Association of interrupted aortic arch, aortopulmonary window with anomalous origin of the right pulmonary artery from the aorta, one-stage repair and postoperative outcomes: A case report

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Introduction

Association of interrupted aortic arch (IAA), aortopulmonary window (APW) with aortic origin of the right pulmonary artery (RPA), first described by Berry and colleagues in 1982, is an extremely rare congenital heart disease (1). IAA is seen in 0.7 to 1%–4% of congenital heart disease. Additionally, it has been reported that APW can be associated with IAA at a rate of 4% (2). Though rare presentations may be seen in adolescents, clinical presentation mostly occurs in the neonatal or infancy period. Early diagnosis plays an important role in the management of this disease, and, with late presentation, hemodynamics may be impaired as a result of ductus arteriosus closure. Surgery is mandatory as this is fatal. One-stage surgery can be performed, even on a low birth weight neonate (3). We report a rare case of a newborn requiring complex surgery who presented with shock and was diagnosed as having an association with type A IAA, type 3 APW, with RPA arising from the aorta.

Case Report

A two-day-old boy was admitted to our emergency unit in a disturbed general condition with prearrest status. At 39 weeks of gestation, he was born by spontaneous vaginal delivery with an Apgar score of 9/10, weighing 3,060 g.

Upon admission to the neonatal intensive care unit, we observed severe congestive heart failure and cyanosis with tachypnea, tachycardia, hypotension and hepatomegaly. His body temperature was 35.7°C, heart rate 180 bpm, respiratory rate 80 bpm, and he had a systolic blood pressure of 40 mm Hg, and a pulse oximetry saturation of 50%. During cardiovascular examination, we observed a grade 3 systolic ejection murmur on the left margin of the sternum with a gallop rhythm. Fluid resuscitation and intubation were performed immediately with a diagnosis of septic or cardiogenic shock. Metabolic acidosis (pH 6.9) and a severely high level of lactate (22 mmol/L) were found in the blood results. On echocardiography (Fig. 1a, 1b), we

observed type 3 APW, with RPA arising from the aorta, type A IAA, and secundum atrial septal defect. Low cardiac output was managed with prostaglandin E1 and dobutamine infusion. After stabilization, he underwent surgery at nine days of life. Through a median sternotomy, the aorta and head vessels were mobilized, and the left and right branch (originating from the ascending aorta) of pulmonary arteries were encircled. The distal ascending aorta was cannulated past the distal extension of the APW. The branch pulmonary artery (PA) snares were tightened when the cardiopulmonary bypass was initiated. Under deep hypothermic total circulatory arrest, the descending aorta was anastomosed to the undersurface of the distal aortic arch, in an end-to-side manner, after all the ductal tissues had been excised. The aortic cross-clamp was placed between the APW and the reconstructed arch and the patient slowly warmed. The APW was approached by way of transverse aortotomy. After the bovine pericardial patch was placed in the origin of the RPA, the RPA and main pulmonary artery (MPA) were connected, and followed by reconstruction of the ascending aorta over the RPA near the anterosuperior portion of the APW. An ostium secundum atrial septal defect was patched with a piece of bovine pericardium. The duration of total circulatory arrest, the lowest nasopharyngeal temperature, and aortic clamp time were 26 minutes, 20°C, and 171 minutes, respectively. In the postoperative period, he was extubated on the second day and transferred to pediatric ward on the thirteenth day of hospitalization.

Due to a progressive increase of RPA stenosis in postoperative follow-up, which was detected as mild stenosis immediately after surgery, a pulmonary balloon angioplasty was planned to be performed at 10 months. In the hemodynamic study, right ventricular systolic pressure was 70 mm Hg, and the systolic main and RPA pressures were 64 and 16 mm Hg, respectively. A stenotic segment of the RPA was measured as having a diameter of 2.6 mm. A balloon angioplasty was applied to the RPA using the 6*20 mm Tyshak II balloon dilatation catheter (B. Braun Interventional Systems, Inc., Bethlehem, PA, USA). There
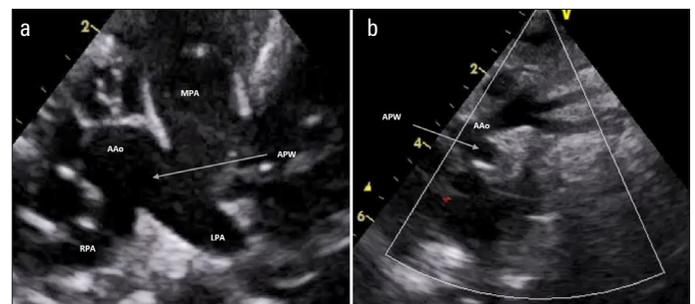

Figure 1. Two-dimensional transthoracic echocardiography images of the patient. a) Parasternal short axis view, showing aortopulmonary window defect between the aorta and main pulmonary artery, anomalous origin of the right pulmonary artery from the ascending aorta. b) Suprasternal view showing an interrupted aortic arch type A (interruption occurs just beyond the left subclavian artery).

AAo - ascending aorta; APW - aortopulmonary window; LPA - left pulmonary artery; MPA - main pulmonary artery; RPA - right pulmonary artery
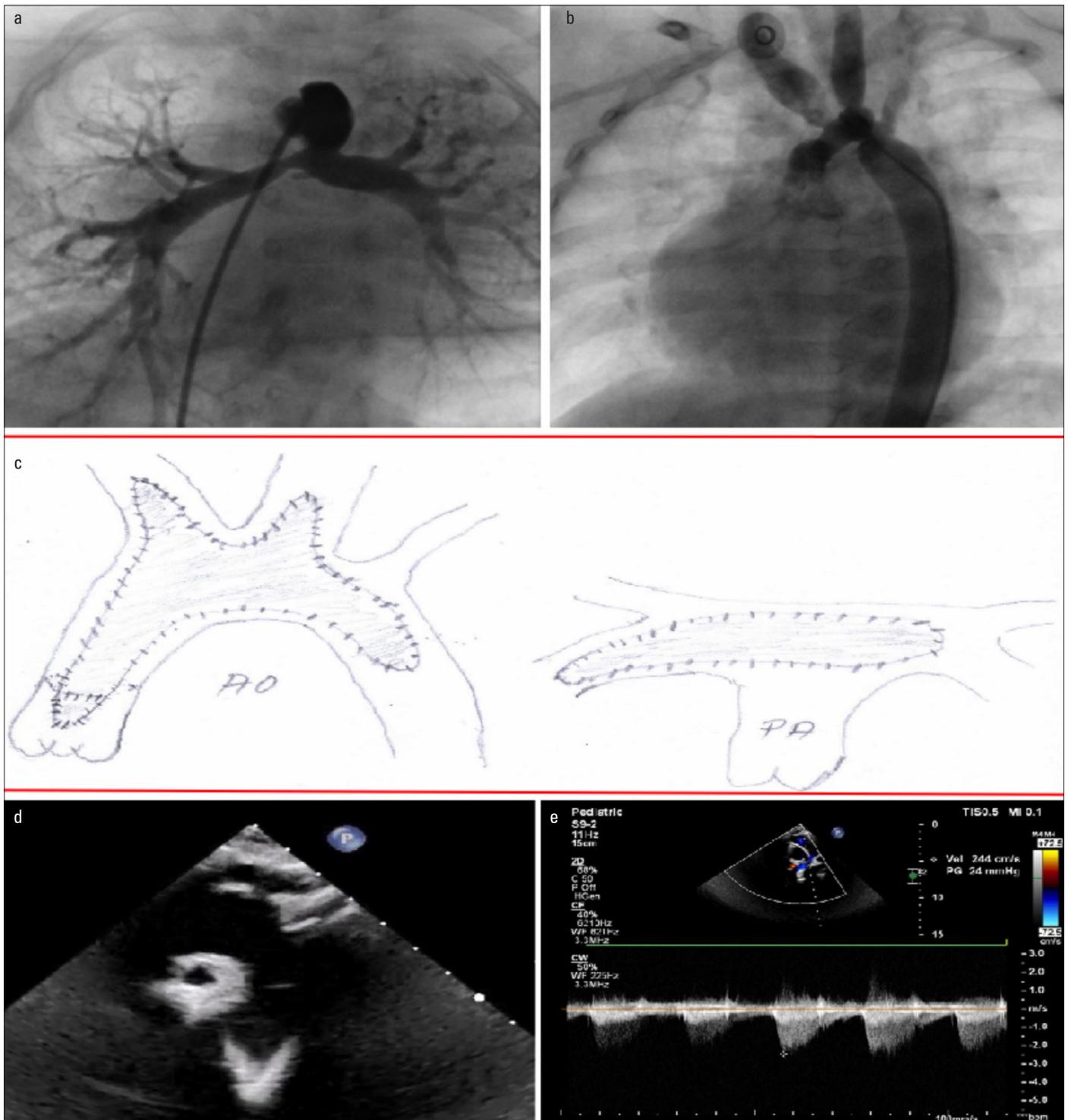
Figure 2. a) Caudal (30°) projection of pulmonary artery angiography showing moderate to severe stenosis of the proximal right pulmonary artery. b) Left anterior oblique (25°) projection of the aortogram revealing significant narrowing of the aorta before the brachiocephalic trunk with hypoplastic arcus aorta. Significant stenosis was also seen in the origin of the innominate and carotid arteries. c) Augmentation of the ascending aorta, transverse arch, innominate artery and left common carotid artery by pericardial patch plasty, and augmentation of the hypoplastic right proximal pulmonary artery and narrow pulmonary bifurcation by bovine pericardial patch. d) Postoperative two-dimensional transthoracic echocardiography images of the patient, suprasternal long-axis view, showing the aortic arch and descending aorta continuity with no stenosis. e) Doppler study demonstrating no significant gradient with a peak flow velocity in the descending aorta of 2.3 m/s.

was moderate relief in the proximal segment of the RPA after the balloon angioplasty with a residual pullback pressure gradient of 30 mm Hg. Because a gradual increase in the pressure gradient of the aortic arch was observed in follow-up, which had not been detected in early postoperative follow-up, catheterization was planned for 16 months after surgery. We observed a sys-

tolic MPA pressure of 82 mm Hg, withdrawal pressure between the RPA and MPA of 50–55 mm Hg, and systolic left ventricular pressure of 215 mm Hg, and withdrawal pressure between the ascending and descending aorta was 110 mm Hg, with a hypoplastic and narrow segment of arcus before the branch of the brachiocephalic trunk and moderate to severe stenosis of the RPA (Fig. 2a, 2b). A reoperation was planned. An arterial cannulation was placed via the innominate artery, and RPA reconstruction was performed with a pericardial patch. Then, the transverse arch and narrow segments of the brachiocephalic and left carotid artery origins were opened from the anterior aspect of the aorta to the distal portion of the left subclavian artery, and aortic arch reconstruction was carried out with a pericardial patch. A stenotic segment of the ascending aorta was widened using a patch, which was extended toward the non-coronary cusp (Fig. 2c). The lowest nasopharyngeal temperature and aortic clamp time were 20°C and 120 minutes, respectively. Total circulatory arrest was not induced; antegrade cerebral perfusion was used.

In postoperative follow-up, there was no gradient in the aortic arch and there was a mild stenosis at the RPA with a peak systolic pressure gradient of 26 mm Hg on echocardiography (Fig. 2d, 2e).

Discussion

Berry syndrome is a rare congenital heart disease with a satisfactory outcome when diagnosed early without clinical deterioration and surgery is performed. When the ductus arteriosus closes, there may be clinical deterioration and hemodynamic instability. In this case, the patient presented with shock and multi-organ failure and was started on prostaglandin E1 immediately after diagnosis. Congestive heart failure, low cardiac output, and pulmonary distress may also be found in clinical presentation due to pulmonary congestion. Tracheal airway compression and respiratory distress may be found in the case of PA dilatation.

Detailed echocardiographic evaluation of the complex anatomy plays a critical role. Echocardiography can be used to assess aortic arch direction, location of interruption, aortic arch length, size of aortic valve and left ventricle, and size and type of APW associated with the lesion (4). Associated lesions are atrial septal defect, ventricular septal defect, left ventricular outflow tract obstruction, tetralogy of Fallot, complete vena cava azygos continuity, left superior vena cava, aberrant right subclavian artery, and pulmonary sling anomaly (5).

Prenatal diagnosis before the closure of the ductus arteriosus, shock or circulatory collapse is important. In the prenatal period, left ventricular outflow tract anatomy, large vessels and semilunar valves should be examined carefully. In our case, an accurate evaluation of the aortic arch anatomy was achieved with transthoracic echocardiography, and there was no need for additional imaging before surgery.

After diagnosis, urgent surgery was necessary. Since the systemic circulation is dependent on the ductus arteriosus,

prostaglandin E1 should be started before surgery to stabilize metabolic condition and prevent the worsening of end-organ perfusion. Inotropic supportive treatment should be planned to ensure systemic perfusion and prevent renal failure. In this case, clinical stabilization was achieved with prostaglandin E1, and emergency surgery was planned after stabilization.

Surgery can be performed as a single-stage or staged repair. For the optimal surgical outcome, it is important to know the complex anatomical features preoperatively and the potential for future growth in large vessels. In a single-stage repair, the APW is closed, and aortic arch continuity is ensured, while RPA binds to the MPA. Aortic arch repair is made with a homograft or autologous pericardial patch, or with direct anastomosis. The most appropriate option for aortic arch repair is an end-to-end anastomosis between the aortic arch and the descending aorta without the use of a prosthetic conduit. There is a potential for the anastomosis to increase in size due to patient growth. In order to not overstress the anastomosis, extensive dissection of the aortic arch and descending aorta should be avoided (6). The APW can be closed with a pericardial or synthetic patch (7). A staged repair has been performed on patients who cannot tolerate cardiovascular bypass and aortic cross-clamping, e.g., those with very low birth weight or a poor general condition. To control pulmonary overcirculation, pulmonary banding and stent implantation of the PDA are performed as a staged repair. Following the initial surgical palliation, a total correction is performed at three to six months of age (8). In our patient, a single-stage repair was planned because his weight was suitable and his hemodynamic status was stabilized. Unfortunately, anastomotic stenosis occurred in the aortic arch and RPA during follow-up. We think that this was mostly due to wrinkle formation caused by the pericardial patch. Foreign body reaction may have been another cause of stenosis. Therefore, we switched to porcine pericardium for the second surgery since it contains less collagen and more water, and is thinner than bovine pericardium.

In the postoperative period, it is important to follow-up closely for any signs of stenosis of anastomosis sites or the PA, bronchial compression, and extrinsic airway compression (7). Reoperation may be needed in the case of anastomotic stenosis, residual APW, left phrenic nerve paralysis, or end-stage cardiomyopathy (9). Percutaneous balloon angioplasty can be performed when stenosis occurs (7, 10). In PA hypoplasia, stent implantation can be performed (10). In this case, balloon angioplasty was applied to the stenosis of the RPA origin in the postoperative period. However, during the first year after PA balloon angioplasty, it was decided to perform surgical augmentation due to significant stenosis occurring in the distal ascending aorta and arcus aorta, the origin of the innominate and left carotid arteries, and also the right MPA division.

Conclusion

Berry syndrome is a rare entity that can progress fatally if not diagnosed early, and may present with shock or low cardiac output conditions due to closure of the ductus arteriosus. A

detailed echocardiographic evaluation of the complex anatomy plays a critical role in surgery, and urgent surgery is necessary. Patients should be monitored closely for stenosis that may develop after surgery and, if this occurs, an interventional approach or reoperation may be planned.

Ethical standards: The authors assert that all procedures contributing to this work comply with the ethical standards of the relevant national guidelines on human medical regulations and with Helsinki Declaration of 1975, as revised in 2008.

Informed consent: Written informed consent was obtained from all patient's parents.

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