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AORTIC ORIGIN OF THE RIGHT PULMONARY ARTERY FROM ASCENDING AORTA IN A 4YEAR OLD CHILD WITH TETRALOGY OF FALLOT.

Anaesthesiology

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ABSTRACT

Aortic origin of the right pulmonary artery from the ascending aorta is a rare and potential fatal kind of cardiac malformation associated with Tetralogy of Fallot (TOF). Right Pulmonary Artery arising from the aorta occurs more commonly as an isolated lesion, but the anomalous origin of the left PA from the aorta is more commonly associated with tetralogy of Fallot (TOF)[1,2]. In our case Right Pulmonary Artery is arising from Ascending Aorta and is in association with Tetralogy of Fallot thus making it one of the rare case presented.

KEYWORDS

Right pulmonary artery, Tetralogy of Fallot, Ascending Aorta, Cardiac malformation, Pulmonary Artery

INTRODUCTION

In 1868, Fraentzel first described that the anomalous origin of a branch pulmonary artery (PA) from the ascending aorta is a rare congenital heart malformation.[3] Anomalous right or left pulmonary artery arising from the aorta are considered relatively rare congenital heart diseases. There are several reported cases, mostly describing anomalous right pulmonary artery arising from the posterior aspect of the ascending aorta as an isolated lesion.[4]

The clinical presentation of these cases is in early childhood with congestive cardiac failure and with the onset of early pulmonary hypertension. In some, there is an absence of cardiac failure or a very short abbreviated period of failure followed by the development of pulmonary vascular disease. This rare disease is thought to be because of failure of migration of the embryonic branch pulmonary artery to reach the pulmonary trunk end of the truncoaortic sac before septation occurs. There are known associations with an anomalous right pulmonary artery (RPA): patent ductus arteriosus (PDA), interrupted aortic arch (IAA), aorto-pulmonary window (AP window) are frequently encountered with Tetralogy of Fallot (TOF). This combination of lesions occurs in 3 of every 10,000 live births, and accounts for 7–10% of all congenital cardiac malformations. The patho-physiological characteristic change of AORPA (Aortic Origin Right Pulmonary Artery) is early and rapid development of pulmonary hypertension, and the several mechanisms involve in which includes (1) circulating vasoconstrictor substances, (2) neurogenic crossover from the unprotected lung to the protected one, (3) the development of pulmonary hypertension secondarily after left ventricular failure.

We are describing the case of a child diagnosed with Fallot's tetralogy, who presented with Bluish discolouration of tongue and nails since few months after birth, chest pain and shortness of breathness. CT angiography disclosed Fallot's tetralogy associated with anomalous origin of a pulmonary artery (PA) branch from the left side of the ascending aorta. Surgical correction of the defect was performed and the postoperative course was favorable.

CASE REPORT:

A 4 years old boy presented to our institution, with a history of bluish discolouration of nails of both extremities, chest pain associated with shortness of breath. He was delivered vaginally at term, weighed 3.0 kg, and had an APGAR score of 9/10,breastfed well, had an uneventful medical history for the first 2 months of life, and attained weights of 7.2 kg. From the third month onwards, he started with aforementioned complaints, and the family sought medical attention from several health facilities. With time, the child became progressively tachypneic and dyspneic, cyanosed, had difficulties in breastfeeding, and started losing weight. The boy underwent echocardiographic (ECHO) examination confirming diagnosis. Physical examination conducted at our institution revealed a small for age boy with bluish discolouration of nails, lips and tongue. The jugular venous pressure was not raised and there was no precordial hyperactivity. The blood pressure was 108/88 mm Hg, heart rate was 131 beats/min and regular, and apex beat was felt at the seventh intercostal space lateral to the midclavicular line. The first (S1) and second (S2) heart sounds were heard but auscultation of the pulmonic area revealed accentuated pulmonary component (P2); no murmur was heard. The respiratory rate was 38 breaths/min, oxygen saturation was 94% at room air, and on auscultation nothing significant was appreciated. Hematological tests revealed hemoglobin 15.9 g/dL, leukocytosis (10.33 cells/μL) with lymphocyte predominance (63%) and platelet count of 147 000/μL. ELISA for HIV, HbsAg, HCV were negative. Chest X-ray revealed bilateral pruning of blood vessels (Figure 1). An ECHO revealed Levocardia, Large mal-aligned perimembranous Ventricular Septal Defect(VSD) with outlet muscular extension shunting bidirectionally with 50% aortic overriding,severe infundibular + valvar+ supra valvar narrowing PS gradient 72mm hg along with doming and thickened pulmonary valve annulus 9.2mm.Confluent branch Pas, RPA = 11mm and LPA = 12.5mm (Fig 2). The child was further investigated with a cardiac computed tomography scan, which finally revealed following:

1) Both pulmonary arteries arising arising separate origin. No pulmonary trunk was visualized.
2) Right subclavian artery arising from arch of aorta, left side cross midline anterior to oesophagus.
3) Both vertebral arising from arch of aorta
4) Right pulmonary artery arising from root of aorta adjacent to the origin.(Fig 3).

Other investigations were within normal range. Patient was planned for surgery.

Operative notes: Patient Supine. Under full asepsis, mid sternotomy done, pericardial patch harvested. CPB established using ascending aortic and bi-venous cannulation. Aorta clamped. Under cold cardioplegic arrest. RA opened and LA vented. MPA opened and RVOT opened. Resection done. VSD closed through RA using Sauvage patch with 5/0 continuous prolene suture. RPA disconnected, Mobilized and connected with MPA. Trans-annular Pericardial patch constructed over RVOT, MPA using prolene 6/0 suture. RA closed. PRV/LV 0.010, no significant gradient between RV inflow Infundibulum and MPA. Heart de-aired. Aorta released. CPB weaned off. De-cannulation. Haemostasis checked. Four drains (Two pleural & two mediastinal). One atrial & one ventricular pacing wire. Sternum closed with steel wires. Wound closed in layers and patient shifted to CTVS ICU for further care. Patient was Extubated on POD (post-operative day) 2 after he met extubation criteria.

DISCUSSION:

Tetralogy of Fallot is often associated with chromosome 22q11 deletion. The frequency of its association has been reported to be between 5% and 20% and the percent of AORPA occupies less than 0.1 % of congenital heart disease. It has also been well documented that TOF often occurs together with anomalies of the aortic arch and its branches, including right aortic arch, and (less frequently) elongation and high positioning of the aortic arch, aberrant origin or isolation of

the SCA, isolation of the left pulmonary artery, absence of the ductus arteriosus, and the presence of major aorto-pulmonary collateral arteries.

Pulmonary artery arising from the aorta was first described by Fraentzel in 1868 and since that time, about 131 cases have been reported.[2,3] This entity is rare, and can involve LPA or RPA or both.[6]

Pulmonary hypertension has been described as starting early in most of the cases reported, and intervention had been mainly carried out in infancy, although there are rare cases of non-operated survivors in adulthood with apparently normal pressures in the lungs.[7] Patients with AORPA who experienced surgical treatment early in life have excellent short-and long-term outcome [8] Various types of surgical techniques are employed for the repair of the anomalous pulmonary arteries, with the most frequently employed being direct anastomosis. Alternative techniques include end to end anastomosis with a synthetic graft, interposition with a homograft patch, aortic flap, etc. The results in the neonatal period have been promising, but due to the rarity of this lesion, it is difficult to predict the optimal timing or the best repair technique.

CONCLUSION:

The results of the correction of Tetralogy of Fallot have been improved. However, Tetralogy of Fallot complicated with other anomalies still has some difficulties for total correction.

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