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GIANT AORTIC ROOT ANEURYSM ASSOCIATED WITH POSTDUCTAL COARCTATION OF AORTA

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All the authors contributed significantly to the research that resulted in the submitted manuscript.

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ABSTRACT

The aorta is a large blood vessel that carries blood from the heart to the rest of the body. The aortic root consists of the aortic valve and the openings for the coronary arteries (the coronary ostia). An aortic aneurysm is an abnormal bulge in the wall of the aorta. If an aneurysm develops in the aortic root, the aorta can dilate and the aortic valve can leak. If the aneurysm continues to expand, it can rupture and can cause life-threatening internal bleeding.

Aneurysms of the aortic root are often due to degenerative disease of media. Aneurysm of the aortic root often affects patients in their second to fourth decades of life. These aneurysms can cause aortic insufficiency, dissection, and/or rupture.Small and slow-growing thoracic aortic aneurysms may not ever rupture, but large, fast-growing aneurysms may rupture.

Depending on the size and growth rate of aortic aneurysm, treatment may vary from watchful waiting to emergency surgery. Ideally, surgery for a thoracic aortic aneurysm can be planned if necessary. Current guidelines recommend surgical treatment when the diameter of the aneurysm exceeds 50 mm. Here we report a case of thoracic aneurysm with postductal coarctation of aorta .

Key Words: Aortic Aneurysm

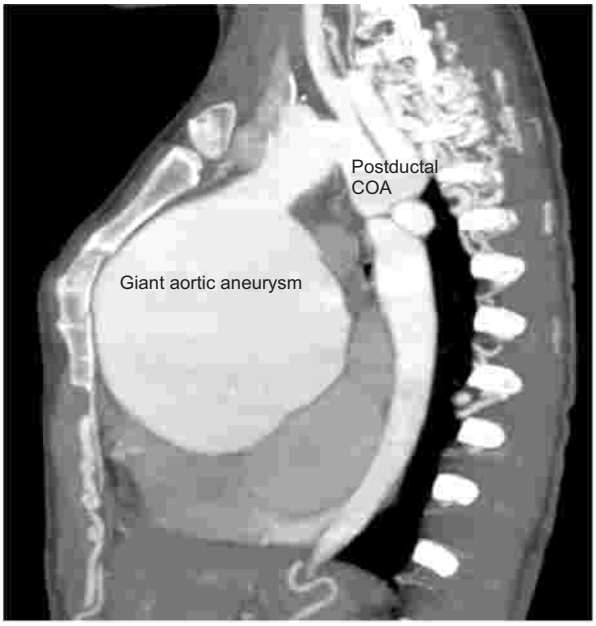
INTRODUCTION

The incidence of aortic aneurysm is 10.4 cases per 100,000 persons per year.[1] Approximately 20 % of thoracic aortic aneurysm is attributed to some form of genetic syndrome.[3] Yet, as in our case, it may also be idiopathic. We report a case of ascending thoracic aneurysm with postductal coarctation of the aorta. Total simultaneous repair was successfully carried out via single stage procedure because of the higher morbidity and mortality rates associated with staged techniques.

CASE PRESENTATION

We present a 22 year old female with a 3 year history of hypertension, dyspnea at rest, 2 pillow-orthopnea, paroxysmal nocturnal dyspnea and headaches. Trans-thoracic echocardiography showed ascending aortic aneurysm extending till

Figure 1: CT scan of Chest Showing Large Aortic Aneurysm and Postductal Coarctation.



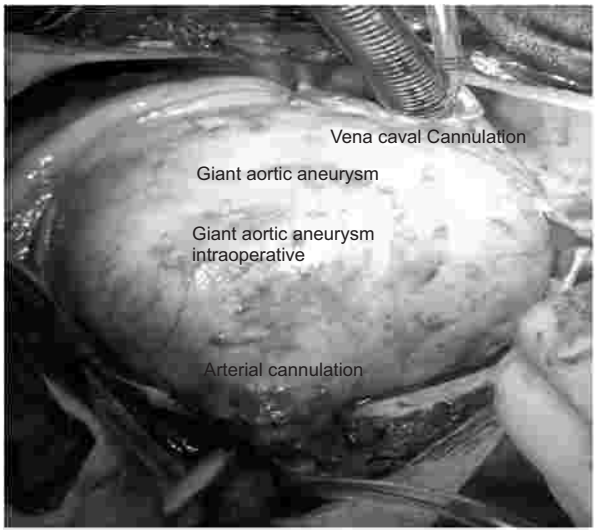
transverse arch, a large aortic valve annulus with severe eccentric aortic regurgitation and mild left ventricular enlargement. The EF was normal 66%. Her CT-chest showed a huge Debakey type II (Stanford type A) ascending aortic aneurysm measuring 11.8 X 10.0 cm associated with severe postductal coarctation of aorta. There were numerous enlarged collaterals and moderate cardiomegaly. She was electively admitted for surgical management. Her vitals were: Pulse 139 beats/min, BP 137/97 mm Hg, Temp 98° F, RR 21 breaths/min, and SpO_2 of 97%. The rest of the examination was unremarkable and lab investigations were normal.

A midline sternotomy observed a 7 cm aneurysm of aortic root. We successfully performed aortic root reconstruction using a composite valved conduit with a bioprosthetic valve of size 23 mm and a Dacron graft of size 26 mm. It was anchored in the aortic annulus using pledgeted mattress sututures with pledgets put in a sub annular position. Extra anatomical graft of 10 mm was proximally anastomosed to the conduit with a retro-caval distal attachment to the descending thoracic aorta, bypassing the coarctation. The left and right coronary arteries were attached to the composite valved conduit. Aneurysmal wall was then sutured over the prosthesis. A section of the aneurysm wall was sent for histopathology which revealed atherosclerotic plaque and fibrosis. The patient had an uneventful postoperative course and was discharged from hospital 10 days after the surgery with no complications.

DISCUSSION

Thoracic aortic aneurysms are divided according to the

Figure 2: Intraoperative Picture of the Aneurysm

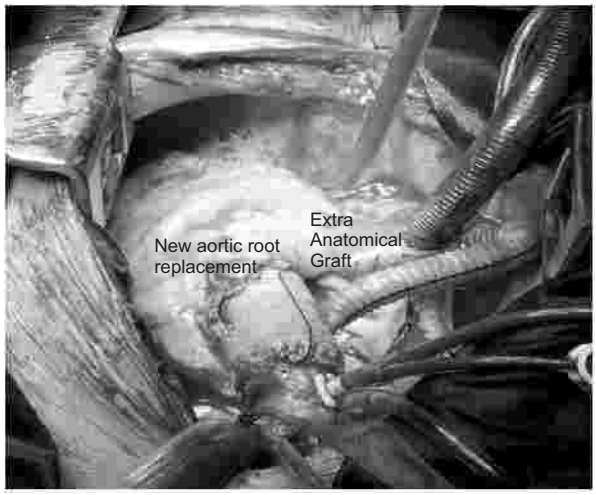


segment of aorta involved: ascending, transverse, descending and thoracoabdominal. The ascending thoracic aorta and/or root are most commonly involved, and the descending aorta least involved. Involvement of aortic arch occurs in only 10 %. The incidence of thoracic aortic aneurysm is only 10.4 cases per 100,000 persons per year.[1] Moreover, its association with coarctation is even rare.

Most thoracic aortic aneurysms are caused by degenerative diseases resulting in dilatation of aorta. Aneurysm formation is attributed to degenerative remodeling of the medial extracellular matrix and Smooth Muscle Cells loss.[2] Risk factors for development of thoracic aortic aneurysm include hypertension, smoking, and COPD. Approximately 20 % of thoracic aortic aneurysms are attributed to some form of genetic syndrome.[3] Yet, as in our case, the pathogenesis of some aneurysms remains unknown. Genetic syndromes with predisposition of thoracic aneurysm include Marfan's syndrome, Ehlers Danlos type IV, Loeys-Dietz syndrome etc. They are also associated with bicuspid aortic valve, coarctation of aorta and other congenital cardiovascular anomalies and inflammatory diseases. The pathogenesis of thoracic aneurysm not associated with known genetic syndromes remains poorly understood. However, alterations in the major constituents of the aortic wall may play an important role in formation and expansion of thoracic and aortic aneurysm. The average rate of expansion of thoracic aortic aneurysm is estimated to be 0.1 to 0.42cm per year.[4,5]

The incidence of aortic dissection and aneurysm in patients with coarctation of aorta is well known, however the reported cases of surgical correction of this condition are very few.[6,7] Fusiform or saccular aneurysms may occur secondary to hemodynamic disturbances caused by the coarctation. If the coarctation is left untreated, 25% of patients die by the end of 2nd decade, 49% die by the end of

Figure 3: Aortic root is Replaced with 26mm Composite Valve Conduit and Extra Anatomical Graft Proximally Anastomosed to Graft



the 3rd decade and approximately 90% die by age 50. The causes of death include congestive heart failure, aortic rupture and dissection, bacterial endocarditis and intracranial hemorrhage.[8]

There are two surgical approaches for patients with coarctation of the aorta and associated aneurysm. One is a single stage simultaneous correction of both lesions through a median sternotomy and the other is a two-stage repair through median sternotomy and lateral thoracotomy. The latter is usually associated with morbidity of two incisions and hemodynamic derangements. Vijaynagar et al were the first to describe aortic valve replacement and concomitant ascending aorta-to-descending aorta bypass through the posterior pericardium with placement of the graft around the left margin of the heart, entirely through median sternotomy.[9] We preferred the single stage, on pump surgical technique via median sternotomy, achieving simultaneous repair of the aneurysm and coarctation. Polytetrafluoroethylene (Dacron) ringed grafts were used for this procedure in an effort to reduce the potential complication of graft kinking and narrowing.[10] For severe AR we chose a composite valved conduit. Although we generally favor the ligation of collaterals or embolization, this particular problem seemed to be better managed with an extra-anatomic bypass, avoiding manipulation of a densely adherent and well-vascularized area, allowing the collaterals to involute. The patient tolerated the single staged repair well and developed no postoperative complications.

CONCLUSION

We conclude that coarctation of the aorta with concomitant cardiac lesions can be repaired simultaneously, through median sternotomy and using extra anatomical graft through posterior pericardial approach minimizing morbidity and mortality.

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