

Management of a Case of Double Aortic Arch with Tracheal Compression Complicated with Postoperative Tracheal Restenosis

Abstract

Tracheal stenosis in association with the double aortic arch (DAA) is uncommon; however, it carries a high risk of morbidity, mortality, and restenosis. Although surgery is the mainstay of managing a case of the DAA with tracheal stenosis, management of tracheal restenosis requires a multidisciplinary approach. In this case report, we present our successful experience in managing a child of DAA with tracheal stenosis who developed tracheal restenosis after sliding tracheoplasty of trachea.

Keywords: Double aortic arch, restenosis, tracheal stenosis

Introduction

Vascular rings account for 1%–2% of all cardiovascular malformations. Double aortic arch (DAA) accounts for 40% of vascular rings with both functional arches being the rarest, comprising only 5% of cases.^[1] We report the perioperative management of a case of codominant DAA with tracheal compression with postoperative tracheal restenosis.

Case Report

A 5-month-old child weighing 6 kg was admitted with complaints of dyspnea and stridor since birth. Two-dimensional (2D) transthoracic echocardiography revealed DAA with patent ductus arteriosus (PDA). Multidetector computed tomography (CT) thorax revealed evidence of DAA [Figure 1] forming a complete vascular ring around trachea and esophagus with resultant concentric narrowing of trachea. Right subclavian and common carotid arteries arise from the right aortic arch, and left subclavian and common carotid arteries arise from the left arch with the descending thoracic aorta on the right side. High-resolution CT (HRCT) thorax revealed tracheal narrowing with prestenotic segment measuring 6.6 mm and the narrowest segment measuring 2.5 mm which was 5 mm proximal to the carina.

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Intraoperative management

The patient was induced and intubated as per the institutional protocols with intravenous narcotics and inhalational agents. The patient was operated on moderate hypothermic cardiopulmonary bypass (CPB). PDA was ligated and divided. The left aortic arch was divided distal to left subclavian artery, and ends were closed. Trachea was divided at the stenotic site, tracheomalacia portion with complete tracheal ring excised and slide tracheoplasty was done using interrupted 6-0 Maxon sutures. Postoperatively, the patient was electively ventilated for 72 h and then extubated. The patient was managed conservatively with intravenous steroids, nasal continuous positive airway pressure, prone position, chest physiotherapy, and humidified oxygen for 8 days.

The patient developed stridor from the 10th postoperative day (POD) which was managed conservatively. In view of respiratory distress, the child was intubated on the 12th POD and rigid bronchoscopy performed which revealed a slit-like opening of tracheal lumen. HRCT thorax revealed narrowest segment of the trachea measured 2.3 mm [Figure 2]. On the 15th POD, balloon dilatation of trachea with local injection of triamcinolone was performed in the hybrid catheterization

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laboratory [Figure 3] and the endotracheal tube passed distal to the site of stenosis. The patient was extubated after 72 h but was reintubated due to persistent stridor and respiratory distress.

In view of failure to wean from mechanical ventilation, tracheal stenting was performed in the catheterization laboratory on the 33rd POD and electively ventilated for 48 h. The patient was extubated on the 37th POD and managed conservatively later on.

The patient was managed for 8 days in step-down Intensive Care Unit (ICU) and discharged on the 51st POD with minimal stridor.

Discussion

Respiratory symptoms account for 91% of presentations of DAA with stridor being the most common presentation (77%), as in this child.^[2] Although DAA carries a good prognosis, associated tracheal anomalies make the prognosis guarded. Hoffer *et al.* accounted that tracheal stenosis with pulmonary or cardiovascular malformations was associated with a 79% mortality.^[3]

Hence, all patients with DAA should be evaluated for associated airway anomalies. Among several methods that have been validated, high-resolution CT with 3D-reconstruction is the modality of choice to diagnose and evaluate the underlying vascular and tracheobronchial anatomy, with the advantage of rapidity needed in children. Bronchoscopy and cardiac catheterization are used to evaluate the functional component of the compression.

The usual approach for repair of DAA is through a thoracotomy; however, midline sternotomy is preferred in patients with associated tracheal stenosis, as surgery requires commencement on CPB.

For management of associated tracheal stenosis, length of the diseased trachea dictates the type of reconstruction. Slide tracheoplasty was preferred for tracheoplasty as it gives better outcome in surgical repair with minimal incidence of tracheal restenosis.^[4] In slide tracheoplasty, native tracheal tissue is used to widen the lumen, giving stability, minimizing granulation tissue formation and providing a complete epithelial surface. Grillo^[5] had shown patent airways and postoperative tracheal growth on long-term follow-up.

Elliott *et al.* reported a mortality of 12.5% and Rutter *et al.*^[6] reported a mortality of 18%, ranging from 0% to 25% after slide tracheoplasty.

Techniques such as patch tracheoplasty and various autografts have been used for long segment narrowing of the trachea which however carried an increased incidence of complications.

Postoperative management of patients with tracheal stenosis is challenging as the rate of complications is high after open tracheoplasty. The common denominator of

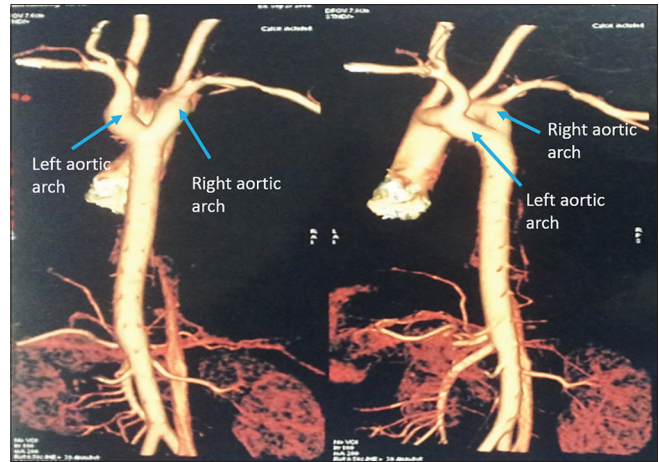


Figure 1: Multidetector computed tomography volume rendered image depicting double aortic arch forming a complete vascular ring

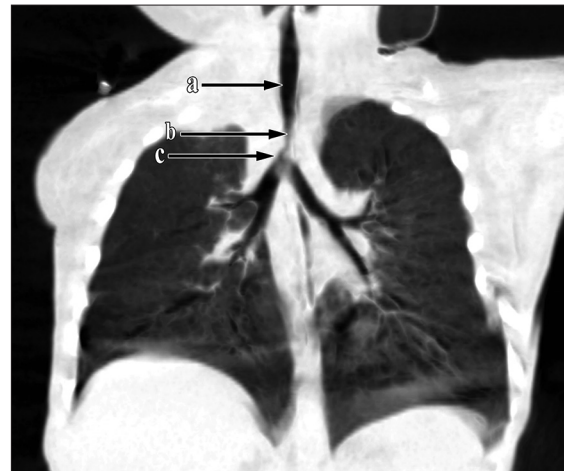


Figure 2: Maximum intensity projection coronal image showing bronchial tree with normal prestenotic segment measuring 6.5 mm (a), stenotic segment measuring 2.3 mm (b) which is 5 mm from the carina (c)

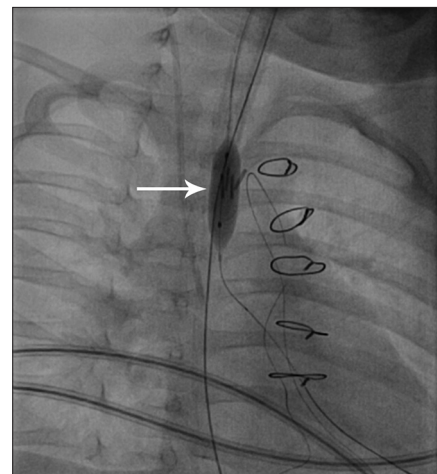


Figure 3: Catheterization image showing balloon dilation of the trachea (white arrow)

all postoperative morbidity is the formation of excessive granulation tissue at the anastomotic line between the

tracheal ends (from exposed sutures) or native trachea and graft materials which was noted in our case also.

Around 14% patients operated for DAA with tracheal stenosis developed tracheal restenosis postoperatively.^[2] The most common presenting symptom is stridor while other symptoms include respiratory distress and persistent lower respiratory infections.

HRCT thorax plays an invaluable role in the diagnosis and management of postoperative tracheal restenosis.

Medical management is the mainstay treatment for tracheal restenosis which includes intravenous steroids, antireflux treatment, antibiotics, chest physiotherapy, and humidified oxygen and helium-oxygen mixture administration. In adjunct, topical steroids (as in our case) or mitomycin C can be used.

In patients with failed medical management, balloon dilation of the stenotic trachea can be performed. Balloon dilation localizes the distending forces in an outward radial direction with minimal trauma and can be moved to multiple positions.^[7]

Patients not responding to tracheal dilation may require intraluminal stents. Stents such as the Dumon silastic stent and the metallic Palmaz stent have gained recent popularity. Absorbable stents are currently under trial and may prove a valuable future asset. Both balloon dilation technique and Dumon silastic stent were used in our case.

Reoperation is the final resort if the child has persistent stridor despite all the medical and interventional measures, but it carries a very high morbidity and mortality.

Current research is being undertaken on reduction of granulation tissue formation which includes vascular endothelial growth factor and tissue engineering to grow trachea from patients own cells.^[8]

Despite the best medical and surgical management, these patients have significant morbidity, as demonstrated by their prolonged intubation times, multiple bronchoscopies, stenting, reoperation, prolonged intensive care stay, and in-hospital stay. Our patient was managed for 38 days in the ICU and was discharged on the 51st POD with minimal stridor.

A committed multidisciplinary approach can optimize the airway and ensure the most favorable outcome for a child with such an anomaly.^[7]

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Conflicts of interest

There are no conflicts of interest.

References

1. Backer CL, Mavroudis C, Rigsby CK, Holinger LD. Trends in vascular ring surgery. *J Thorac Cardiovasc Surg* 2005;129:1339-47.
2. Alsenaidi K, Gurofsky R, Karamlou T, Williams WG, McCrindle BW. Management and outcomes of double aortic arch in 81 patients. *Pediatrics* 2006;118:e1336-41.
3. Hoffer ME, Tom LW, Wetmore RF, Handler SD, Potsic WP. Congenital tracheal stenosis. The otolaryngologist's perspective. *Arch Otolaryngol Head Neck Surg* 1994;120:449-53.
4. Cotter CS, Jones DT, Nuss RC, Jonas R. Management of distal tracheal stenosis. *Arch Otolaryngol Head Neck Surg* 1999;125:325-8.
5. Grillo HC. Slide tracheoplasty for long-segment congenital tracheal stenosis. *Ann Thorac Surg* 1994;58:613-9.
6. Rutter MJ, Cotton RT, Azizkhan RG, Manning PB. Slide tracheoplasty for the management of complete tracheal rings. *J Pediatr Surg* 2003;38:928-34.
7. Gastón BM, Marcelo MF. *Pediatric Tracheal Stenosis and Vascular Rings*. Autonomous City of Buenos Aires, Argentina: Hospital Foundation, Private Children's Hospital; 2012. p. 206-19.
8. Nissen NN, Polverini PJ, Koch AE, Volin MV, Gamelli RL, DiPietro LA. Vascular endothelial growth factor mediates angiogenic activity during the proliferative phase of wound healing. *Am J Pathol* 1998;152:1445-52.