Case Report

A Rare Case of Acyanotic Congenital Heart Disease, Large Patent Ductus Arteriosus with Pre-ductal Coarctation of Descending Thoracic Aorta with Patent Ductus Arteriosus Closure and Extra Anatomical Bypass Grafting

Abstract

We report a case of 18-year-old female patient with large patent ductus arteriosus (PDA)-preductal coarctation of descending thoracic aorta. She underwent large PDA closure with a prosthetic graft from ascending aorta to descending thoracic aorta by mid-sternotomy on cardiopulmonary bypass machine under total hypothermic circulatory arrest.

Keywords: *Cardiopulmonary bypass machine, coarctation of aorta, patent ductal arteriosus, total hypothermic circulatory arrest*

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Introduction

Patent ductus arteriosus (PDA) is a relatively common congenital heart defect. The condition occurs more often in premature infants (on average, occurring in about 8 of every 1000 births).[1] However, PDA also occurs in full-term infants (on average, occurring in about 2 of every 1000 births). In children who were born at term, the incidence of PDA has been reported to be ≈1 in 2000 births.[2] The reported incidence of PDA varies due to methodological differences related to the population group studied, age of consideration, and method of detection.[3] This accounts for ≈5% to 10% of all congenital heart disease. However, if we include children with "silent" patent ductus (those discovered incidentally by echocardiography performed for another purpose), the incidence has been estimated to be as high as 1 in 500. The female to male ratio is ≈2:1 in most reports.[4] The incidence of preductal coarctation of aorta in adults is a rare phenomenon which is our case. A heart murmur (an extra or unusual sound heard during the heartbeat) may be the only sign that a baby or child has PDA The ductus arteriosus is a normal and essential fetal structure that becomes abnormal if it remains patent after the neonatal period. Coarctation of the aorta (COA) is a congenital (present at birth) heart defect involving a narrowing of the aorta. The narrowed segment called coarctation can occur anywhere in the aorta, but is most likely to happen in the segment just after the aortic arch. Most cardiac surgical procedures can be accomplished using cardioplegia-induced cardiac arrest and cardiopulmonary bypass (CPB) to maintain perfusion of other organs. In some situations, however, the underlying pathology or the nature of the surgery proposed necessitates complete cessation of the circulation. The use of profound systemic hypothermia to preserve organ function during cessation of the circulation is termed deep hypothermic circulatory arrest (DHCA). The theory is to cool the patient, cease blood flow to the brain, and rely on the hypothermic protective effects of decreased cerebral metabolic rate of oxygen ($CMRO_2$). $CMRO_2$ decreases significantly with hypothermia; there is a 6%–7% decrease in metabolism per 1°C decreased.

Case Report

We report a case of an 18-year-old female patient who was a known case of PDA and preductal COA, admitted in our hospital for invalidating shortness of breath on exertion, palpitation for the past 1½ year. On admission, the blood pressure was 126/78 mmHg in the left arm supine position and lower limb blood pressure

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was 90/50 mm hg. Oxygen saturation was 95% in all the extremities. The resting electrocardiography showed suspected left ventricular hypertrophy, slightly depressed ST segment (lateral), and prolonged QT [Figure 1]. The X-ray chest showed cardiomegaly and prominent bronchovascular margins. The transthoracic echocardiography [Figure 2] revealed an preductal aortic coarctation with a 70 mmHg gradient, and showed a large PDA with left to right shunt (systolic/diastolic pressure = 50 mm Hg/20 mmHg) with dilated left atrium and left ventricle (LV). The computed tomography (CT) chest angiography [Figure 3] showed an abnormal communication between distal pulmonary trunk toward left side and descending aorta with a defect of about 9.7 mm leading to dilatation of pulmonary artery (PA) and left pulmonary vein: suggestive of PDA. There was an evidence of short segment abrupt stenosis of the distal arch of the aorta with the maximal caliber of about 19 mm: suggestive of coarctation of aorta. The heart was enlarged in size with cardiothoracic ratio >60%: suggestive of cardiomegaly with left ventricular configuration. Ascending thoracic aorta 28 mm, arch of aorta 27 mm, descending aorta 26 mm. The CT Abdominal Angiography was normal. On admission, the patient had no fever and the laboratory tests showed normal leukocytes (4700/mm^3) and the C-reactive protein was negative. The patient was hemodynamically stable and considering all other parameters such as normal renal function test, liver function test, thyroid profile, and normal blood sugar level. We decided to proceed with PDA closure and extra-anatomical bypass grafting (using 18 mm knitted polyester collagen-coated intergard tube prosthetic graft) from ascending aorta to descending thoracic aorta by mid-sternotomy on CPB [Figure 4].

Operative findings

Large PDA around 1.30 cm in usual position with pre-ductal COA and severe LV hypertrophy. Gradient of >35 mm of Hg between radial and femoral arterial pressures (femoral < radial).

Operation

We proceeded with after routine monitoring lines placed, injection ranitidine, avil, dexamethasone were given as a premedication followed by fentanyl then injection etomidate as an induction agent and injection vecuronium as a mucle relaxant. Oxygen and sevoflurane were the gases used. Parts were cleaned, chest opened through midline sternotomy. Above mentioned external findings noticed and systemic heparinization done to achieve activated clotting time of >480 s. Aortic and two stage single venous cannulation done and hypothermic (28°C). CPB established and the LV was vented by a left superior pulmonary venting cannulae.

Aorta X – Clamped. Modified St. Thomas cold sanguinous cardioplegia delivered in the root. PA was opened but there was excessive flow from aorta to PA from PDA which was not manageable, so flows were reduced to 0.5 l/min and tried to close PDA. Still the flow was high so it was decided to go for total circulatory arrest (TCA) on deep hypothermia. Patient was cooled to 18°C, TCA was achieved. For neuroprotection injection thiopentone sodium 5 mg/kg, injection methylprednisolone 1 g was given. Head was packed with ice packs. PDA closed from PA side using pericardial pledgeted 4–0 prolene. Total duration of TCA was 4 min. Full flow CPB was resumed and the patient was rewarmed to 28°C over 15 min. Heart vented and lifted up to expose descending thoracic aorta posteriorly. Distal end of 18 mm knitted polyester tube graft anastomosed at this position using 4–0 prolene. Graft was directed upward from space below inferior vena cava and anterior to right inferior pulmonary vien, right lateral aspect of right atrial and anastomosed to the right lateral aspect of ascending aorta using 5–0 prolene. After adequate de-airing X clamp removed and intrinsic sinus rhythm achieved. Patient gradually weaned off bypass, after gradual re-warming and electrosystolic training by an external pace maker, with stable hemodynamics. De-cannulation done and protamine

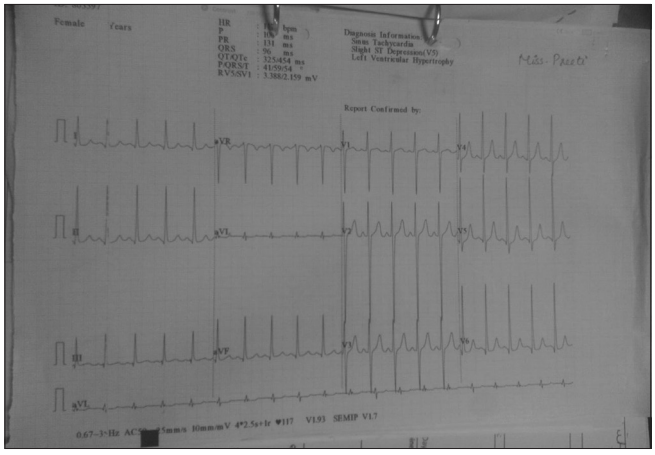

Figure 1: Electrocardiography showing suspected left ventricular hypertrophy, slightly depressed ST segment (lateral), prolonged QT

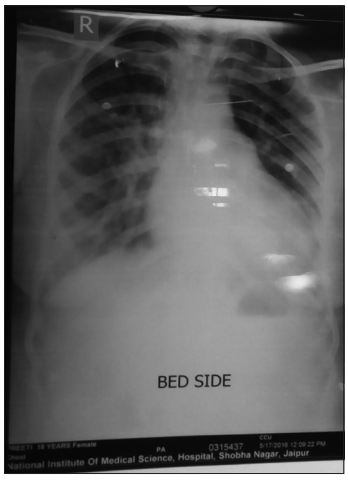

Figure 2: X-ray chest showing cardiomegaly and prominent broncho-vascular margins. The transthoracic echocardiography

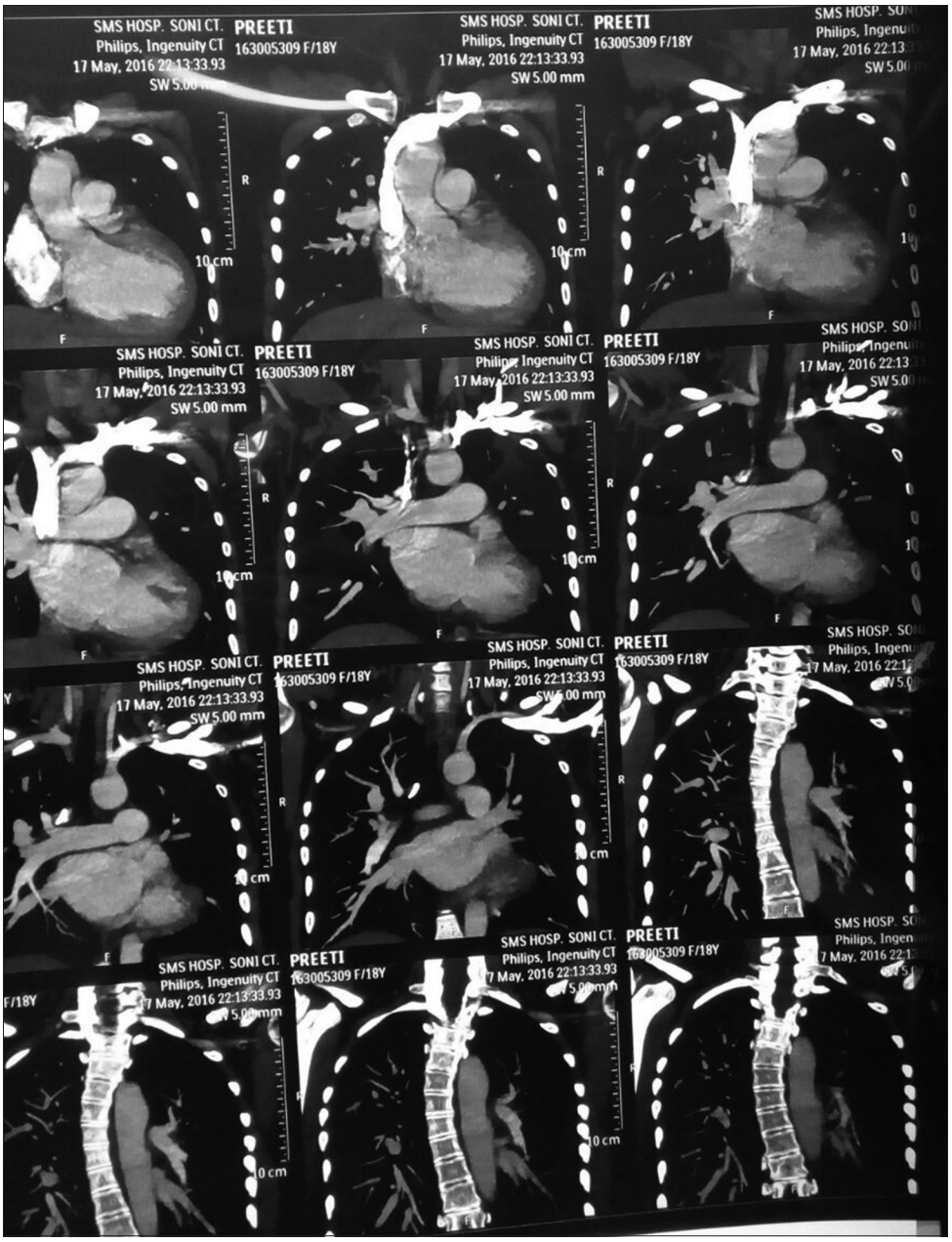

Figure 3: Computed tomography chest angiography showing an abnormal communication between distal pulmonary trunk toward left side and descending aorta with a defect of about 9.7 mm leading to dilatation of pulmonary artery and left pulmonary vein

reversal done. Proper hemostatis secured and chest drained using two mediastinal drains. One ventricular and one atrial pacing wire applied prophylactically. Sternum closed followed by skin and patient was shifted to cardio thoracic vascular surgery Intensive Care Unit in a satisfactory condition.

Discussion

Aortic coarctation is a well-known congenital cardiovascular malformation, which can remain asymptomatic and unknown until adulthood and result in cardiovascular complications and premature death. It can be associated in more than 50% of patients to major cardiac pathologies such as bicuspid aortic valve diseases and ventricular septal defect, which makes the surgery more challenging. The ductus arteriosus is an essential fetal structure. A failure of constriction of smooth muscle within the wall of this ductus in postnatal period leads to PDA. It is the second most common form of congenital heart defect, accounting for about 5%–10% of all congenital heart disease, with a reported incidence of nearly 1 in 2000 births. On the other hand, coarctation of the aorta is the fifth most common form of congenital heart defect, accounting for about 6%–8% of all congenital heart disease, with an estimated incidence of nearly 1 in 2500 births. There are few cases in which both conditions,

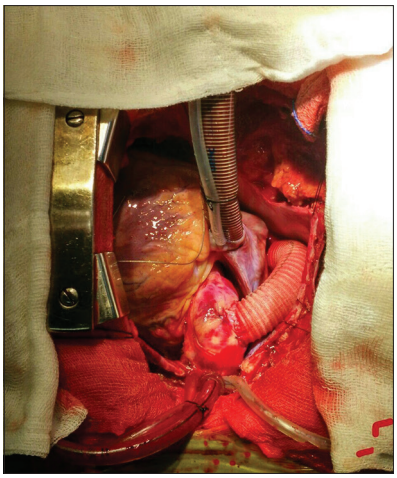

Figure 4: Patent ductus arteriosus closure and extra anatomical bypass grafting (using 18 mm knitted polyester collagen-coated intergard tube prosthetic graft) from ascending aorta to descending thoracic aorta by mid sternotomy on cardiopulmonary bypass

PDA and COA, may coexist. Surgery is usually suggested in patients with PDA and coarctation of the aorta. Although the recommended treatment for the combination of aortic coarctation and PDA in infants and children is surgery, transcatheter treatment approaches can be effectively applied as a non-surgical option. The major benefits of transcatheter interventions are avoidance of thoracic surgery, thoracotomy scar, and prolonged hospitalization associated with it. Several reports suggest that both lesions can be treated successfully, either simultaneously or sequentially, using transcatheter treatment approach. The sequential approach avoids catheter and wire manipulation and reduces the risk of aortic dissection and device mal-positioning. However, this approach is associated with high cost and a need for second hospitalization and intervention, which are unpleasant for the child and family. Hence, an effort should be made to treat both the conditions simultaneously, when possible. Death in these patients is usually due to heart failure, coronary artery disease, aortic rupture/dissection, concomitant aortic valve disease, infective endarteritis/endocarditis, or cerebral hemorrhage[5,6] There are a few reports of patients first diagnosed with uncorrected aortic coarctation at very late age[7,8] and there is no consensus on how to manage them. In this report, we present the case of an 18-year-old female first diagnosed with severe aortic coarctation. The patient was relatively asymptomatic until she presented with palpitations and exertional dyspnea. Classically described, the narrowing could be proximal (preductal) to the ductus arteriosus and the blood flow in aorta distal to the narrowing is dependent on the ductus arteriosus and this is the most common type in neonates. Hence, closure of the duct will result in early severe neonatal presentation. Hence, our is a rare case. In postductal COA, the narrowing is distal to insertion of the ductus arteriosus and this is the common type in adult patients. Weak or absent femoral pulses are present in 92% of patients.[9] Pulse oximetry measurement at pre- and post-ductal sites should be monitored. In the case of critical coarctation of the aorta, the preductal saturation is higher than the postductal saturation and differential cyanosis exist.[10] Mortality rates of between 8% and 15% have been reported following DHCA with stroke rates of 7%–11%. Predictors of stroke after DHCA are increased age, longer length of DHCA, and atheroma or thrombus in the aorta. Subtle, long-term cognitive dysfunction is common, particularly problems with short- and long-term memory and information processing.[9]

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Conflicts of interest

There are no conflicts of interest.

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