Common arterial trunk and double aortic arch: a rare association

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Abstract

Background: The association of double aortic arch and common arterial trunk is extremely rare. The initial surgical approach depends on the patient's clinical condition and associated cardiac anomalies. Aim: To report a rare association of common arterial trunk with double aortic arch in a 4-month-old female infant. Methods: description of case of a rare association where double aortic arch was not diagnosed initially, surgical repair was done successfully. Results and conclusions: associated cardiovascular anomalies may have an impact on management and outcome. Magnetic resonance imaging and computed tomography may be useful in assessment of this rare association. Complete repair has favorable outcome.

INTRODUCTION

Common arterial trunk (CAT) is a rare congenital malformation characterized by a single arterial trunk arising from the heart, overriding the interventricular septum and gives rise to the systemic, pulmonary, and coronary circulations[1]. The incidence of defect is 6–10/100,000 live births, it is occasionally associated with aortic arch anomaly such as right aortic arch and aortic arch interruption[1, 2]. Double aortic arch (DAA) is the most common vascular ring, and results from persistence of both 4^{th} arches, compressing the trachea or esophagus or both[2]. Common arterial trunk and double aortic arch are rarely associated and there are only a few cases in the literature research about this association. We present one case that were approved by the local institutional review board. The approval included a waiver of informed consent because it does not show personal data of patients.

CASE REPORT

A 4-month-old, female child presented with recurrent episodes of tachypnea and diaphoresis. She had signs of cardiac heart failure, respiratory distress, fever, costosternal retractions and bilateral rales; resting oxygen saturation was 40% and systolic murmur. On the chest X-ray, increased vascular marking and cardiomegaly were noted. Echocardiography showed type I CAT with mild regurgitant three-cusp truncal valve overriding a large ventricular septal defect and bidirectional shunt. Computerized tomography (CT) showed (Figures 1 and 2) DAA with a dominant left-sided arch, right aortic arch gave rise to the right common carotid and right subclavian arteries, and the left arch gave off the left common carotid and left subclavian arteries. Surgery was performed via a standard median sternotomy. The establishment of cardiopulmonary bypass included ascending aorta, bicaval cannulation and cooling to 25 °C. Cardiopulmonary bypass and aortic cross-clamping time was 102 and 72 minutes respectively with anterograde cerebral flow. Surgical management included division of the right aortic arch along with CAT repair: pulmonary artery was disconnected from the arterial trunk and the aorta was reconstructed with a bovine pericardium patch; the ventricular septal defect was closed with a bovine pericardium patch. A 14 mm home-made valved Woven Dacron© conduit was used to reestablish the continuity between the right ventricle and the pulmonary arteries (Figure 3).

Postoperative course was uneventful, and the infant was discharged on postoperative day 15, she is currently on follow up.

DISCUSSION

CAT with DAA remains a very rare condition. Patients with CAT usually present with respiratory distress, cyanosis, diaphoresis, and poor feeding. Patients with DAA also present with respiratory distress, stridor, and feeding difficulty in infancy due to tracheal and esophageal compression by the vascular ring[3,4]. Therefore, when DAA coexists with CAT, symptoms associated with tracheal compression may be mistakenly attributed to increased pulmonary blood flow and severe pulmonary arterial hypertension in CAT, thus missing the important contribution of the arch anomaly to the symptoms[3]. In our patient, we found predominant symptoms attributable to the markedly increased pulmonary flow. Echocardiography allows reliable assessment of the hemodynamics, anatomy, and physiology of CAT[5], but it appears to be of little value for the diagnosis of DAA. CT scan is useful for its definitive diagnosis and can show compression of the trachea, bronchus, and esophagus[4,6]. In our case, echocardiography diagnosed a CAT, but no vascular ring could be seen. A CT was then performed for additional anatomic information and showed a DAA. Therefore, MRI and CT can be performed preoperative for the determination of the exact anatomy to minimizes the risk of misdiagnosis or inadequate treatment. The initial surgical approach depends on the patient's clinical condition and associated cardiac anomalies; increased pulmonary pressure due to hyper blood flow should not contraindicated the repair.

CONCLUSION

The association of the CAT with DAA is extremely rare and diagnosis is difficult because symptoms attributable to CAT may mask those of the DAA. It is important to bear all these possible associations in mind when assessing the patient with CAT because it may have an impact on management and outcome. Complete repair has favorable outcome. The behavior of the pulmonary vasculature is variable and should be individualized in each case and patients should not be excluded from treatment solely based on age and type of defects.

AUTHORS' CONTRIBUTIONS:

Concept/design: Cervantes-Salazar, Peralta-Santos, and Flores-Sarria.

Drafting article: Flores-Sarria and Peralta-Santos.

Critical revision/approval: Cervantes-Salazar, Peralta-Santos, Flores-Sarria, Ramirez-Marroquin and Calderon-Colmenero.

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FIGURE LEGENDS

Figure 1: CT angiography reconstruction: (A) there is a common trunk of the ascending aorta and main pulmonary artery; (B) posterior view of the double aortic arch with a dominant left-sided arch and right aortic arch forming the descending aorta; (C) cranial view of right and left aortic arches forming complete vascular ring. **AO:**ascending aorta; **CAT:** common arterial trunk**; DA:**descending aorta; **MPA:** main pulmonary artery; **LAA:**left aortic arch; **RAA** : right aortic arch.

Figure 2: CT axial section shows: (A) main pulmonary artery and its two branches right pulmonary artery and left pulmonary artery, (B) aortic arches encasing the trachea in a circular fashion. (C) CT coronal view show a double aortic arch with the descending aorta. **DA:**descending aorta; **MPA:** main pulmonary artery; **LAA:**left aortic arch; **LPA:** left pulmonary artery; **RAA:**right aortic arch; **RPA** : right pulmonary artery**.**

Figure 3: (A) intraoperative photograph showing the origin of the main pulmonary artery from the common trunk. (B) Reconstruction of the aorta with a bovine pericardium patch. (C) 14 mm home-made valved Woven Dacron© conduit. (D) Proximal anastomosis of the conduit to the right ventricle. **AO:** ascending aorta; **CAT:** common arterial trunk; **MPA:** main pulmonary artery.