Case Report

A Rare Cause of Wheezing in a Child with Pulmonary Atresia

Abstract

The determination of the exact cause for symptomatic airway obstruction in pediatric patients not responding to medication can be a clinical dilemma. Very rarely external vascular compressions can produce airway obstruction symptoms unresponsive to usual bronchodilator medications. The successful management of a child with pulmonary atresia and an innominate artery compression syndrome with respiratory compromise due to tracheal compression is described.

Keywords: *Airway obstruction/etiology, brachiocephalic trunk/abnormalities, bronchoscopy, tomography, tracheal stenosis/etiology, X-ray computed*

Madan Mohan Maddali, Pranav Subbaraya Kandachar[1], Nishant Ram Arora[2], Francois Lacour-Gayet[1]

Departments of Cardiac Anesthesia and [1]Cardiothoracic Surgery, National Heart Center, Royal Hospital, [2]National Heart Center, Royal Hospital, Muscat, Oman

Introduction

In patients with pulmonary atresia with ventricular septal defect (with Tetralogy of Fallot type of morphology) the aorta is often quite enlarged. The aortic dilation is sometimes progressive in these patients, and secondary tracheal compression can occur in the process of innominate artery dilation. We describe a case of innominate artery compression syndrome resulting in wheezing due to distal tracheal compression in a child with pulmonary atresia. To the best of our knowledge, the association between pulmonary atresia with ventricular septal defect and innominate artery compression syndrome with airway compromise has not been described, making this report rather unique. A consent from parents of the child as well as Institutional Scientific Research Committee approval (SRC#CR17/2017) was obtained for publishing this case report.

Case Report

A 14-month-old girl child (weight: 11.3 kg and height: 90 cm) presented to the authors' institute with a history of cyanosis since birth and a wheeze. At 5 days of age, the child was diagnosed to have pulmonary atresia with a large ventricular septal defect by transthoracic echocardiography. An emergency right Modified Blalock–Taussig shunt was performed between the innominate artery and the right pulmonary artery through a median sternotomy as the baby had low arterial oxygen saturations persistently despite prostaglandin E1 infusion. A median sternotomy was preferred as against a thoracotomy as it would avoid any lung retraction that might contribute further toward arterial oxygen desaturation. The pulmonary arteries are usually larger in their central part, and the surgeon would have the option of performing the shunt on cardiopulmonary bypass in case the baby became further unstable. Finally, a median sternotomy approach would give the surgeon an opportunity to ligate the ductus arteriosus after performing the shunt. For the Modified Blalock–Taussig shunt through the median sternotomy, the right innominate artery was opted instead of the subclavian artery as it is the most accessible artery among the arch vessels, and it is usually considered safe to clamp the innominate artery for short periods of time since the left carotid is left to perfuse the brain. During the subsequent definitive operation, taking down an innominate to pulmonary artery shunt would be easier compared to a subclavian to pulmonary artery shunt.

The baby developed a wheeze when she was 7 months old and was initiated at a peripheral health center on ipratropium bromide nebulization (250 μg QID), salbutamol by an inhaler (400 μg/3 hourly), and dexamethasone tablet (4 mg OD) with poor response. The child continued to receive these medications intermittently until she arrived at our center. As it was felt

How to cite this article: Maddali MM, Kandachar PS, Arora NR, Lacour-Gayet F. A rare cause of wheezing in a child with pulmonary atresia. Ann Card Anaesth 2019;22:449-51.

Address for correspondence:
Dr. Madan Mohan Maddali, Department of Cardiac Anesthesia, National Heart Center, Royal Hospital, P. B. No: 1331, P. C: 111, Seeb, Muscat, Oman. E-mail: madanmaddali@hotmail.com

Videos Available on: www.annals.in

Access this article online

Website: www.annals.in

DOI: 10.4103/aca.ACA_92_18

Quick Response Code:

by the pediatric cardiologists that she was outgrowing the shunt and it was time for definitive surgery, a computed tomography (CT) angiography scan of the chest was performed as part of preoperative evaluation for pulmonary artery anatomy. The CT angiography revealed compression of the distal trachea by a rather large innominate artery arising more posteriorly than usual from the arch of aorta, and forming a tortuosity which was displacing the trachea posteriorly and compressing the distal trachea from its right anterolateral aspect [Figure 1].

The child underwent a Rastelli operation along with right brachiocephalic arteriopexy. At the time of intracardiac repair, an intraoperative bronchoscopy was performed, and distal trachea was visualized [Figure 2 and Video 1]. The arteriopexy was monitored by intraoperative bronchoscopy which revealed relief of pulsatile obstruction of the distal trachea as the brachiocephalic artery was retracted away from the trachea and fixed to the upper end of the right blade of sternum [Figure 3 and Video 2]. The dynamic lung compliance before arteriopexy was 7.5 mL/cm H_2O, and following arteriopexy in the postoperative period was 4.9 mL/cm H_2O [Figure 4]. Tracheal extubation was performed after elective mechanical ventilation of 24 h.

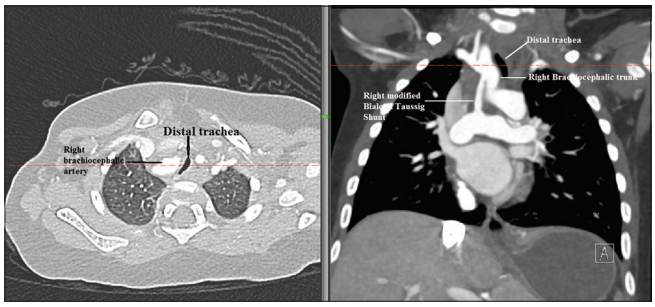

Figure 1: A computerized tomography angiogram of chest displaying both the right brachiocephalic artery and the distal trachea in the transverse plane in the left half of the image and in the coronal plane in the right half

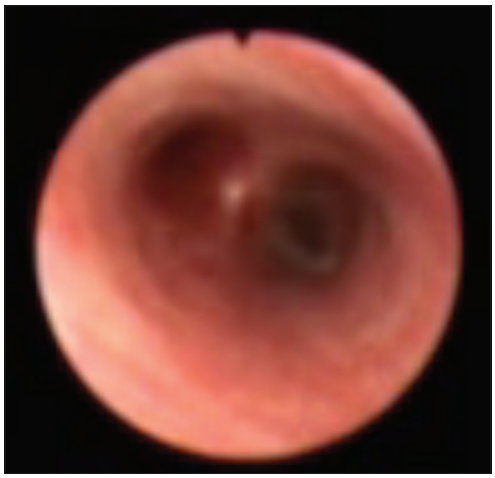

Figure 3: Intraoperative bronchoscopy depicting the distal trachea with its patency restored after a right brachiocephalic arteriopexy

Discussion

The successful surgical management of a child with pulmonary atresia and innominate artery compression syndrome that presented as a continuous, coarse, whistling sound during breathing (wheezing) not responding to medical treatment is presented.

Innominate artery compression syndrome with respiratory compromise due to tracheal compression by the brachiocephalic artery that was attributed to an anomalous origin of the artery to the left of the trachea and producing tracheal compression while crossing it was described earlier.[1] In patients with lesser degrees of displacement of the origin of the artery, it has been noted that the left common carotid artery arises very close to the brachiocephalic trunk and the trachea gets trapped in a vice. Rarely, anterior mediastinal masses can displace the innominate artery resulting in vascular compression of the trachea.[2]

The management options include reimplantation of the innominate artery more proximally on the aorta thereby eliminating the anatomic cause of tracheal compression.[3]

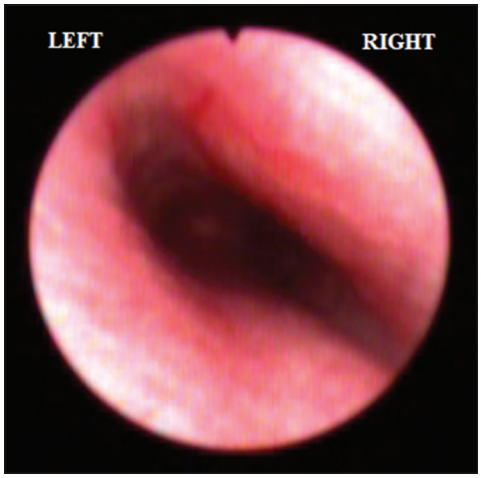

Figure 2: Intraoperative bronchoscopy depicting the distal trachea

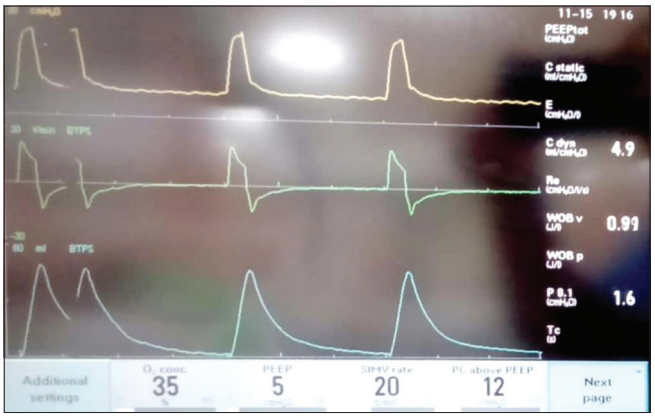

Figure 4: Postoperative image of the front panel of the ventilator displaying a dynamic compliance of 4.9 mL/cm H_2O

The other option is innominate arteriopexy or by aortopexy wherein, the innominate artery is mobilized and fixed to the sternum.[4] Backer *et al*. have suggested that the indication of innominate arteriopexy is when there is more than 80% compression of the tracheal lumen as confirmed by bronchoscopy.[5]

The anesthesiologists can contribute toward the management of these cases by performing intraoperative bronchoscopy and by invasive arterial pressure monitoring of the right upper limb. The role of intraoperative bronchoscopy during surgical correction of bronchial compression has been described earlier in this journal.[6] Intraprocedural bronchoscopy is useful during specific interventional cardiology procedures as well, to avoid airway complications.[7,8] Backer *et al*. highlighted the importance of intraoperative bronchoscopy to confirm the success of the innominate arteriopexy operation as well as suggested monitoring of the right radial artery pulse to identify and prevent stenosis of the innominate artery by the suspension maneuver.[5] In this baby, intraoperative bronchoscopy during brachiocephalic arteriopexy ensured that the external vascular compression was removed restoring an unobstructed airway. The right radial artery was cannulated, and pressure was monitored continuously perioperatively in accordance with Backer *et al*. suggestions.

Conclusion

Mechanical airway obstruction may masquerade as reactive airway disease in children and the astute clinician with a high index of suspicion can make the diagnosis and thus, a difference.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the legal guardian has given his consent for images and other clinical information to be reported in the journal. The guardian understands that names and initials will not be published and due efforts will be made to conceal patient identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Fawcett SL, Gomez AC, Hughes JA, Set P. Anatomical variation in the position of the brachiocephalic trunk (innominate artery) with respect to the trachea: A computed tomography-based study and literature review of innominate artery compression syndrome. Clin Anat 2010;23:61-9.
2. Hennington MH, Detterbeck FC, Molina PL, Wood RE. Innominate artery and tracheal compression due to aberrant position of the thymus. Ann Thorac Surg 1995;59:526-8.
3. Hawkins JA, Bailey WW, Clark SM. Innominate artery compression of the trachea. Treatment by reimplantation of the innominate artery. J Thorac Cardiovasc Surg 1992;103:678-82.
4. Gardella C, Girosi D, Rossi GA, Silvestri M, Tomà P, Bava G, *et al.* Tracheal compression by aberrant innominate artery: Clinical presentations in infants and children, indications for surgical correction by aortopexy, and short- and long-term outcome. J Pediatr Surg 2010;45:564-73.
5. Backer CL, Mongé MC, Popescu AR, Eltayeb OM, Rastatter JC, Rigsby CK. Vascular rings. Semin Pediatr Surg 2016;25:165-75.
6. Maddali MM, Junghare SW, Nadarajan K, Kandachar PS, Al-Maskari SN, Valliattu J. Left pulmonary artery stent causes ipsilateral pulmonary complication. J Cardiothorac Vasc Anesth 2017;31:395-7.
7. O'Byrne ML, Rome N, Santamaria RW, Hallbergson A, Glatz AC, Dori Y. Intra-procedural bronchoscopy to prevent bronchial compression during pulmonary artery stent angioplasty. Pediatr Cardiol 2016;37:433-41.
8. Grohmann J, Stiller B, Neumann E, Jakob A, Fleck T, Pache G, *et al.* Bronchial compression following pulmonary artery stenting in single ventricle lesions: How to prevent, and how to decompress. Clin Res Cardiol 2016;105:323-31.