Pulmonary artery sling: An uncommon cause of stridor

Filipa Neiva^{a,*}, Maria José Oliveria^a, Marta João Silva^a, Milagros Garcia^a, Augusto Ribeiro^a, Aida Sá^b, Bernhard Frey^c and René Prêtre^d

^a*Pediatric Intensive Care Unit, Centro Hospitalar de São João, Porto, Portugal*

^b*Pediatric Department, Centro Hospitalar de Trás-os-Montes e Alto Douro, Vila Real, Portugal*

^c*Department of Intensive Care and Neonatology, University Children's Hospital, Zurich, Switzerland*

^d*Department of Congenital Cardiovascular Surgery, University Children's Hospital, Zurich, Switzerland*

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Abstract. Pulmonary artery sling is a rare congenital condition often associated with congenital tracheal stenosis. Untreated pulmonary sling carries a high morbidity and mortality, most of which is due to the airway and other associated anomalies rather than the aberrant artery itself. We report the case of an infant presenting progressive respiratory distress after a viral infection. The workup revealed a left pulmonary sling with concomitant tracheal stenosis. This anomaly was successfully corrected by slide tracheoplasty and left pulmonary artery reinsertion. *Conclusion* pulmonary artery *sling* is a rare entity with a nonspecific clinical presentation, a high degree of clinical suspicion is needed in order to obtain the correct diagnosis.

Keywords: Pulmonary artery sling, stridor, respiratory failure, slide tracheoplasty

1. Introduction

Pulmonary artery sling is a rare congenital condition in which the left pulmonary artery (LPA) has its origin extrapericardially from the posterior aspect of the right pulmonary artery. The LPA passes leftward around the right bronchus and between the lower trachea and esophagus, thereby indenting these structures, and forming a partial sling around the trachea [1,2]. A patent ductus arteriosus (PDA) or ductal ligament may contribute to encirclement of the trachea [2]. A genetic influence on the LPA sling anomaly is apparent by its occurrence in identical twins and trisomies 18 and 21 [2]. Recently an association with Mowat-Wilson syndrome was described with a mutation in gene ZFX1B of chromosome 2 [3].

Two types of LPA sling are recognized. In type I, the LPA sling is located just above the carina adjacent to the distal trachea and proximal right main stem bronchus, is associated with bronchomalacia and generally resolves with the LPA reanastomosis. The type II is more common and frequently associated with long segment tracheal stenosis. The surgical approach of this entity involves the resolution of the tracheal stenosis along with the LPA reanastomosis [2].

Pulmonary sling frequently presents with respiratory symptoms (up to 90%) in the first months of life due to tracheal stenosis (chronic stridor and wheezing) [1,4], but the pulmonary sling could remain unrecognized until late childhood or even adult life and be misdiagnosed as asthma [5]. The beginning of the symptoms, as well as the clinical severity, is highly dependent on the airway stenosis degree and may be precipitated by an acute respiratory infection [1,2]. The presentation may be catastrophic due to acute airway obstruction

*Corresponding author: Filipa Neiva, Alameda Prof. Hernâni Monteiro, 4200-319 Porto, Portugal. Tel.: +351 9 6526 4804; E-mail: afneiva@gmail.com.

often triggered by physical activity or acute infections and non-recognition of these symptoms can lead to sudden death in newborns and infants [1,5]. Untreated pulmonary sling carries a high morbidity and mortality, most of which is due to the airway and other associated anomalies. Symptoms of airway obstruction are secondary to both impingement of the aberrant pulmonary artery on the tracheobronchial tree as well as to concomitant long-segment congenital tracheal stenosis [3,6]. Feeding difficulty without explicit respiratory symptoms may be the presenting symptom. Pulmonary sling can occasionally be found in a less symptomatic or even asymptomatic older child or adult [2]. About half of patients have other associated congenital defects; heart defects are the most frequently described (30–50%): persistent left superior vena cava, septal defects and PDA [4].

Despite all the surgical advances LPA sling associated with tracheal stenosis continues to be a challenge to the pediatrician and the cardiothoracic surgeon [1].

We report a case of pulmonary artery sling detected in the workup of an infant with severe respiratory distress by a CT pulmonary angiogram.

2. Case history

A 2 months-old male infant was admitted to the Pediatric Intensive Care Unit (PICU) with severe respiratory distress with respiratory failure.

He was born in Switzerland (country where the parents had emigrated). He is the first child of a young couple, non-consanguineous, without relevant family antecedents. He was delivered by caesarean section at 39 wk of gestation with birth weight of 3135 grams and Apgar score of 8/9/9 at one, 5 and 10 minutes of life, respectively. Stridor was noted at the end of his first month of life.

The family was in Portugal for holiday and five days before being admitted to PICU he was hospitalized in the *Centro Hospitalar de Trás-os-Montes e Alto Douro* with complaints of fever, cough, nasal obstruction, and dyspnea. In the initial clinical examination stridor and signs of moderate respiratory distress were described. There were no significant changes on chest radiograph and human metapneumovirus was isolated in respiratory secretions. After an initial period of clinical improvement, a progressively increasing work of breathing was noted with hypoxemia and need for non invasive ventilation with nasal continuous positive airway pressure. Because of severe respiratory distress with acidosis

and eventual respiratory failure (fraction of inspired oxygen- $\text{FiO}_2 = 1$: pH 7.3, PaCO_2 73 mmHg; PaO_2 329 mmHg; HCO_3 29,3 mmol/L, BE 1 mmol/L) he was intubated and placed on invasive mechanical ventilation (IMV) and transferred to the PICU in *Centro Hospitalar de S. João*.

On PICU admission he was sedated with midazolam and fentanyl continuous infusion, and put on IMV [peak airway pressure (Ppeak) 40 cmH_2O ; positive end-expiratory pressure 5 cmH_2O]. He was afebrile. Clinical assessment revealed an infant with normal thoracic expansion, crackles and wheeze on pulmonary exam, liver palpable 2 cm below right costal margin. He was hemodynamically stable, with no inotropic support (Mean arterial pressure P5-50). Chest radiograph was normal (Fig. 1). Progressive respiratory instability with short periods of marked hypoventilation was observed and elevated Ppeak (maximum 55 cmH_2O ; P plateau 29 cmH_2O) was applied. Arterial blood gas: pH 7.22; pO_2 89,9 mmHg; pCO_2 77 mmHg; HCO_3 31 mmol/L (FiO_2 0,6). Persistence of human metapneumovirus in respiratory secretions.

A thoracic computerized tomography on the third PICU day described a stenosis of the lower third of the trachea, with reduction of 70% of the tracheal diameter and an extension of 2.5 cm. There were no areas of extrinsic compression. Alveolar consolidation of the right lower lobe and to a lesser extent the left lobe, suggestive of pneumonia, was found (Fig. 2).

The study was completed on the fourth day with a computerized tomography (CT) angiogram, which showed LPA sling. LPA has its origin in right pulmonary

Fig. 1. Normal X-ray.

Fig. 2. CT angiogram – abnormal LPA origin from RPA.

Fig. 4. Traqueal stenosis on CT angiogram.

Fig. 3. CT angiogram – abnormal LPA origin from RPA.

Fig. 5. Echocardiogram – abnormal LPA origin.

artery, then follows its course between the esophagus and the trachea. The tracheal stenosis was described above this malformation; complete tracheal rings could not be excluded. (Fig. 3 and 4)

Echocardiography on the same day confirmed the LPA anomalous origin and described normal intracardiac anatomy. (Fig. 5)

Given the impossibility of resolution of tracheal stenosis along with LPA correction in one surgical session, the patient was transferred to the pediatric surgical center in Zurich (Switzerland) on the seventh PICU day.

Shortly after arrival in Zurich, the infant was again in severe respiratory failure and therefore he was operated even on the same day on cardio-pulmonary bypass: tracheal repair by slide tracheoplasty, LPA reinsertion

and PDA ligation. In the first post-operative night he developed a tension pneumothorax which was drained by chest tube. A tracheoscopy 5 days after the operation showed an open distal trachea. He was on IMV for a week after surgery, and needed oxygen supplementation until day 9 post-operation. Post-procedure serial echocardiography was performed. By the 25th post-operative day echocardiogram was normal (no pulmonary hypertension); the assessment of the LPA anastomosis was difficult but it showed normal flow without acceleration. A normal venous return in the pulmonary left veins was described.

He was discharged home on 27th post-operative day with normal breathing. Actually with four months

of age, there is no stridor or any other sign of respiratory difficulty. The patient maintains regular visits to pediatric cardiology, and the pediatrician.

3. Discussion

Pulmonary artery *sling* (PAS) is a rare entity with critical prognosis when it is not diagnosed in time. There are few studies about the true prevalence of PAS in part because the asymptomatic remain undiagnosed, nevertheless there is an increase in case report and small series of vascular anomalies and PAS in older children and adults in recent years, suggesting that this group of abnormalities are likely to be under-diagnosed. A study conducted in Taiwan revealed a prevalence of PAS of about 59 per million school-aged children [5]. Because it has a nonspecific clinical presentation (which is common to other diseases causing airway obstruction), a high degree of clinical suspicion is needed in order to obtain the correct diagnosis. In the current clinical case, stridor started about a month before the hospitalization with sudden deterioration coinciding with a viral infection, as it is described in the literature as the most frequent presentation form [1,2,7]. Some data, such as the high Ppeak with normal Pplateau, suggested an airway obstruction to be more likely than a serious pulmonary disease. Imaging studies were of extreme importance in obtaining the diagnosis.

The procedure of choice to establish the diagnosis of pulmonary artery sling is echocardiography [2,7], which is simple and comfortable for the patient, but has low sensitivity and it has limitations in defining the aortic arch anatomy [14]. CT and magnetic resonance are good methods with higher sensitivity and should be used when there is a high clinical suspicion of malformation and normal echocardiography. They allow the diagnosis of most vascular malformations also allowing airway assessment, particularly the tracheal diameter. 3D image reconstruction allows a more effective evaluation of tracheal stenosis extension, also making the planning of the surgical approach easier. CT, contrary to magnetic resonance, has the advantage of much better visualization of lung parenchyma and airways, higher spatial resolution, and faster scanning with lower requirement for sedation or anesthesia. For that reason CT angiography is the best technique for evaluating PAS complete anatomy [8].

Although asymptomatic patients with a type I sling can be followed clinically [8], the treatment of aberrant LPA is surgical and in most cases must be

accomplished as soon as the diagnosis is established to avoid the possibility of death by hypoventilation [5,9–11]. In the current case, given the severity of respiratory distress with increasing difficulties in effective ventilation, the immediate surgical resolution of the malformation was mandatory. The cooperation between the two hospitals (Centro Hospitalar de S. João, Porto and University Children's Hospital, Zurich) was an example, allowing the patient rapid transfer to the surgical center and resolution of the malformation in time.

The crucial step in preoperative workup is to identify complete tracheal rings (because this implies the need for tracheoplasty under cardiopulmonary bypass, and not just reimplantation of the LPA). Bronchoscopy is still used for this but with modern three-dimensional CT techniques, including virtual bronchoscopy allow a noninvasive study of the trachea [12]. The ideal surgical option remains controversial with debates focusing the ideal management of the LPA itself. The controversy is whether the best is the translocation of the LPA with distal tracheal resection versus reimplantation, especially in type II *sling* where the tracheal repair should be done in the same session [2,5,9,11,13]. There were some reports of complications after translocation such as the possibility of LPA kinking and anterior compression of the trachea by the left pulmonary artery. In addition it was also noted that the translocated LPA could cause anterior compression of the trachea or left main bronchus. The main concern about reimplantation is obstruction of the LPA. However, several reports have described excellent long-term patency of implanted LPA [9]. The reimplantation must be technically perfect in order to maintain artery patency [13].

Slide tracheoplasty is the preferred technique for repair of congenital tracheal stenosis [5,8,11]. This technique, proposed in 1989 by Tsang et al, was used in the present case and has the advantage of limited dissection and of using the trachea itself for the repair, so the airway is lined by normal mucosa and can grow with the child [10]. The excellent post-operative result of our patient supports the use of slide tracheoplasty and LPA reimplantation as the surgery of choice for Type II LPA sling. Post-operative outcome depends on the associated malformations, previous clinical state of the patient, as well as the experience of the surgical team involved [10]. These patients require close cooperation between the two surgical specialties (pediatric and cardiothoracic), as well as their medical counterparts [2].

The authors present this case for its rarity, highlighting the seriousness of the clinical presentation and the

collaboration between the two units of different countries in the resolution of the case. We feel that this anomaly should be included in the differential diagnosis of airway obstruction of the newborn/infant despite its relative rarity. The early execution of imaging exams allowed a prompt diagnosis and surgical repair.

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