



Combination of Dissecting Ascending Aortic Aneurysm and Post-Ductal Coarctation of the Aorta: Case Report from Istishari Arab Hospital

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Abstract

Aortic dissection is a relatively rare but dreadful illness, often presenting with tearing chest pain and acute hemodynamic compromise. Early and accurate diagnosis and treatment are essential for survival. Coarctation of aorta (CoA) can be defined as a cardiac anomaly that results in obstruction of blood flow in the aorta leads to upper extremity hypertension. CT scan and MRI are the most important imaging studies for evaluation of aortic dissection and Coarctation of aorta. In the present review, a case of undiagnosed post-ductal coarctation of aorta presents with ascending aortic aneurysm with early signs of aortic dissection will be discussed. The general clinical manifestations, diagnosis and management of aortic dissection and post-ductal coarctation of will also be reviewed.

INTRODUCTION

The primary event in aortic dissection is a tear in the aortic intima. Data concerning the incidence in the general population are limited; estimates range from 2.6 to 3.5 per 100,000 person-years. The most important predisposing factor for acute aortic dissection is systemic hypertension. Other predisposing factors include disorders of collagen (Marfan syndrome, Ehlers-Danlos syndrome, annul aortic ectasia), bicuspid aortic valve, aortic coarctation, Turner syndrome, coronary artery bypass graft surgery, previous aortic valve replacement, crack cocaine use, strenuous resistance training and trauma(1-4).

Aortic dissection is generally suspected based on a patient's history and physical examination. Patients with an aortic dissection typically present with severe, sharp or 'tearing' back pain (in dissection distal to the left subclavian artery) or anterior chest pain (in ascending aortic dissection). Painless dissection has been reported (5), but is relatively rare. In an analysis of 977 patients from the International Registry of Acute Aortic Dissection (5), only 63 patients (6.4%) had no pain. Patients with painless dissection were slightly older (mean age 67 years versus 62 years) and had a type A dissection more often (75% versus 61%). A history of diabetes, aortic aneurysm or cardiovascular surgery

was more common in patients with painless dissection. Presenting symptoms of syncope, heart failure or stroke were seen more often in this group. In-hospital mortality was significantly higher than for patients presenting with pain (33% versus 23%) (6, 7)

Coarctation of aorta (CoA) can be simply defined as a cardiac anomaly that results in obstruction of blood flow in the aorta, occurring at any site in the thoracic and abdominal aorta(8). The most common site for it is just distal to the origin of left subclavian artery at the connection site between ductus arteriosus and aorta(8). The narrowing of the aorta raises the upper body blood pressure, causing upper extremity hypertension. Unrepaired coarctation leads to premature coronary artery disease, ventricular dysfunction, aortic aneurysm/dissection, and cerebral vascular disease by the third or fourth decade of life(8). Transthoracic echocardiography is mainstay of making initial diagnosis and routine follow-up. Cardiac magnetic resonance imaging (MRI) and computed tomography (CT) are great advanced imaging tools for two dimensional and three-dimensional imaging of aortic arch in evaluating CoA(9, 10). Based on type of coarctation, size of patient, severity of lesion, and associated abnormalities various management options like surgical treatment, transcatheter balloon angioplasty and transcatheter stent implantation are available(11).

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CASE PRESENTATION

A 34 years old male patient with free past medical and surgical histories presented to IAH emergency department on Jan 2023 with a sudden onset complain of diffuse chest pain with radiation to the back. It was associated with dizziness and decreased level of consciousness. His vital signs on presentation showed hypotension around 80\50 with tachycardia. Resuscitation started at emergency department with IV fluid. ECG was done which showed non-specific changes. His labs were within normal limits with no elevation in Troponin. Patient blood pressure improved with reading about 110\60. Chest x-ray showed signs of winded mediastinum. Patient was evaluated by Cardiology team to rule out ischemic heart disease with echocardiography was done and showed severe Aortic Regurgitation with dilatation of ascending aorta. So that, patient was suspected to have aortic aneurysm with signs of aortic dissection. Patient was referred to radiology department where he underwent Aortic CT angiography. Findings were (Figure 1-5):

- **Marked fusiform aneurysmal dilatation of the ascending aorta with peripheral anterior flap suggestive of early dissection.** The diameter is around 6.7 x 6.6 cm at the tubular ascending aorta. (Fig. 1)
- An extensive well developed collateral system is noted in the anterolateral chest wall, thorax, abdomen and in the muscular plane along the back.
- Angiographic examination of the aorta shows a focal area of severe narrowing and constriction seen just distal of origin of left subclavian artery (subtotal luminal occlusion as only a thread-like follow is appreciated) with relative narrowing of descending and abdominal aorta. These findings are associated with collateral pathways have developed through the internal mammary, long thoracic, intercostal, spinal, vertebral and superior epigastric arteries. The above-mentioned findings are suggestive of **post-ductal coarctation of the aorta.** (Fig. 2 + 3)
- Mild to moderate amount of pericardial effusion with HU in NCCT about 40 suggestive of blood with no obvious signs of extravasation. Thickness of effusion is about 1.6 cm ±.
- Inferior ribs notching of the upper ribs.
- Mildly enlarged heart size with intervertebral septum measuring about 2 cm.
- There is reflux of contrast into IVC and hepatic veins associated with dilated IVC and hepatic veins. Findings are suggestive of heart failure.
- Liver is enlarged measuring 19.5 cm in mid-clavicular line associated with nutmeg appearance and peri-portal edema. **These findings are suggestive of congested liver.** (Fig. 4)

- Edematous wall of gallbladder likely secondary to the liver pathology. (Fig. 5)

Impression:

- Ascending aorta aneurysm with peripheral flap suggestive of dissection.
- Post-ductal Coarctation of the aorta with multiple collaterals.
- Mildly enlarged heart with signs of heart pump failure including liver congestion.

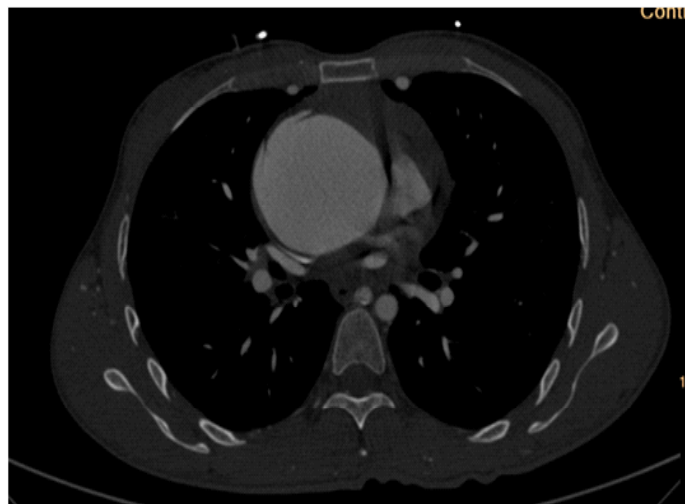


Fig. 1: Ascending aortic aneurysm with signs of intimal flap suggestive of aortic dissection.

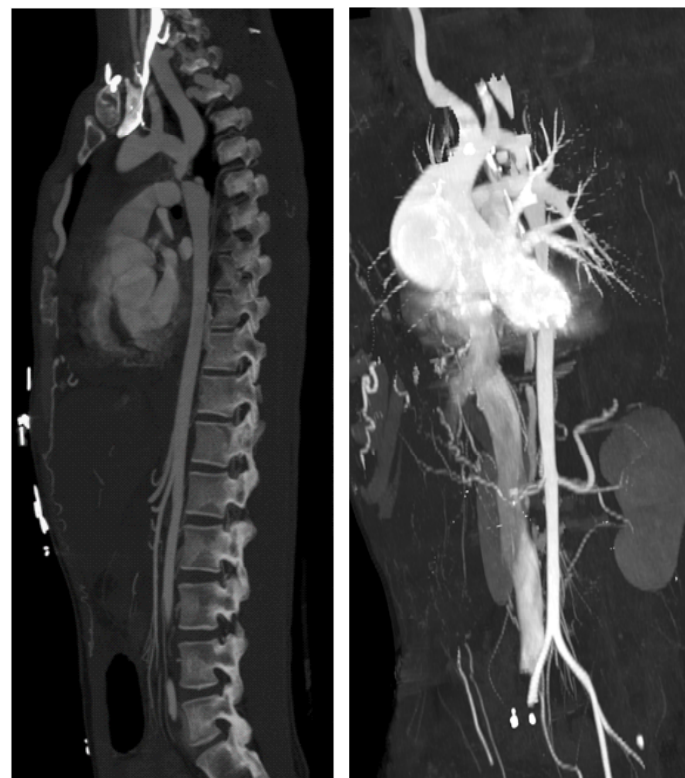


Fig 2: Sagittal view with MIP of aorta that shows post-ductal coarctation of the aorta

Fig 3: 3D view of aorta with post-ductal coarctation of the aorta



Fig 4: Sagittal view with MIP of aorta that shows post-ductal coarctation of the aorta

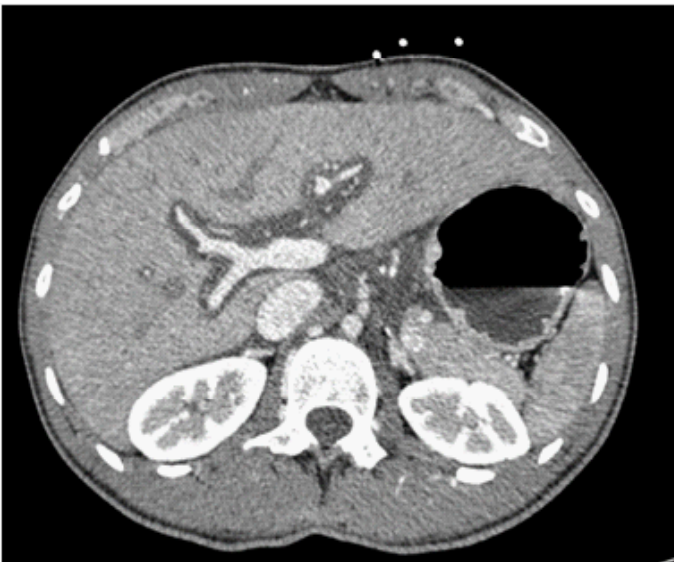


Fig 5 : Nutmeg appearance of liver with peri-portal edema (liver congestion)



Fig 6: Edematous wall of gall bladder.

After that patient was transferred urgently to operation department where he underwent:

- Mid-sternotomy was performed
- The ascending aorta aneurysm was resected
- Aortic valve replacement was done (Bentall Procedure with 23 mechanical Onix Valve and a 28 mm Albograft with Re-implantation of the coronary Arteries)
- A graft was anastomosed between ascending aorta and descending aorta bypassing post-ductal coarctation of the aorta

During surgery due to significant blood loss, patient was given 6 units PRBC, 16 units platelets, 8 units Cryoprecipitate and 6 units fresh frozen plasma.

Patient was transferred to CCU department. He was intubated on mechanical ventilator with high dose inotropes (nor-adrenaline and adrenaline).

Vital signs were HR: 100-110 bpm, BP: 120-125/65-70mm, Hg SPO2: 100%

Patient was intubated for two days. His condition was improved and patient was off inotropes with stable vital signs and normal blood pressure. After two days extubation was done and patient was transferred to ward to continue follow-up.

After few days post-Operative Echo was done and it showed

- LVH with Preserved LV Systolic function, EF: 45 - 50 %
- Metallic Aortic Valve with no Stenosis
- Mild Para-valvular Leak
- No Pericardial Effusion

After few days aortic CT angiography was done which showed (Figures 6-8) aortic valve replacement with shunt that bypassing post-ductal aortic coarctation in addition to normal diameter of ascending aorta with no aneurysm.



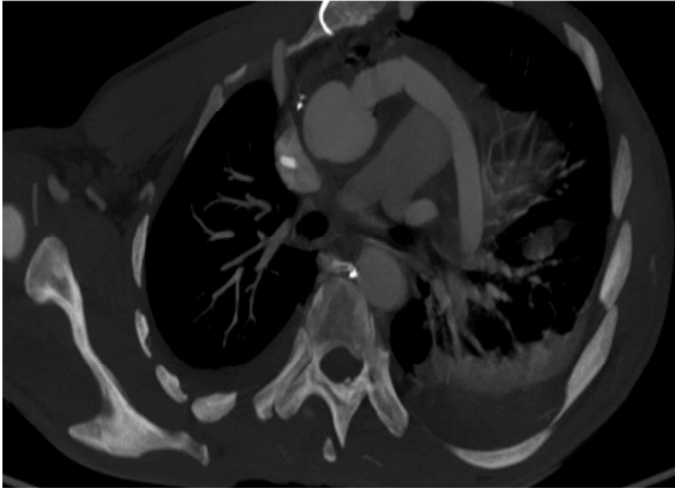


Fig. 7+8: An anastomosis graft between ascending aorta and descending aorta bypassing post-ductal coarctation of the aorta



Fig. 9: Coronal MIP view of ascending aorta shows aortic valve replacement with normal ascending aorta diameter.



Fig.10: Sagittal view with MIP of aorta that shows post-ductal coarctation of the aorta

Patient condition continue to improve and he was conscious oriented alert with stable vital signs (blood pressure was equal in all limbs). Patient was discharged home in good general condition with follow-up at open heart surgery clinic.

DISCUSSION

Due to reduced flow and an atypical aortic wall, coarctation of the aorta is a congenital abnormality usually associated with aneurysm(12). Congenital cardiac abnormalities have an incidence of about 5% for aortic coarctation(13). It is frequently linked to aneurysms, which can occur in the proximal (32%) or distal (51%) regions. Since the 1950s, surgery has been the primary choice for repair. Aneurysms can develop at proximal or distal anastomotic sites even after surgical repair(14). Endovascular operations are a fantastic therapy option, especially in difficult patients with a high risk of morbidity and death. They have opened a new dimension in the management of thoracic aorta diseases(15). In contrast to an isolated dissection or aneurysm, aneurysmal dilatation of a dissected ascending aortic aneurysm (AA) is a rare occurrence. Even less frequently occurs (0.2% to 2%) after replacement of the aortic valve(16). An extremely rare consequence of prosthetic aortic valve endocarditis is the coexistence of dissected ascending AA with various fistulae to the nearby tissues, such as the pulmonary artery and left atrium, as well as the concurrent occurrence of paravalvular leaks(16). According to research, aortic valve replacement is an independent factor in the emergence and development of ascending AAs, which may eventually result in rupture or dissection(17).

The risk of postoperative IE, aortic dilatation, dissection, and paravalvular leaks increases with aortic valve replacement. On the other hand, having a paravalvular leak may increase the risk of developing IE and a fistula in the future(17). The primary surgical procedures that cause the disruption of supporting tissues, such as the release of tissue from the aortopulmonary groove, iatrogenic injury from the needle's tip to the aortic wall, extensive decalcification of the aortic ring, calcification of the aortic wall, perforation of the coronary ostium vault by turbulent flow of the cardioplegia catheter's tip, and disruption of the aortic-mitral fibrous continuity in the primary either an infection of the prosthetic valve and aortotomy suture line, or uncontrolled tension on the aortic wall by the surgeon's assistants (18, 19).

Treatment consist of Following that, the patient was hurriedly taken to the operating room, where he underwent:

- A mid-sternotomy;
- The ascending aorta aneurysm was removed;
- An aortic valve replacement was completed (Bentall Procedure with a 23 mechanical Onix Valve and a 28 mm Albograft with Re-implantation of the coronary Arteries);
- A graft was anastomosed between the ascending and descending .

Patient received 6 units of PRBC, 16 units of platelets, 8 units of cryoprecipitate, and 6 units of fresh

frozen plasma after surgery due to severe blood loss. An echocardiogram was performed a few days after surgery and revealed preserved LV systolic function and an EF of 45 to 50%.

No pericardial effusion; Metallic Aortic Valve without Stenosis; Mild Para-Valvular Leak After a few days, an aortic CT angiography was performed, revealing (Figures 7-9) that the post-ductal aortic coarctation had been bypassed by the replacement of the aortic valve with a shunt in addition to the ascending aorta's normal diameter and the absence of an aneurysm. Patient condition continue to improve and he was conscious oriented alert with stable vital signs (blood pressure was equal in all limbs). Patient was discharged home in good general condition with follow-up at open heart surgery clinic.

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