Case Report

Double aortic arch presenting with respiratory distress: A case report and review of the literature

ABSTRACT

Tracheal compression by vascular structures in infants is uncommon and may be masked by nonspecific respiratory symptoms. Double aortic arch (DAA) is the most common vascular ring. We describe a case of a 9-month-old male infant presented with respiratory distress and found to have a DAA. In this report, the authors emphasize the consideration of this pathology-induced respiratory distress and discuss its anesthetic management.

Key words: Airway compression; anesthesia management; double aortic arch; infant

Introduction

Double aortic arch (DAA) is a rare congenital vascular malformation in which persistence of two aortic arches forms a complete vascular ring that can compress the trachea and/or esophagus. Early diagnosis is difficult because it has a wide clinical spectrum.[1] We present a rare case of a 9-month-old male infant with DAA who presented with respiratory distress. Through this case report, the authors emphasize the consideration of this pathology-induced respiratory distress and discuss its anesthetic management.

Case Report

A 9-month-old full-term male infant was admitted to our neonatal intensive care unit for the management of respiratory distress. He had a history of permanent stridor. At presentation, the infant was moderately dehydrated and was in severe respiratory distress: tracheal tugging, marked intercostal and subcostal recessions, and was cyanosed. His respiratory rate was 50 cycles/min; SpO_2 was 50% at room air improved to 70%–80% on supplemental O_2 with mask. The cardiovascular findings were unremarkable. The chest X-ray showed hyperinflated lung fields bilaterally. Then, the patient was intubated with a 3.5 mm endotracheal cuffed tube and ventilated in controlled pressure mode. Fiber-optic bronchoscopy examination revealed tracheal narrowing 8 mm just above the carina. He was planned for computerized tomography (CT) scan detecting the presence of vascular ring surrounding the trachea [Figure 1]. Reconstruction of CT arteriography showed both right and left aortic arches around the trachea [Figure 2]. The right side arch was bigger and dominant. Surgical indication was retained. On admission to the theater, venous access was secured with peripheral venous line. General anesthesia was induced with fentanyl (5 μg/kg) and propofol (3 mg/kg). After verification with bronchoscopy of correct placement

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of tip, a dose of 20 mg of rocuronium was administered. Then, a central right jugular internal venous with a left radial arterial line was set up. Anesthetic was maintained by sevoflurane (2%–4%). Surgical approach was performed by left posterolateral thoracotomy [Figure 3]. After surgery, the symptoms improved strikingly with regression of stridor. He was discharged on the postoperative 6th day.

Discussion

Vascular ring is reported to account for <1% of congenital heart disease. DAA is the most common form of it. The first anatomical case of a DAA was described in 1737 by Hommel, and the first surgical correction was performed in 1945 by Robert Gross.[2] Classically, DAA has three types: right dominant aortic arch (75%), left dominant aortic arch (20%), and balanced type (5%).[3]

Clinical symptoms of DAA usually appear in the first 6 months of life. Compression from complete anomalies leads to respiratory and gastrointestinal symptoms [Table 1]. DAA usually causes respiratory symptoms in 91% and gastrointestinal symptoms in 40% of cases.[3] The respiratory symptoms include stridor, choking, episodes, and recurrent respiratory infections. Respiratory arrest has been reported in 8% of reported cases.[4] In this observation, tracheal compression was revealed by respiratory distress in a 9-month-old infant.

Diagnostic methods of DAA have evolved from chest radiography, esophagocardiogram, echocardiography to CT scan, and magnetic resonance imaging (MRI). Flexible bronchoscopy would be the first line of investigation for the patients typically present with respiratory symptoms. Typical compression of the trachea was visualized by bronchoscopy which was indicated for respiratory. Nature of extramural obstruction would be best shown by spiral CT scan, MRI, or color-coded three-dimensional[5] It showed in our case an external compression on the trachea.

DAA is usually found as an isolated cardiovascular malformation. Associated cardiac anomalies occurred only in eight patients (7%) in a large study of 113 patients with DAA[3] and appeared in 14 patients (17%) in another study of 81 patients with DAA.[6] In our patient, no associated cardiac abnormality was noted.

Airway management in patients with DAA is challenging for anesthesiologists. The preoperative condition of these patients is often serious. They must frequently be accepted for anesthesia with severe respiratory infection still present which are a source of aggravation of chronic airway obstruction through edema and inflammation. The evaluation should also include research into bronchial fibroscopy (FB) of any associated tracheal lesions such as

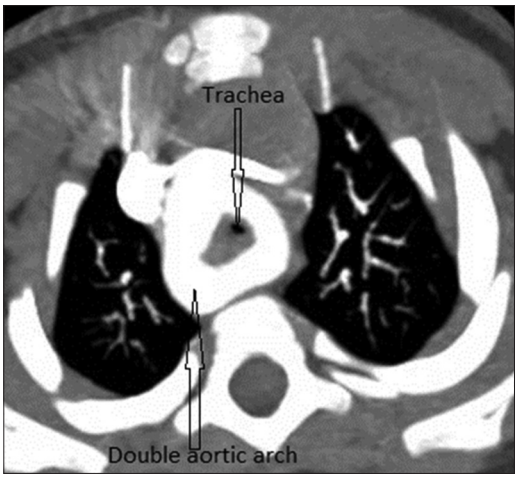

Figure 1: Contrast-enhanced computerized tomography axial section image showing the double aortic arch

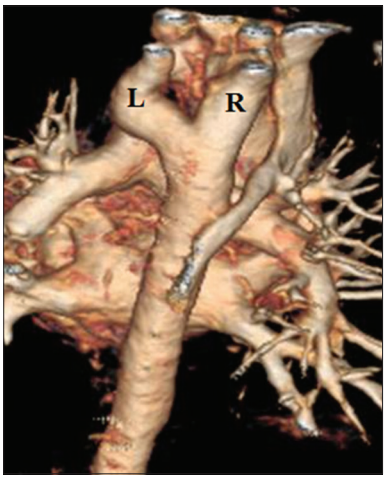

Figure 2: Computerized tomography reconstruction arteriography posterior view showing the right (R) and left (L) aortic arch making a vascular ring

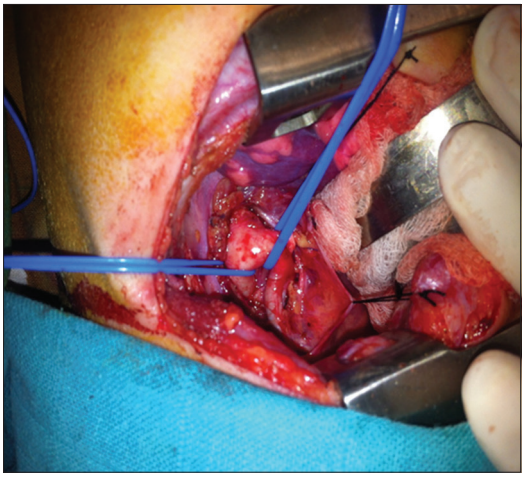

Figure 3: Intraoperative photo with individualization of the descending aorta and the left arch

Table 1: A contemporary review of the literature regarding double aortic arch

	DAA		
	Backer *et al.*[3] (*n*=113)	Alsenaidi *et al.*[4] (*n*=81)	Ruzmetov *et al.*[6] (*n*=67)
Patient symptoms, *n* (%)			
Stridor	46 (57)	62 (77)	43 (64)
Recurrent respiratory infection	22 (27)	8 (10)	28 (42)
Coughing	17 (21)	28 (35)	-
Dysphagia	12 (15)	4 (5)	11 (16)
Respiratory distress	8 (10)	6 (7)	-
Associated cardiac defect, *n* (%)	8 (7)	14 (17)	7 (11)
Dominant arch, *n* (%)			
Right	85 (75)	56 (72)	55 (82)
Left	20 (18)	16 (20)	12 (18)
Balanced	8 (7)	7 (9)	-

DAA: Double aortic arch

tracheobronchomalacia and assess the degree of stenosis to identify patients at risk for respiratory failure who may present difficulties in ventilation.[7]

The site of tracheal obstruction, when present, is important to the anesthetist. If situated just above the carina, placing the tip of the endotracheal tube (ETT) distal to the obstruction may mean using one-lung anesthesia only. A selection of ETTs of different sizes and lengths must be carefully assembled. FB confirmation of ETT tip position is helpful, and ETT should be secured firmly by adhesive plaster or thread. Induction of anesthesia must be performed without curare until the airway is secured. Blood loss may be sudden and severe and must be immediately replaced. The infant's line arterial and a central venous should be placed.[7]

In the postoperative period, airway obstruction may persist in approximately 30% of cases despite surgical correction, requiring artificial ventilation, or continuous positive airway pressure ventilation.[8]

Conclusion

DAA may cause persistent nonspecific complaints to life-threatening respiratory distress in infants. In these patients, the airway is precarious and complete cooperation between anesthetist and surgeon is essential.

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Conflicts of interest

There are no conflicts of interest.

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