



Case report

A ruptured descending thoracic aorta pseudo aneurysm in a child presented with chest pain having untreated aortic coarctation: A case report

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ABSTRACT

Introduction: Coarctation of aorta is a congenital narrowing of the aorta commonly involving periductal segment of descending aorta. It commonly occurs as an isolated lesion and if left untreated it may result in hypertension, aneurysm and aortic rupture.

Case presentation: A previously healthy 10 years old male child presented with sudden onset of chest pain associated with shortness breath. Contrast enhanced computed tomography of the chest revealed a narrowed segment of proximal descending aorta with vascular enhancing out pouching surrounded by massive non enhancing posterior mediastinal collection. Resection and interposition grafting with synthetic tube was done for ruptured pseudo aneurysm complicating coarctation.

Discussion: Coarctation of aorta incidence is two to five times higher in males and its incidence increases with congenital cardiac lesions associated with decreased blood flow through the aorta. Clinical presentation varies with age at presentation so related with severity of the disease. Diagnosis can be made with echocardiography and computed tomography. Options of management and materials to be used are controversial issues but also determined by age and mode of presentation.

Conclusion: Aortic pseudo aneurysm is a rare complication of coarctation which may rupture requiring urgent surgical intervention.

1. Introduction

Coarctation of the aorta (CoA) is a hemodynamic ally significant congenital narrowing of the aorta. It accounts for more than 5 % congenital heart diseases with a prevalence that exceeds 40 per 100,000 live births [3]. Even though any segment of aorta can be involved, the commonest site is just proximal to the attachment of patent ductus arteriosus/ligamentum arteriosum to the descending aorta (1–3).

About 82 % of Coarctation occurs as an isolated lesion, with 11 % having a coexisting Ventricular septal defect, while 7 % have other complex cardiac anomalies [2]. If left untreated Coarctation of aorta might result in hypertension, ascending aortic aneurysm, cardiac failure, endarteritis, and descending aortic aneurysms. The aortic pseudo aneurysm usually occurs in patients with endarteritis [4]. Enlarged third and fourth intercostal arteries at the aortic origin may also result in saccular aneurysms, but not before the age of 10 years [2]. We report a

10 years old boy with restrictive perimembranous ventricular septal defect (VSD) coexisting with uncorrected aortic Coarctation complicated by a contained ruptured descending aortic pseudo aneurysm. To our knowledge, this is the first reported case in a pediatric patient having non mycotic ruptured pseudo aneurysm contained by mediastinal structures complicating aortic Coarctation. patient was managed by resection and Dacron interposition grafting at a public teaching hospital in a low income country. This case report has been reported in line with SCARE criteria [5].

2. Case presentation

A 10 year old boy presented to the emergency department with shortness of breath and severe sudden left side chest pain of 4 days duration which he felt initially while he was playing with his peer. Associated with these he had also difficulty of swallowing solid food,

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palpitation and diaphoresis. Other than few visits to clinics for upper respiratory tract infection he was well thriving child, has no limitation in physical exercise and had no family or personal history of hypertension.

On physical examination he was in severe respiratory distress; with heart rate 122beats/min, respiratory rate 34breaths/min and temperature 36.4°C . His blood pressure was on the upper limbs 128/85 mmHg which was above 99th percentile, which was stage two and on the lower limb 76/50mmhg. He had weakly palpable pulse of femoral arteries bilaterally, radio femoral delay and grade 4 holosystolic murmur best heard at the left lower sternal boarder.

On investigation his hemoglobin was 12.8 mg/dl, platelet count 253,000/ μl and white blood cell count was 16,300/ μl . His serum electrolyte and renal function tests were within normal range. On transthoracic echocardiography assessment there was 7 mm perimembranous ventricular septal defect which was restricted by tricuspid valve septal leaflet with flow peak systolic gradient of 98 mmHg and left ventricle hypertrophy with ejection fraction of 69 %. There was turbulent flow across the descending aorta having a peak systolic gradient of 77 mmHg with transverse arch measured 11 mm, isthmus 9 mm and the narrowest segment of descending aorta measuring 3 mm. Computed tomographic angiograph revealed narrowing of proximal descending aorta just below the origin of left subclavian artery (Fig. 1). The bilateral subclavian arteries, bilateral internal mammary arteries and posterior intercostal arteries were seen dilated. There was also a vascular type enhancing out pouching from right side of the descending aorta having a length of 2.1 cm, largest diameter of 1.2 cm and a neck communicating with aorta of 5 mm (Fig. 2). There was non-enhancing posterior mediastinal collection compressing the left atrium and esophagus with extension to the bilateral pleural cavities and to the retro-peritoneum (Fig. 3 & Fig. 4). With these findings Coarctation of aorta with ruptured contained pseudo aneurysm was made.

In the emergency room the patient was treated with propranolol for the hypertension and analgesics for his pain. We had short discussion with our cardiac surgery team, patient's family was informed about the disease and written consent was then taken for surgery.

After the patient was taken to the operation theatre general anesthesia was induced with single lumen endotracheal intubation. Central venous line catheter was inserted through right internal jugular vein and right radial and right dorsalis pedis arterial line was also secured. The

systolic pressure difference between radial and dorsalis pedis arterial line was more than 50mmhg before starting surgery.

The left groin was cleaned and with small incision femoral vessels were exposed. Then, the left common femoral artery and vein were cannulated and appropriate connections were made to the pump oxygenator to establish partial cardiopulmonary bypass during aortic clamping (Fig. 5). The patient was then positioned in right lateral position and secured with strapping across the hip and onto the operating table. Skin was cleaned with antiseptic solution and draping made. Approach was made through a left posterolateral thoracotomy incision. Intercostal muscles were divided over the upper border of the fifth rib and parts of latissimus dorsi and trapezius muscles were divided for maximum exposure.

Rib spreader was inserted and opened step by step, upon entering into chest cavity there was about 200 ml of blood within the pleural space (Fig. 6A). There was a retropleural clotted hematoma extending from below the transverse aorta to the diaphragm (Fig. 6B). The lung was retracted anteriorly, and the mediastinal pleura was opened carefully over the aorta, then stay sutures were placed to the pleural edges for retraction. The Coarctation site was visible externally having a waist below the origin of left subclavian artery and just below the ligamentum arteriosum (Fig. 7A). There was no visible pulsation of the descending aorta below the site of constriction. On the right and partly anterior side of descending thoracic aorta there was an organized dark clot with no clearly visible aneurysm sac. A careful dissection was made around the distal aortic arch by keeping close to the aortic adventitia and vascular tape passed around it making sure aortic clamp can be applied easily when needed. The same step was done for descending thoracic aorta some distance below the Coarctation. The upper two intercostal arteries were ligated and divided and aorta beyond the Coarctation dissected. The ligamentum arteriosum was ligated and divided.

The aortic cross clamp was then applied both proximal and distal to the Coarctation site. There was back bleeding into the field and after ligating and dividing one medial branch the bleeding stopped. The Coarctation segment including the ruptured aneurysm sac was completely resected. The pseudo aneurysm neck was visible on the resected segment (Fig. 7B). The sac of the pseudo aneurysm had already ruptured and it was not possible to define the peripheral extent of its wall. The clotted hematoma was evacuated mostly and there were no

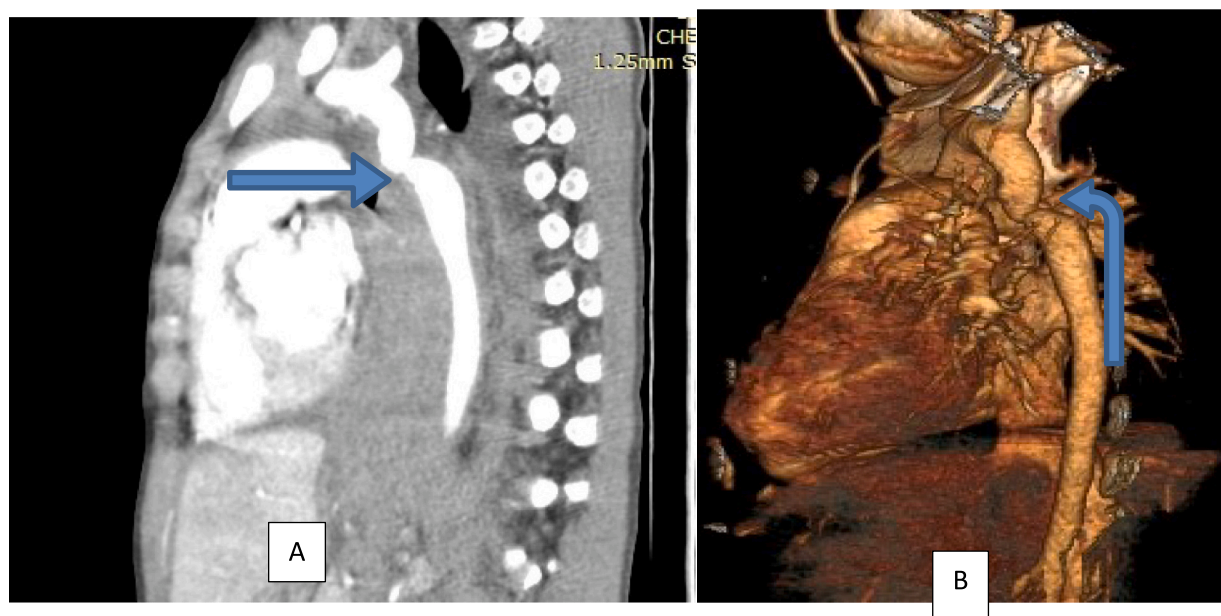


Fig. 1. A sagittal section CT scan image showing narrowing of proximal descending aorta (arrow) just below left subclavian artery origin (A). Coarctation of descending aorta (arrow), a 3-D CT image (B).

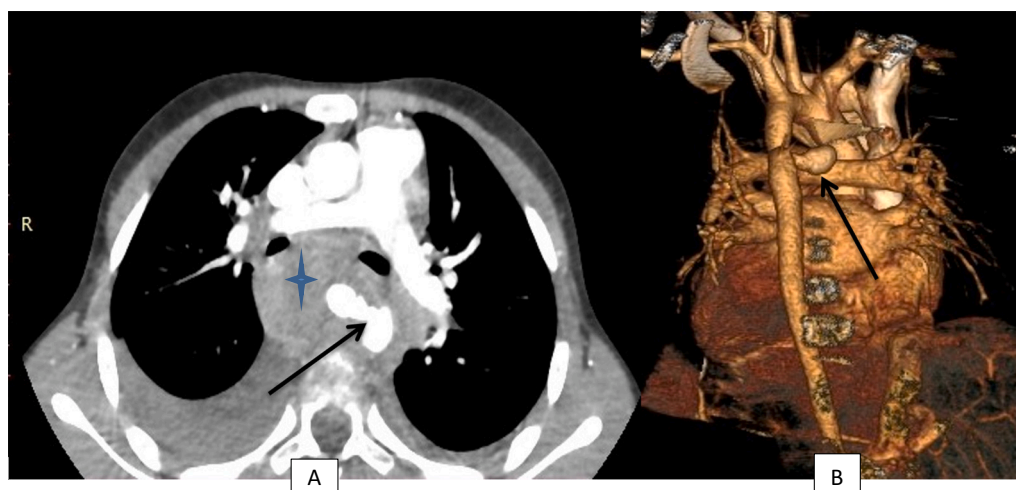


Fig. 2. A vascular enhancing outpouching from right side of descending aorta communicating through narrow neck (arrow), non-enhancing collection around it extending to pleural space bilaterally (star) (A). A 3- D reconstructed CT image showing a saccular outpouching (arrow) from descending aorta (B).



Fig. 3. Axial section CT scan shows non enhancing posterior mediastinal collection compressing the left atrium (arrow), extending also to the pleural spaces bilaterally more on the right side.

signs of infection or abscess collection around the surgical field. The proximal and distal anastomoses were performed to the interpositioned prosthetic 16 mm Dacron tube. The cross clamps were removed, bleeding points were reinforced with additional sutures. The pulsation of descending aorta was clearly visible distally and there was no pressure difference between the upper and lower limb arterial line pressure.

The chest cavity was irrigated with warm saline, a drain was placed in the left thoracic cavity and chest closed in layers. The femoral canulas were removed and inguinal wound sutured. After the patient was repositioned into supine, right thoracostomy tube was inserted for right hemothorax which was significant. The total operation time was 4 h, aortic cross clamp time of 67 min and total blood loss of 500 ml. The patient was transfused with 2 units of packed red blood cells.

Histopathologic examination report of the excised specimen showed tubular tissue composed of mainly fibrous wall with extensive areas of cystic medial degeneration with some infiltration with lymphocytes and neutrophils.

The patient's postoperative course was uneventful and chest tube drains were removed on the 5th post-operative days. Hypertension was controlled with sodium nitroprusside and labetalol, patient was discharged from the hospital on 8th post-operative days. He was then reevaluated on 15th post-operative days and he had no subjective complaint. On Doppler echocardiography there was good flow with gradient of less than 20mmhg (Fig. 8A), on chest x-ray lungs were



Fig. 4. Coronal section CT image showing posterior mediastinal collection extending to the retroperitoneal space surrounding the pseudo aneurysm sac (double arrow).

expanded well with no pleural collection (Fig. 8B). Improvements were also observed in the blood pressure difference of upper and lower limbs, with ankle brachial index (ABI) of 1.02 on the left and 1.14 on the right.

3. Discussion

The term Coarctation is derived from the Latin word *coarctare*, which means to compress [3]. In a localized stenosis hemodynamically important pressure gradient is caused when the aortic lumen is reduced by more than 50 %. Coarctation with or without patent ductus arteriosus but without other major associated cardiac anomalies is termed isolated Coarctation [2]. Morgagni in 1760 did the first description of an aortic Coarctation found at autopsy examination. Crafoord and Nylin in October 1944 did the first operation for aortic Coarctation on a patient. They did resection of the aorta having the most constricted segment and anastomosis of the aorta having comparable lumen (2,6).

Coarctation of aorta incidence is two to five times higher in males. It may be associated with genetic anomalies like gonadal dysgenesis

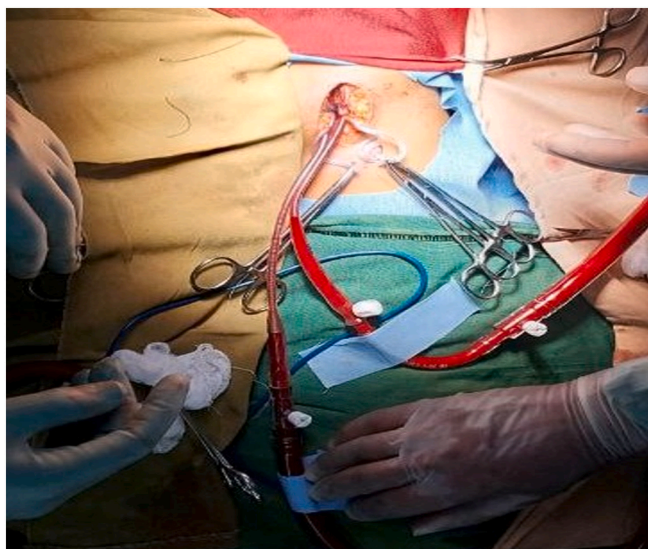


Fig. 5. Femoral vessels cannulated and connected to pump oxygenator.

(Turner syndrome) and Noonan syndrome. A bicuspid aortic valve occurs in about 50 % of cases with Coarctation and these patients have greater risk of aortic root dilation and dissection than those with Coarctation and tricuspid aortic valves (3,7). Generally, cardiac anomalies associated with a reduced aortic flow, such as aortic stenosis, ventricular septal defect (VSD), and mitral valve stenosis or regurgitation have higher incidence of aortic Coarctation. In our patient, Coarctation was associated with a restrictive perimembranous type of ventricular septal defect (VSD). In lesions that result in increased aortic flow, such as pulmonary stenosis or atresia, tetralogy of Fallot and tricuspid atresia with concordant ventriculoarterial connection incidence of Coarctation is close to 0 per 100,000 [2].

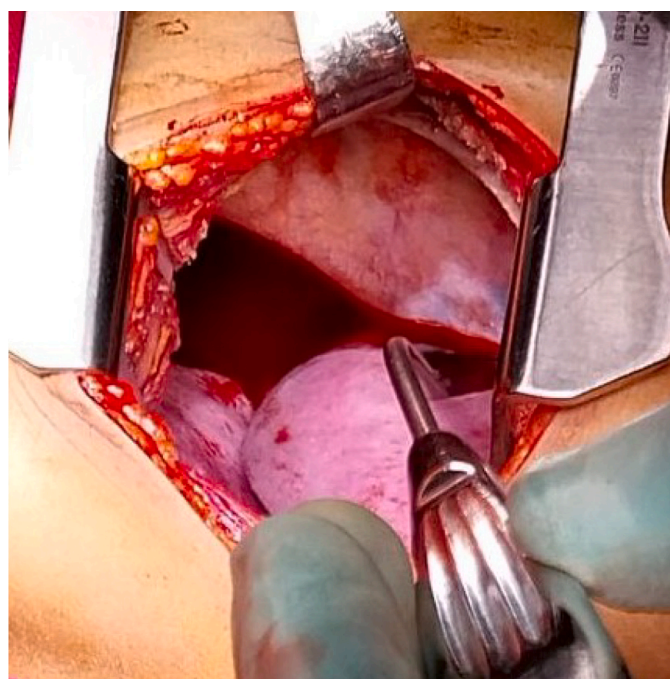
Clinical presentation of aortic Coarctation is determined by severity

of the disease, so mode of presentation depends on age of the patient. In the neonatal period signs and symptoms of heart failure like poor feeding, irritability, tachypnea, and weak or absent femoral pulses and differential oxygen saturations between upper and lower extremities may be there. Many older children and adults who are not diagnosed during infancy as a result of less severe disease may remain asymptomatic and are diagnosed at a routine clinical examination. In symptomatic cases exercise intolerance, lower limb claudication, headache, epistaxis, visual field disturbances, and stroke may be a chief complaint (2,3). In our case the chief complaint of the patient was sudden severe chest pain associated with dyspnea in the absence of diminished exercise tolerance.

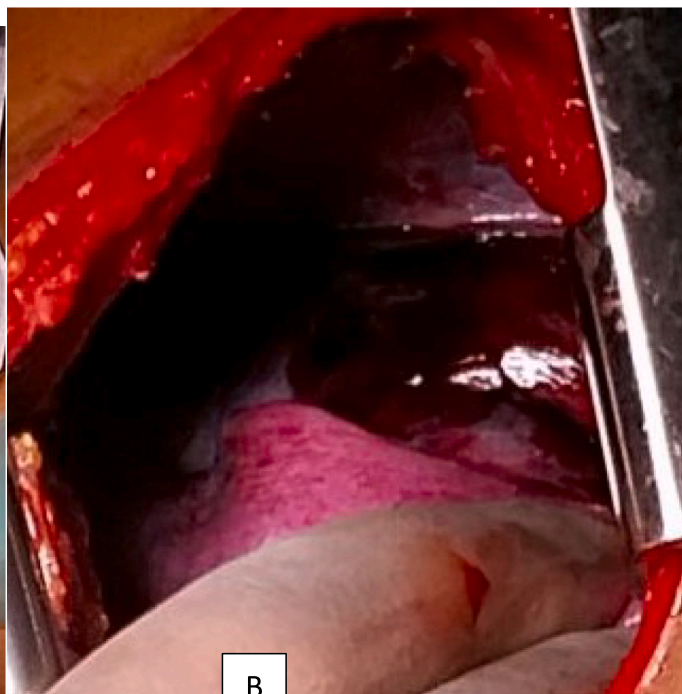
The natural history of patients having Coarctation of aorta is generally poor. The average survival age of patients with untreated Coarctation beyond childhood is 34 years, with 75 % mortality by the age of 43 years. About 76 % of deaths are attributable to complications of Coarctation including heart failure, aortic dissection or rupture, infective endocarditis, and intracranial hemorrhage due to ruptured cerebral aneurysm (1,2).

Aneurysms complicating Coarctation may be single or multiple and may occur in the aorta, intercostal arteries, Abbott's artery, or the cerebral vessels [8]. The anatomic distribution of aneurysms is: 32 % proximal to the Coarctation, 51 % distal, and 17 % involve the left subclavian artery [9]. Aortic rupture is the major cause of death in untreated Coarctation complicated with aneurysm [4]. Apart from the risk of rupture where patient may die suddenly or develop a cerebrovascular accident, pressure on the anterior spinal artery or the spinal cord from an intercostal artery aneurysm can also cause paraparesis [8]. In our case, according to preoperative contrast enhanced computed tomography there was post stenotic pseudo aneurysm medially having a narrow neck communicating with aorta, surrounded by low attenuating posterior mediastinal collection with compressive effect on the left atrium which is consistent with a contained ruptured pseudo aneurysm.

The presence of abnormalities of many arteries suggests that, a Coarctation is probably a feature of a more generalized arterial disease



A



B

Fig. 6. Shows free blood within the pleural space (A), and contained retropleura clotted hematoma (B).

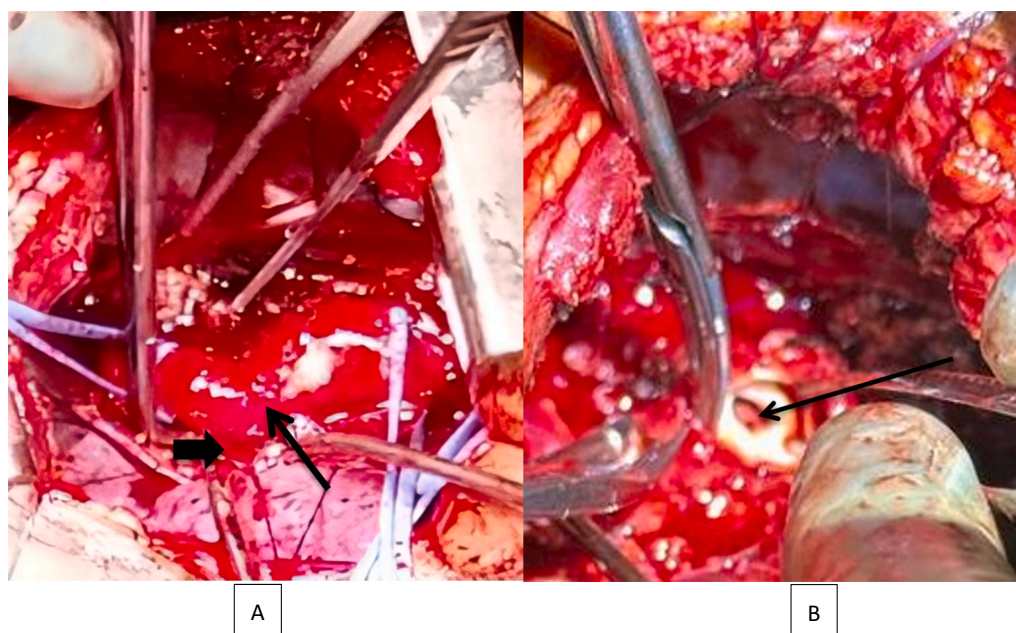


Fig. 7. (A), shows externally visible coarctation site having waist (slender arrow) just distal to ligamentum arteriosum (wider arrow), (B) shows the neck of pseudo aneurysm communicating with the aorta (arrow).

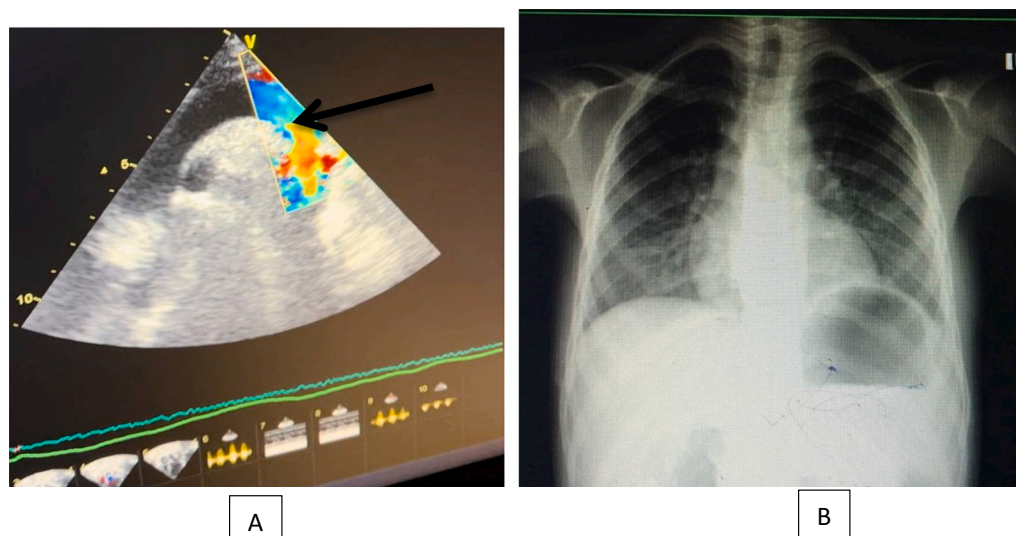


Fig. 8. (A) Doppler echocardiography showing flow through the replaced descending aorta (arrow). (B) PA chest x-ray on 15th post-operative days, shows well expanded lungs with pleural space collection.

as it is also frequently associated with bicuspid aortic valve [8]. The causes of aortic pseudo aneurysm are unclear. But, incriminated factors are hypertension in the upper part of the body, infection, congenital weakness in the aortic wall, and jet stream through the Coarctation. The presence of Coarctation leads to endothelial injury with subsequent infection by the same pathogenic mechanism as endocarditis of the heart valves. The infective lesion happens in the aorta downstream from the high-gradient stenosis at the site of impact of the high velocity jet [4]. In our patient pseudo aneurysm is located at post stenotic site, so hypertension would not be the cause and there was no evidence of endarteritis.

A hemodynamic change in Coarctation as blood flows from a high pressure to a low pressure through a narrow orifice has a jet stream resulting in abnormal stress on the aortic wall and endothelial injury (4, 8). The third and fourth intercostal arteries acting as outflow vessels

from the collateral system to the descending thoracic aorta may also develop saccular aneurysm at the site of their aortic origin as it is the weakest point. But, this phenomenon is rare before the age of 10 years [2]. In our case the pseudo aneurysm is located on the anteromedial surface of the aorta, so saccular aneurysm from intercostal artery origin would not be the cause. Probably, the inherent arterial wall weakness in Coarctation and post stenotic jet stream might have caused the aortic pseudo aneurysm which was ruptured and luckily contained by mediastinal structures.

The diagnosis of aortic pseudo aneurysm can be made by echocardiography and computed tomographic angiography. But, many aortic pseudo aneurysms are discovered when rupture or external compression occurs (4,10). In our case, the patient complained severe sudden chest pain and shortness of breath as a result of ruptured pseudo aneurysm. Transthoracic echocardiography revealed ventricular septal defect,

aortic Coarctation with gradient and ill-defined echogenic mass compressing the left atrium. Then, contrast enhanced computed tomography was done which revealed Coarctation of aorta with contained ruptured post stenotic pseudo aneurysm.

There is a controversy on surgical treatment, timing of the operative intervention, and the type of material used for the aortic reconstruction. Generally, surgical correction is indicated for all symptomatic patients and in asymptomatic patients with upper-to-lower extremity mean blood pressure gradient more than 20 mmHg [3]. In situ reconstruction of the aorta with homograft material after excising the entire infected tissue might be the preferable surgical treatment for mycotic aneurysm located at the site of aortic Coarctation. Another option is excision of the infected aortic tissue and end-to-end anastomosis. The interposition of a synthetic graft between the origin of left subclavian artery and the descending aorta below the pseudo aneurysm is a technique of choice with excellent outcome (4,10). Endovascular approach, such as percutaneous balloon dilatation and stent implantation can be done as well safely in adult patients (11,9).

In our patient consultation for urgent surgical intervention was made to cardiothoracic surgery unit by a pediatric cardiology unit after pre-operative workup. After proper preparation resection of the aorta having narrowing and pseud aneurysm was done with Dacron tube interposition. The patient may require revision surgery in the future as the synthetic tube will not grow with the child. Since the ventricular septal defect was restrictive one its closure was not attempted. We decided to follow and manage after the patient is fully recovered.

4. Conclusion

The aortic pseudo aneurysm is a relatively rare complication in untreated Coarctation which may result in rupture. Urgent surgical intervention is required in pseudo aneurysm complicating Coarctation. Rupture should be considered in patients with untreated aortic Coarctation who present with sudden chest pain.

Consent

Written informed consent was obtained from the patient's family for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Ethical approval

Ethical approval letter RC/DOS/97/23 was given on 28 July 2023.

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Author contribution

WTD did the pre-operative patient evaluation, assisted the operation, conceptualization, writing, editing and submission of the manuscript for publication.

ABT did the operation, conceptualization, reviewing and editing of the manuscript.

HGA did the pre-operative and post-operative patient management, reviewed the manuscript and collected post-operative patient data.

AHG assisted the operation, collected post-operative patient data and reviewed the manuscript.

HAA did the pre-operative and post-operative patient management and reviewed the manuscript.

MTD assisted the operation and reviewed the manuscript.

Guarantor

Workneh Tesfaye Deme.
Abebe Bezabih Tessema.

Research registration number

NA.

Conflict of interest statement

The authors declare that they have no competing interests.

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