare phenomenon. The dissecting aneurysm with pseudoaneurysm is a serious complication of aortic coarctation and, however, aortic dissection with an aneurysm in patients with coarctation of the aorta is well known and some few case reports are observed in medical literature (3-5), but dissected ascending aorta complicated with pseudoaneurysm and coarctation has not been reported so far.

Case Report

A 45-year-old man was referred to our hospital with reduced consciences. The patient was, pale, sweaty, restless, and agitated and had not retrosternal pain. On clinical examination, a diastolic murmur was heard in the aortic area. He has not been diagnosed with the cardiac disease, reporting only a recent diagnosis of hypertension. His jugular veins were not distended; the blood pressure in his upper extremities was 190/110 mmHg. The carotid and radial pulses existed and were symmetrical, whereas femoral pulses were absent. Chest radiography, performed at the bedside, demonstrated mediastinal widening, and evidence of bilateral rib notching and electrocardiography showed sinus bradycardia with left ventricular hypertrophy. Transthoracic and transesophageal echocardiography (TEE) demonstrated no fluid in the pericardial cavity, dilated ascending and aortic root (approximately 15 cm), advanced aortic regurgitation and typical signs for intimal flap in the ascending aorta and large pseudoaneurysm in dissected flap filled with

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thrombosis, and a coarctation of the descending aorta (Figures 1-5).

Because ascending aorta was very large, and echocardiography revealed intimal flap, acute dissection with a higher risk of rupture, was considered, and the patient's clinical condition deteriorated so dramatically that he was taken immediately to the operating room. After median sternotomy, the pericardium was attached severely to cardiac structures and showed pericarditis and the previous leak of blood from the ruptured aorta to the pericardium. It was carefully dissected and opened. The aneurysmal ascending aorta actually was a pseudoaneurysm with wide mouth extended to right hemithorax.

The right atrial appendage was hidden below pseudoaneurysm. Ascending aorta below subclavian artery was normal size and cannulated for cardiopulmonary bypass. Venous drainage was established with a single "two-stage" venous cannula placed into the right atrium, and cardiopulmonary bypass (CPB) was routinely instituted. An aneurysm was limited to the ascending aorta.

The ascending aorta was cross-clamped and incised transversely. There was a flap in chronic dissection that inferiorly limited to Valsalva sinus with the intact ostium of coronary arteries. The sinuses of Valsalva were abnormal, they were suspended, and the coronary ostia were displaced. The aortic valve was tricuspid, without fusion or, thickness, but aortic root had severe annuloectasia. The aortic valve was excised, and the segment of the ascending aorta containing huge pseudoaneurysm with thrombosis was resected. The patient underwent classic Bentall procedure as described in the literature (6). At the end of the procedure, the patient was easily weaned off CPB and transferred to Intensive Care Unit (ICU) unite.

The upper and lower extremity blood pressure was 230/110 and 70/10 subsequently. Acute renal failure occurred, and the patient became oliguric. The patient transferred to angiography unit for evaluation of coarctation and possibility of nonsurgical intervention for opening of a stenotic segment of coarctation. Angiography revealed complete interruption of upper and lower segments of aorta separated by coarctation (Figures 6,7). Nonsurgical intervention was unsuccessful, and patient scheduled for surgery, 24 hours after the repair of an aneurysm. The patient underwent emergency correction of the aortic coarctation. A left posterolateral thoracotomy was performed out at the fourth intercostals space, and we observed marked dilatation of the left subclavian artery.

Descending aorta and huge left subclavian artery completely released. Partial-clamps were then placed proximally and distally to these arteries and bypassed by Dacron tube (Number, 20). Due to appropriate descending aorta perfusion pressure (70 mmHg), we did not need an atrio-femoral shunt for spinal protection. The completely fused isthmic segment was entirely bypassed and the bypass was performed between the huge left sub claveian artery and the thoracic aorta, with end to side method with a segment of Dacron graft with a continuous 4/0 prolene suture. The patient was tried for extubation in the next morning. The patient was agitated, and his neurologic function was not appropriate for weaning from a ventilator. After thoracotomy and bypass of coarctation segment, blood pressure abruptly reduced and maintaining the normal blood pressure needs low dose inotropic drugs. Urine output increased and the patient became polyuric, and Blood Urea Nitrogen (BUN) and creatinine decreased. The patient was discharged on the 7th postoperative day. On routine follow-up, the patient was in excellent condition. He returned to full-time work. On the 2nd month of follow-up, he was free of complications and two-dimensional echocardiography revealed normal heart contractility, good function of the aortic valvular prosthesis.

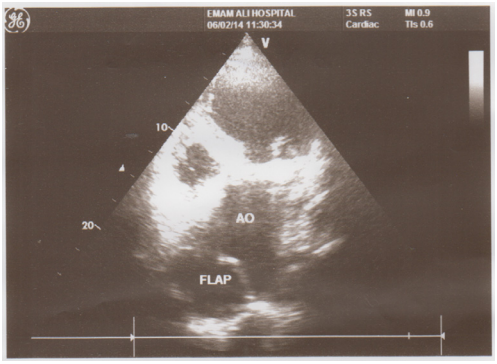

Figure 1. Shows flap in ascending aorta

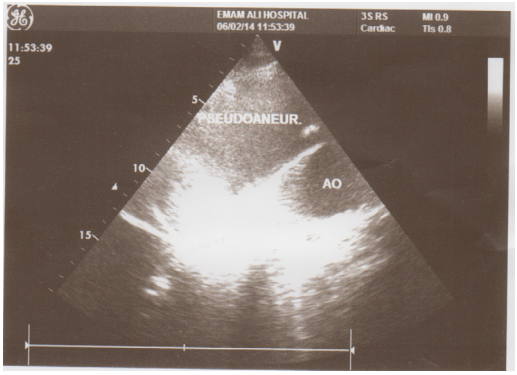

Figure 2. Shows huge pseudoaneurysm of aorta

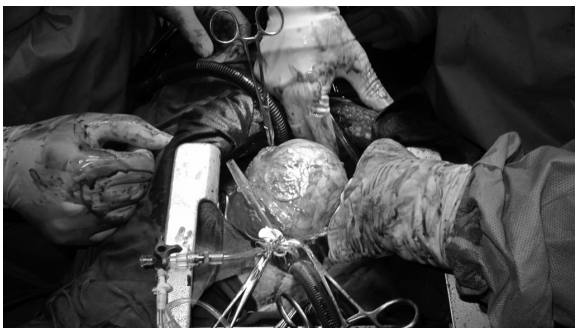

Figure 3. Shows intraoperative view of pseudoaneurysm

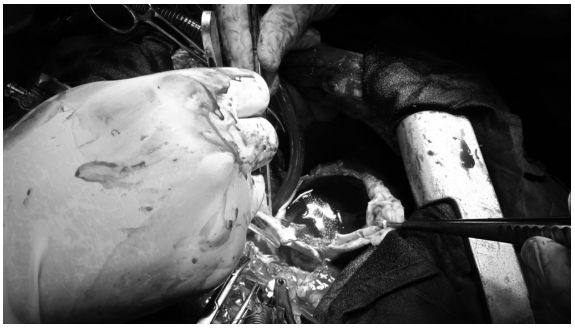

Figure 4. Shows flap in pseudoaneurysm

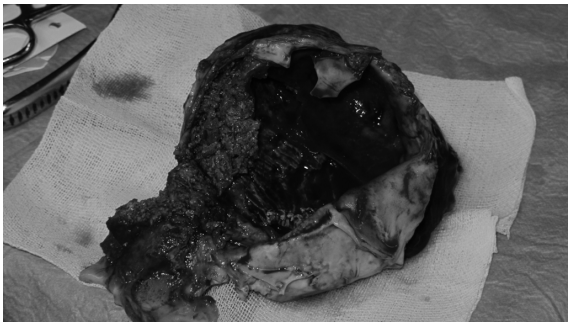

Figure 5. Shows gross pathology of huge pseudoaneurysm filled with clot

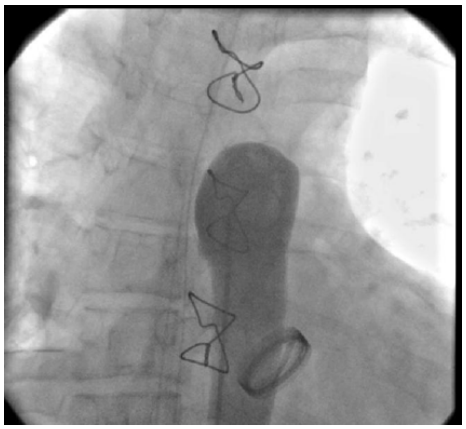

Figure 6. Shows postoperative angiography of descending aorta and coarctation, black arrow shows prosthetic aortic valve of composite graft

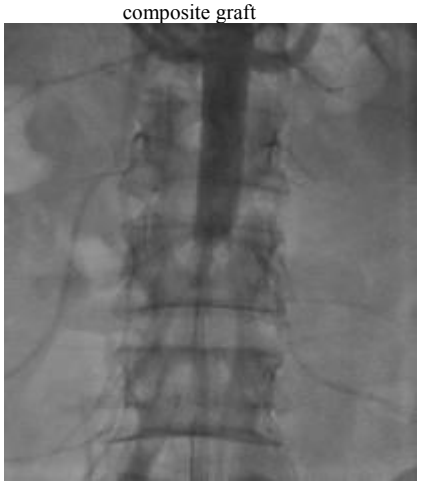

Figure 7. Shows left internal mammary artery as a large collateral artery to distal coarctation

Discussion

Kirsch found that the most common cause of ascending aortic aneurysms and related dissection is idiopathic but genetic and congenital and acquired disease such as connective tissue disorders, coarctation of aorta, bicuspid aortic valve, and pregnancy are major contributor to development of ascending aortic aneurysms as a clinical component of the syndrome (7). Lilly LS exhibits that coarctation of the aorta, also as a congenital disorder, is known to have an increased risk of ascending aortic aneurysm and aortic rupture as well (8). Coarctation of the aorta and bicuspid aortic valve aortic disease have common findings, regarding interstitial connective tissue disease including medial degeneration, increased some enzyme activity, and decreased interstitial fibrillin-1 level in the aortic wall. Lehoux found that coarctation with increasing tensile and shear stresses across the stenotic segment or ascending aorta play a role in the pathogenesis of coarctation aortic disease (9).

Ascending aorta wall tension is increased as hypertension exerts perpendicular wall stress to the aortic wall and is evenly this shear stress distributed along the circumference of the aorta. According to La Place's law tensile stress has a positive correlation with increased aortic radius. Robiscek revealed that hypertension, a product of pre coarctation stenosis , not only exerts force in parallel to the aortic wall by Laplace law but also by stimulation of the endothelial surface, and increased cellular signaling cascades resulting in increased expression of interstitial aortic wall proteolytic enzymes and caused matrix degradation and gradual dilatation of aortic wall (10). keane showed that shear

stress is also exerts increased turbulent blood flow through the lower stenotic segment and causes post stenotic dilatation (11).

In coarctation, lower extremity hypoperfusion needs higher cardiac stroke volumes and hypertension, to compensate perfusion through small collateral vessels leading to higher wall tension in the ascending aorta. The severity of coarctation correlates with severity of hypertension and degree of aortic root dilatation in chronic untreated coarctation .The acute hypertensive crisis may be associated with rupture or dissection of dilated aorta. Dore found that the increased risk of dissection and rupture associated with coarctation is due to the higher prevalence and rate of hypertension, which occurs at a significantly younger age relative to idiopathic ascending aortic aneurysms (12).

In patients with ascending aortic dissection, together with aortic coarctation, initially acute aortic dissection must be repaired, in order to lessen proximal hypertension, decrease the chances of progressive dissection or rupture, but this method does not enable to institute safe perfusion during correction of an aortic aneurysm (3). In our case, both ascending aortic aneurysm and aortic dissection occurred besides aortic coarctation. We could not treat coarctation initially because of acute dissection suspension and rupture risk resulting from ascending aortic aneurysm (almost 15 cm). However, opposed to preoperative diagnosis, the dissection was chronic, for this reason, coarctation pathology could be corrected initially by urgent thoracotomy.

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