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SHORT REPORT

Anomalous ascending aorta causing severe compression of the left bronchus in an infant with ventricular septal defect and pulmonary atresia

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Abstract Tracheobronchial compression by cardiovascular structures complicates the course after surgery of congenital heart disease. A 2-month-old boy with ventricular septal defect, pulmonary atresia, and velocardiofacial syndrome had severe left main bronchus obstruction due to external compression by an anomalously oriented ascending aorta. The patient's hypoxemic episodes necessitated extracorporeal membrane oxygenation. We inserted a stent in the left bronchus to open the airway, but the stent was crushed by the anomalous aorta. We later surgically rerouted the aorta and finally restored the patency of the left main bronchus. However, the patient died of fungemia, without being weaned from extracorporeal membrane oxygenation. We conclude that surgery is necessary instead of stent implantation to relieve the external compression of the left bronchus from a vessel with systemic arterial pressure.

Keywords Congenital heart disease · Tracheobronchial stenosis · Vascular anomaly

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Introduction

Tracheobronchial compression by cardiovascular structures in childhood is uncommon, but complicates the course after surgery of congenital heart disease [4, 8]. The intimate anatomic relationship between cardiovascular and airway structures plays an important role in this phenomenon [7]. Critical airway stenosis usually requires aggressive intervention, including stenting [9], rerouting the surrounding vessels [3], and tracheoplasty [2, 10]. We report an infant with ventricular septal defect (VSD) and pulmonary atresia (PA), whose left main bronchus was compressed by his anomalously oriented ascending aorta. An airway stent failed to improve his hypoxemia and surgery was required to relieve the left bronchial obstruction.

Case report

A 2-month-old boy presented with cyanosis and tachypnea since birth. Physical examination showed grade III/VI continuous murmurs over the left upper sternal borders and marked wheezing in the left chest. Echocardiography revealed VSD with PA, major aortopulmonary collateral arteries (MAPCAs), and a right aortic arch. The aortic arch arose from the posterior part of the left ventricle (LV). Electrocardiography-gated computed tomography (CT) confirmed the diagnosis and showed the anomalous ascending aorta traversing just above the left main bronchus. As a result, the anomalous aorta compressed the left main bronchus (Fig. 1). The patient also had velocardiofacial syndrome. He underwent right-side modified Blalock-Taussig shunt placement and ligation of the MAPCAs at 24 days of age.

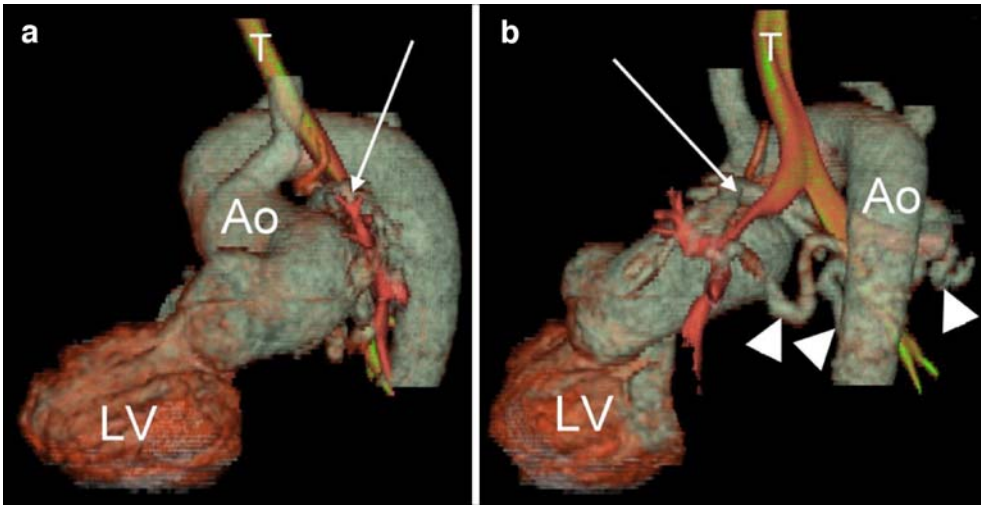

Fig. 1 Multiple tissue segmentation computed tomography (CT) with three-dimensional volume rendering of the left ventricle (*LV*), aorta (*Ao*), pulmonary artery (*arrow*), major aortopulmonary collateral artery (*arrowheads*), and the tracheobronchial tree (*T*). (**A**) Left lateral and (**B**) left posterior oblique views show the redundant sigmoid course of the ascending aorta, with a posteriorly displaced aortic root that pushed the hypoplastic ventral pulmonary artery backwards and compressed the proximal portion of the left main stem bronchus

After surgery, he encountered difficulty weaning from the ventilator. High-pressure ventilator support was required, but hypercapnia ensued frequently. Chest radiography showed poor aeration of the left lung and right upper lung field (Fig. 2). Bronchoscopy confirmed that the left bronchus was significantly narrowed by a pulsatile mass. We assumed that this was the cause of failure to wean from the ventilator. Progressive hypoxemia and bradycardia ensued 1 month after surgery and extracorporeal membrane oxygenation (ECMO) was instituted during cardiopulmonary resuscitation. We then put a Palmaz stent (2.5×12 mm, Johnson and Johnson Interventional Systems Co., Warren, NJ) into the stenotic left bronchus. The left lung expanded temporarily, but on repeat bronchoscopy and CT the next day, both imaging modalities revealed the crushed stent lodged in the left bronchus (Fig. 3).

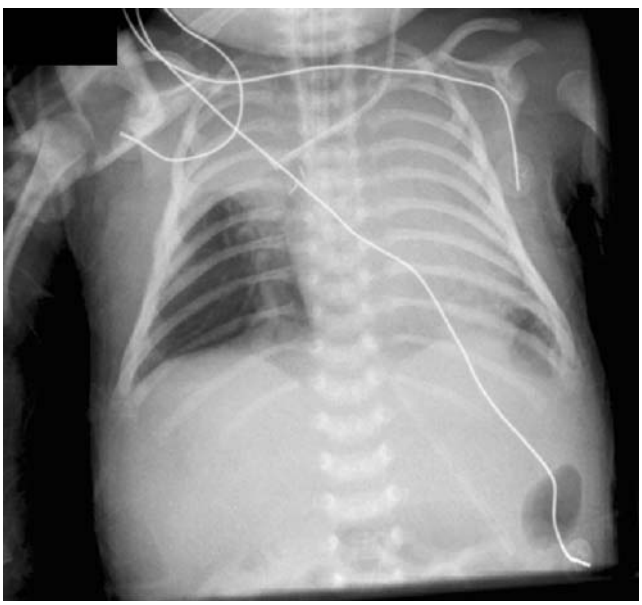

Fig. 2 Chest radiograph after the Blalock-Taussig shunt operation shows poor aeration of the left lung and collapse of the right upper lung

We then decided to restore left bronchial patency by surgical intervention on the fifth day of ECMO support. Under cardiopulmonary bypass, the ascending aorta was elongated with an 18-mm ePTFE tube conduit and repositioned away from the left main bronchus. Aortopexy was also accomplished by suture fixation to the posterior sternum. Finally, the Palmaz stent was removed bronchoscopically, which also confirmed the patency of the left bronchus after aortic arch reconstruction and repositioning. Postoperative chest radiography showed re-expansion of the left lung. Despite these efforts, the patient failed to wean from ECMO because of an overwhelming fungal infection, and he later died.

Discussion

Our patient had a torturous ascending aorta, VSD, PA, and concomitant chromosome 22q11.2 deletion (velocardiofacial) syndrome. We found no report of this stunning picture in the medical literature. The patient's torturous aorta originated in the very lateral aspect of the left ventricle crossed over the left bronchus. It then crossed the lower trachea to form a right aortic arch.

Patients with chromosome 22q11.2 deletion have a high incidence (75%) of cardiac anomalies, especially complex conotruncal anomalies, as in our patient [5]. In this patient with VSD and PA, the pulmonary arteries failed to grow, thus, yielding room for the development of the malpositioned aorta. The anomalous aorta served as the only outlet from the heart, which gradually became enlarged. Thereafter, the left main bronchus lying underneath the aorta was squeezed and grew smaller than normal.

Airway anomalies in infants cause excessive morbidity or mortality in the perioperative course [8]. These underrecognized airway problems are mainly caused by anatomic cardiorespiratory interactions. The mechanisms include

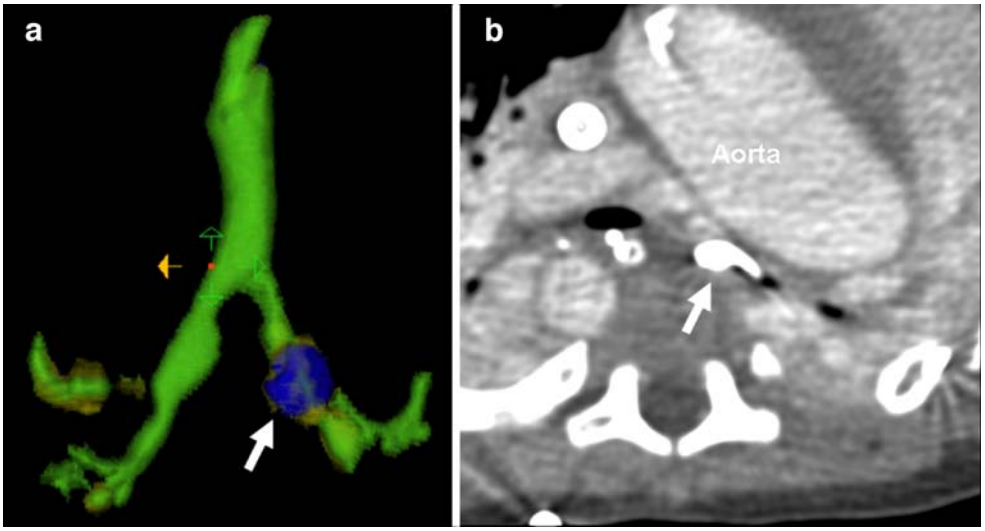

Fig. 3 Three-dimensional CT (**A**) and conventional CT (**B**) show the stent (*arrow*) collapsed and lodged in the left main bronchus

anomalous relationships between the tracheobronchial tree and vascular structures (complete or partial vascular rings) or result from extrinsic compression caused by dilated great vessels, left atrial enlargement, massive cardiomegaly, or postoperative reconstructed aortic arches [7]. Survival outcome analysis among patients showed that patients with cardiac/syndromic anomalies had inferior survival compared to patients with primary airway stenosis [6]. In this situation, flexible bronchoscopy is necessary to identify complete tracheal rings, the severity of stenosis, and any dynamic collapse of the airway [2]. Additionally, CT helps to delineate the relationship between the cardiovascular system and the tracheobronchial tree [11]. Severely symptomatic patients require aggressive treatment once the diagnosis is established.

To relieve our patient's severe bronchial stenosis, we opted to first place a stent in the left bronchus. However, the heavy, pulsatile aorta lying above it crushed the stent in the airway. Patency of the left bronchus was established only after we moved the aorta away from the left main bronchus. A variety of management approaches have been provided in children with airway obstruction, including internal wire mesh Palmaz stents, external Gore-Tex Hagl stents, aortoplasty, aortopexy, and slide tracheoplasty [1, 4]. Some patients require tracheostomy and long-term home ventilation [1]. Because the optimal treatment is highly individualized, prudent patient selection for operative repair is paramount and should be based upon the patient's condition and operative risks. Based on our experience, in patients with obvious external compression by a vessel supporting systemic circulation, an internal stent seems inadequate to relieve this type of severe obstruction.

In our patient with chromosome 22q11.2 deletion, VSD, PA, and a rare anomalous ascending aorta compressing the left main bronchus, stent placement failed to maintain left bronchial patency. Thus, surgical rerouting of the anomalous aorta or vessel should be considered to relieve tracheobronchial stenosis and restore patency.

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