Surgical Management of Pulmonary Artery Sling in a Pediatric Patient

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Patient: Male, 12-year-old
Final Diagnosis: Pulmonary arteriovenous malformations
Symptoms: Chest tightness
Clinical Procedure: —
Specialty: Cardiac Surgery

Objective: Congenital defects/diseases
Background: Pulmonary artery sling (PAS) is an anatomical vascular anomaly due to the origin of the left pulmonary artery from the right pulmonary artery, which runs posteriorly between the esophagus and trachea, resulting in compression of adjacent structures. Accurate evaluation for malformation of the pulmonary artery and severity of airway obstruction is essential to surgical strategy. This report presents the diagnosis and surgical management of pulmonary artery sling in a 12-year-old boy.

Case Report: A 12-year-old boy had chest tightness and wheezing after exercise for 6 years. He was diagnosed with PSA based on findings from imaging tests, demonstrating the left pulmonary artery originated from the middle of the right pulmonary artery and the tracheal carina was located at the site of the T6 thoracic vertebra. The main bronchus and esophagus were compressed by the left pulmonary artery due to its ectopic origin. Then, after comprehensive preoperative assessment, the patient underwent surgical repair of PAS.

Conclusions: This report highlights the importance of pulmonary artery sling diagnosis, imaging, and surgical planning, and the role of a multidisciplinary team in preoperative and postoperative patient management. An individualized strategy based on the preoperative assessment, intraoperative coordination among cardiologists, surgeons, and perfusionists, and careful postoperative management are the core elements for successful PAS repair.

Keywords: Stenosis, Pulmonary Artery • Thoracic Surgery • Pulmonary Heart Disease

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Introduction

Pulmonary artery sling (PSA) is an anatomical vascular anomaly leading to persistent compression of the trachea and esophagus. It has an abnormal origin from the posterior wall of the right pulmonary artery instead of the main pulmonary artery, while, the left pulmonary artery arises from the right pulmonary artery and runs posteriorly between the esophagus and trachea, causing a series of compression-related symptoms. Once diagnosed, PAS often coexists with respiratory and congenital cardiac malformations. Patients require a full work-up, including imaging and surgery planning [1]. Various imaging technologies, including echocardiography, CT, MRI, and bronchoscopy, are widely used for accurate diagnosis of PAS, including assessment of airway stenosis and the severity of tracheal compression [2]. This report presents the diagnosis and surgical management of pulmonary artery sling in a 12-year-old boy.

Case Report

A 12-year-old boy was admitted with a 6-year history of persistent chest tightness and wheezing after exercise. Imaging showed the left pulmonary artery (inner diameter 0.74 cm) originated from the middle of the right pulmonary artery (inner diameter 1.20 cm). The widened main pulmonary artery had an inner diameter of 2.49 cm (Figure 1A-1C). 3D-CT reconstruction of cardiovascular system showed the right pulmonary artery was 17.0 mm and the tracheal carina was located at the site of the T6 thoracic vertebra. The main bronchus and esophagus were compressed by the left pulmonary artery (10.1 mm) due to its abnormal origin (Figure 2A-2F).

After comprehensive preoperative assessment with no contraindications, the patient underwent surgical repair of PAS. The median sternotomy incision was selected and cardiopulmonary bypass (CPB) was established normally by ascending aorta and vena cava cannulation. The assisted CPB without cardiac arrest under moderate hypothermia was initiated after activated clotting time (ACT) exceeded 480 s. Visually, it was found that the left pulmonary artery originated from the right pulmonary artery. Then, the origin of left pulmonary artery was cut and the remaining incision of the right pulmonary artery was sutured completely. Another 1-cm incision was made in the left side of the main pulmonary artery at the anastomotic site between the isolated left pulmonary artery and the main pulmonary artery in front of the trachea. After qualified anastomosis, rewarming was initiated in a timely manner. The CPB was weaned when the temperature exceeded 36°C. Finally, heparin neutralization, hemostasis, and chest closure were performed, as planned.

After the surgery, the patient recovered smoothly and the PAS repair was satisfactory, in accordance with the encouraging Figure 1. (A-C) The preoperative images of echocardiography. The left pulmonary artery with inner diameter of 0.74 cm originating from the middle of the right pulmonary artery, with inner diameter of 1.20 cm. The widened main pulmonary artery with inner diameter of 2.49 cm was measured.
result of re-checking echocardiography, including grafted left pulmonary artery with inner diameter of 0.82 cm, remaining right pulmonary artery with inner diameter of 1.05 cm, and main pulmonary artery with inner diameter of 1.92 cm (Figure 3A-3C). During the perioperative stage, cardiologists were involved in the clinic management, including preoperative heart function adjustment and postoperative infection prevention, mechanical ventilation maintenance, and organic function protection.

**Discussion**

Generally, for special patients with uncommon diseases, the importance of cautious baseline evaluation, reasonable surgical strategy, and multidisciplinary cooperation should be fully emphasized. PAS is an uncommon congenital heart disease that is often misdiagnosed due to nonspecific symptoms and limited clinical knowledge [3]. Infants with PAS may also experience negative effects on lung volume, function, and lung development due to secondary tracheal stenosis [4]. Structurally, there are 2 types of PAS, depending on the location of tracheal carina. For Type I PAS, the tracheal carina is located at the site of the T4-5 thoracic vertebra, mainly compressing the distal trachea. For Type II PAS, abnormal tracheal carina, even with inverted “T” bifurcation, is located at the site of the T6 thoracic vertebra, so-called pseudo-tracheal carina, and trachea stenosis is often detected [5]. At the initial stage, due to absence of special or characterized symptoms, PAS may be misdiagnosed...
Figure 2. (A-F) The right pulmonary artery measured as 17.0 mm and tracheal carina located at the site of T6 thoracic vertebra and both main bronchus and esophagus were compressed by the left pulmonary artery (10.1 mm) (red and white arrows).
as a common respiratory infection or allergic diseases, including asthma [6]. Etiologically, the process of PAS is induced and advanced under a multifactorial mechanism, and there is an association between PAS and trisomy 18 and 21. Similarly, a case report stated there is a potential association between PAS and 18q del syndrome [7].

Elective surgical repair is considered the optimal strategy for alleviating clinical symptoms, correcting anatomical abnormalities, and improving quality of life in PAS patients. It is important to note that, besides PAS repair, addressing other complex cardiac vascular anomalies is important for prognostic assessment. In our case, although diagnosed as Type II PAS, a relative simpler lesion without any other complex malformations and severe tracheal stenosis from preoperative imaging and moderate or mild clinical symptoms were positively associated with a satisfactory outcome. Preoperatively, for this case, 3D-CT reconstruction was used to assess the airway stenosis instead of conventional bronchoscopy due to the history of intolerance to endoscopic procedure. The compression caused by left pulmonary artery was also observed and assessed objectively, including stenosis degree, which mostly agrees with the literature [8,9]. CBP without cardiac arrest under moderate hypothermia can help protect organ function, decreasing perfusion injury, and maintaining stable vital signs, which is consistent with the literature [10]. However, for PAS patients with complex pulmonary or cardiac malformations, especially complicated with tracheal stenosis, a reasonable and well-prepared surgical strategy is associated with long-term improvement. Chenet et al [11] indicated that slide tracheoplasty should be tailored and individual in accordance with the clinical assessment of infants complicated with congenital tracheal stenosis. For our case, slide tracheoplasty was not the preferred option. Preoperative comprehensive individual assessment plays an important role in selection of surgical strategy. To some extent, the procedures for both CPB and mechanical ventilation are prolonged with the aim to complete extra-slide tracheoplasty [12]. Moreover, underdevelopment of the right lung is negatively associated with the effect of tracheoplasty, which means tracheoplasty is more difficult and challenging due in patients with right-lung hypoplasia [13]. Furthermore, it has been found Type II PAS is associated with higher incidences of postoperative tracheomalacia and recurrent stenosis, which means that long-term stenosis after surgery may not be avoided even when slide tracheoplasty is implemented [14].

Conclusions

We report our center’s first case of PAS, emphasizing the importance of pulmonary artery sling diagnosis, imaging, and surgical planning, and the role of a multidisciplinary team in preoperative and postoperative patient management, as well as improving the local medical system’s ability to treat rare congenital heart diseases, with the aim to serve more pediatric patients with congenital diseases.

Declaration of Figures’ Authenticity

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References: