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Case Report

A Rare Long Term Complication of Interrupted Aortic Arch Surgery: Asymptomatic Descending Aortic Aneurysm

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Abstract

A 28 year old male with rare congenital heart disease: Interrupted aortic arch anomaly developed a large descending aortic aneurysm years after multiple surgeries. He underwent successful conduit replacement.

Keyword: *Interrupted Aortic Arch; MRI 3D Reconstruction; Long-Term Complications*

Introduction

Interrupted aortic arch (IAA) is a very rare congenital cardiac anomaly. IAA accounts for 1% of all critically ill neonates with congenital cardiac defects. Celoria and Patton [1] in 1959 classified the defect as type A if the interruption is distal to the origin of the left subclavian artery, type B if between origins of left carotid and left subclavian artery and type C if between origin of two carotid arteries. This interruption can occur in both the left and the right sided aortic arch. DiGeorge syndrome with chromosome 22q11.2 deletion is commonly associated with this anomaly.

Type B are most common representing 53% of all cases, followed by type A in 43% and type C in 4%. Type A interruption tends to occur with aortopulmonary septal defect and inlet ventricular septal defect (VSD) [2]. Type B usually has large malaligned VSD with posterior displacement of the infundibular septum causing subaortic obstruction. Type C interruption is very rare.

The purpose of this presentation is to share our experience of a very rare complication with one of our patients born with IAA who developed a large pseudoaneurysm of the descending thoracic aorta in adult life (Figure 1). Surgical repair was performed.

Typical presentation of this anomaly in neonatal life is ductal dependent left sided obstructive lesions with acute cardiovascular collapse following spontaneous closure of patent ductus arteriosus. They are managed with infusion of prostaglandin E1 to open up the patent ductus arteriosus and medical cardiovascular and metabolic support. Two dimensional echocardiography is the most commonly inpatient tool to confirm the diagnosis.

Type of primary surgery depends upon the size of the sub aortic area [3]. Suggested data indicating the need for surgical therapy to the left ventricular outflow tract are an absolute sub aortic diameter of less than 3.5 mm and a sub aortic area of < 0.7 cm^2/m^2 [3] as well as a diameter of the aortic valve annulus of < 4.5 mm [3].

Surgical options are resection or single ventricular repair by direct end-to-end anastomosis of the proximal and distal arch. Artificial tube graft connecting ascending and descending thoracic aorta is usually avoided when possible because the infant outgrows the size requiring multiple surgeries in the future.

Case Report

A 28 year old male with confirmed DiGeorge syndrome (22q deletion) had undergone neonatal palliation for interrupted aortic arch type B employing 8 mm Gore Tex tube aorto-aortic interposition graft and pulmonary artery banding in 1990 via left thoracotomy.

Two years later (1992) debanding and MPA patch plasty and VSD closure followed. At six years of age (1996), in the second surgery an ascending to descending aortic conduit (12 mm Hemashield tube) was implanted.

He continued to do well, completed college and is currently employed. Physical examination prior to this surgery revealed he is in good health. The Vitals were: Weight. 61 Kg. Height. 172 cm. Heart rate 83/min. Respiratory rate 18/min. Blood pressure: Right arm 123/57 and Right leg 120/67 mm Hg. Cardiac examination revealed quiet precordium, no palpable heave or thrill. Both first and second heart sounds were normal. A grade 2/6 ejection systolic murmur was heard at the left sternal border and a grade 1/4 high pitched early diastolic murmur at the left sternal border, cardiac rhythm was normal.

2D and Doppler echocardiography revealed normal LV wall thickness, size and function. Doppler velocity within the descending thoracic aorta with CW Doppler showed systolic flow velocity 2.1 m/s with mild diastolic runoff. The descending thoracic aorta could not be well imaged.

A routine, standard chest X-ray (Figure 1) showed normal heart size. An opacity typical for descending aortic aneurysm is seen along the left upper cardiac border.

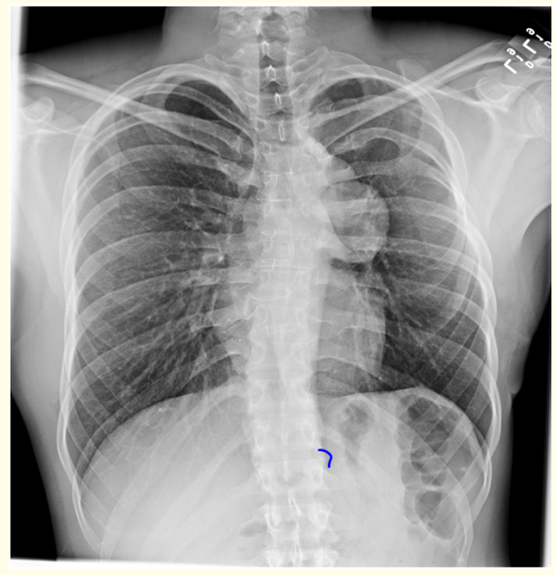

Figure 1: *P-A chest x-ray showing (black arrow) large opacity in left upper (posterior) mediastinum, corresponding with proximal descending aortic aneurysm.*

Cardiac MRI showed a large pseudo aneurysm of the descending thoracic aorta at the anastomosis of the ascending to descending aortic conduit measuring 5 cm X 7 cm (Figure 2).

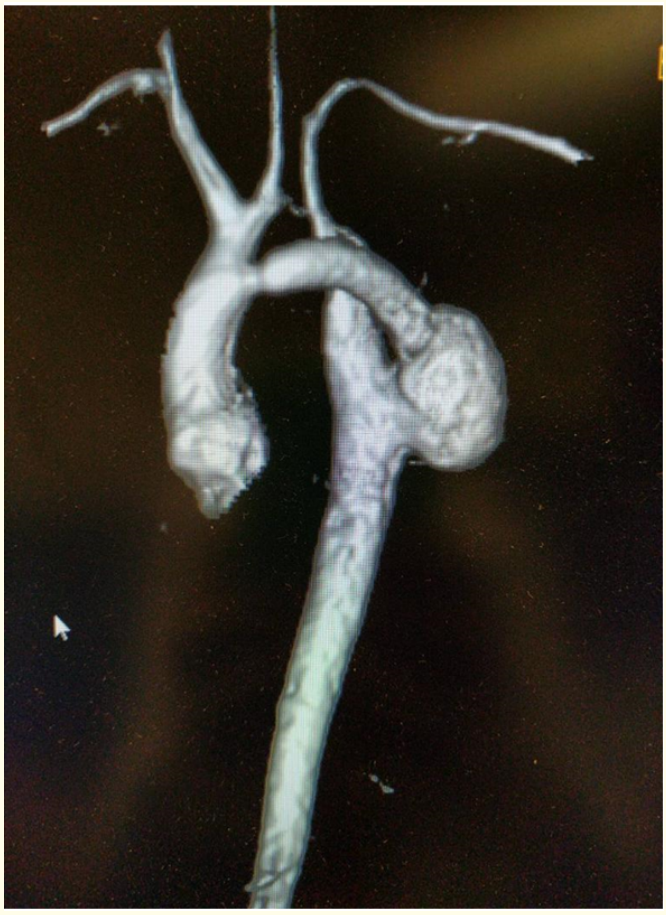

Figure 2: *3-D reconstruction MRI showing 5X7 cm large descending aortic aneurysm, suggestive of almost circumferential dehiscence of distal anastomotic suture line of ascending to descending aortic conduit. Note the tiny patent lumen of original 26 yo 8 mm Gore Tex tube aortic arch interposition graft.*

At surgery through a limited clam shell incision standard left lateral thoracotomy extended anteriorly with oblique sternotomy and anterior right thoracotomy (Figure 3) under normothermic beating heart surgery was performed. On extracorporeal circulation with arterial double cannulation (L femoral artery, ascending aorta) and right atrial two stage cannula stepwise descending aorta and then ascending aorta were side bite clamped. Ascending to descending aortic conduit was completely removed and replaced with new 18 mm Gelweave R Dacron tube (Figure 3a).

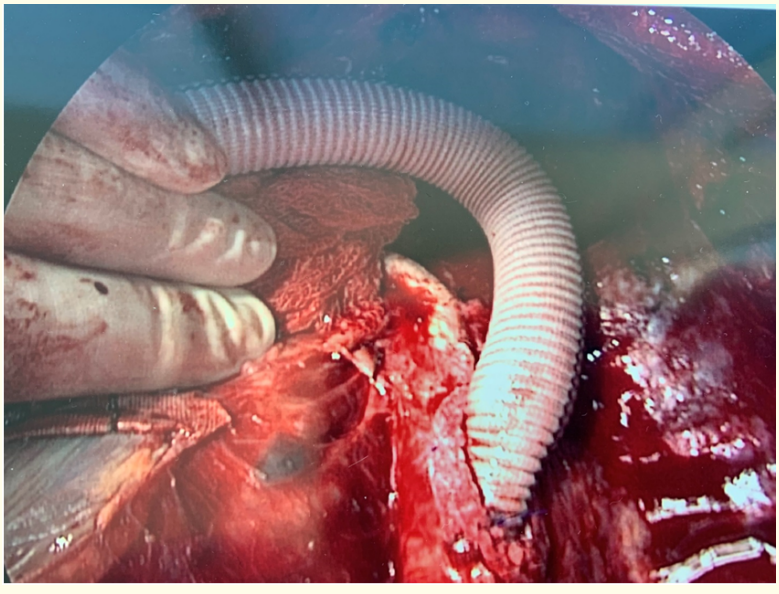

Figure 3a: *End to side anastomosis of 18 mm Dacron graft (Gelweave R) to previous anastomotic site.*

Severely calcified old Gore-Tex interposition graft from twenty six years ago was left in place (Figure 3b).

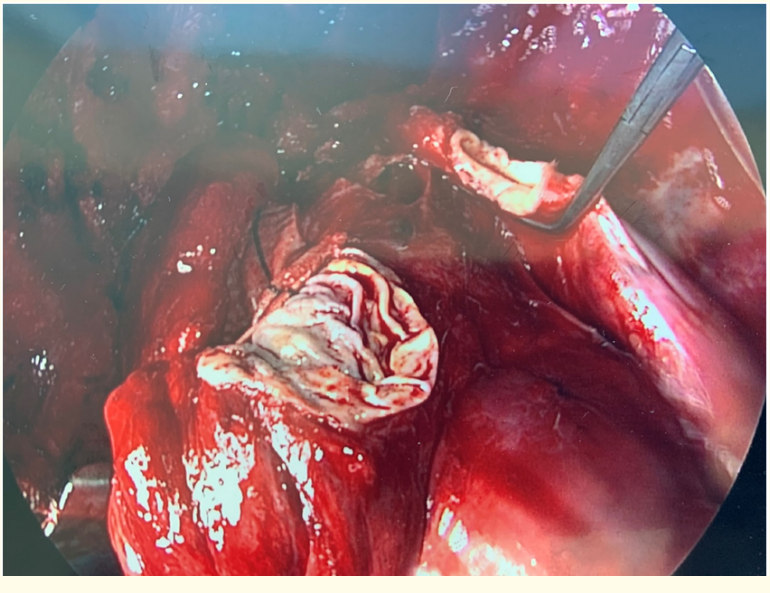

Figure 3b: *During beating heart surgery with arterial double cannulation (L femoral artery, ascending aorta), descending aorta is side bite clamped. In front, crimped false aneurysm, anastomotic dehiscence, contained by visceral pleura. Note dacron graft still attached, as before for maximal 30% of the circumference to the aorta, also.*

The Postoperative course was complicated by significant bleeding form extensive left pleural dissection in combination with coagulopathy. A large hematoma required surgical evacuation through limited re-thoracotomy on postoperative day 4.

Five months post op, patient is doing well and back to his job. In the last clinic visit his BP: Right arm 102/55 mm Hg. and right leg 114/81 mm Hg.

Discussion

While pseudoaneurysm may develop at any anastomotic site between native tissue and artificial tube graft over time, ascending to descending aortic conduits implanted in early childhood needs closer follow up, as tension with growth may give rise to traction on suture lines, increasing the risk of pseudoaneurysm development.

Conclusion

Our experience with this patient suggests the importance of close follow up. Even though 2-Dimentional echocardiogram was done periodically during follow up, 2D echocardiogram is not adequate enough for follow up for this condition. Cross-sectional imaging with cardiac MRI or CTA is recommended for better evaluation of the anatomy of the area of aneurysm. Early recognition and restriction of strenuous physical activities are extremely important to prevent sudden cardiac death.

We are following the patient every six month with Chest X-ray and echocardiogram besides history and physical examination and doing MRI scans every year so far. If any changes on Cheat X-Ray, MRI will be done as needed.

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