Case report

Vertical vascular ring around right mainstem bronchus

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Abstract

An 18-year-old male patient was referred with an atypical form of vascular ring consisting of a right aortic arch, a right descending aorta, a persistent right ligamentum arteriosum, and a dilated right pulmonary artery in the setting of discontinuous pulmonary arteries with a single functional right lung. The initial presentation was that of pneumonia of the right lung and later evaluation revealed the right mainstem bronchus was squeezed between the descending aorta and the dilated right pulmonary artery. Surgical exploration disclosed the right-sided ligamentum arteriosum was running under the right mainstem bronchus and hereby was completing a vertical vascular ring. Decompression was achieved through surgical and bronchoscopic treatment.

Keywords: Vascular ring; Pulmonary artery discontinuity; Aortopexy; Endobronchial stent

1. Introduction

Tracheal compression of vascular origin most commonly is caused by variations in anatomy of the aortic arch and pulmonary artery system, and may or may not be accompanied by congenital anomalies of the heart [1,2]. However, the diagnosis of atypical airway compression around a bronchus may easily be overlooked because respiratory disease in children with congenital heart disease is common and often multifactorial (change in pulmonary blood flow, pulmonary hypertension, tracheobronchomalacia).

2. Case report

An 18-year-old male patient was referred to our hospital at the age of 4 months with signs of severe respiratory distress. A chest X-ray on admission showed a right middle and upper lobe infiltrate with an abnormal left pulmonary branching pattern and elevated left hemidiaphragm. The patient was treated successfully with invasive ventilation and broad-spectrum antibiotics. However, 1 month later, a ventilation-perfusion scan showed absent perfusion of the left lung and normal ventilation of both lungs. Echocardiography was normal. Angiocardio graphic evaluation disclosed a right-sided aortic arch-type 'mirror-image' branching, a right-sided descending aorta, an 'agenesis' of the left pulmonary artery, a hypoplastic left lung perfused by bronchial collaterals from the left internal mammary artery, and the remnants of a right- and left-sided arterial duct. Retrograde wedge injection in a left pulmonary vein showed thrombosis of the left pulmonary artery. The right pulmonary artery pressure was 38/10 mmHg (mean 22 mmHg). Genetic examination revealed a familial transmaternal 22q11 deletion (CATCH) syndrome (cardiac anomaly-abnormal facies-thymic hypoplasia-cleft palate-hypocalcemia). During the following years, the patient developed recurrent infections in the right lung accompanied in recent months with severe bronchospasms and dyspnea. Computed tomography of the chest demonstrated that the right mainstem bronchus was squeezed between the right descending aorta and a dilated right pulmonary artery (24.7 mm) (Fig. 1 A). Bronchoscopy showed a distinct pulsatile compression of the right bronchus from its posterior wall, reducing the latter to a slit-like opening along with signs of distal tracheomalacia. Right heart catheterization was repeated to exclude progression of pulmonary hypertension (PAP 31/12 mmHg, mean 20 mmHg). In order to relieve the bronchial compression, surgery was performed. Through a right fourth thoracotomy incision, the right pleural cavity was entered. Surgical exploration revealed a right-sided ligamentum arteriosum running under the right mainstem bronchus and completing a vertical vascular ring (Fig. 2). Division of the ligamentum rendered the right main bronchus additional space. With the aid of fiberoptic endoscopy and dimming the light in the operating
room, the site of aortopexy was correlated with the pulsatile stenosis. Posterior aortopexy was achieved by wrapping a double piece of Dacron patch 18 mm around the descending aorta and fixing (prolene 4/0) the lifted aorta at the lateral thorax wall. The aortopexy patch was protected by a Gore-tex (WL Gore & Associates, Flagstaff, AZ) membrane to avoid erosion of the adjacent lung. Intraoperative bronchoscopy showed a residual mild lumen reduction of approximately 20%. The patient was extubated 5 days later, and the postoperative course was uncomplicated. However, at 5 months of follow-up, he was readmitted with pneumonia of the right lung. Bronchoscopy showed a residual stenosis of the right main bronchus caused by extrinsic compression of the dilated pulmonary artery. Relief was obtained by bronchoscopic silicone stent placement. Computed tomography confirmed a satisfactory repair (Fig. 1B).

3. Discussion

Tracheobronchial compression in patients with congenital heart disease may become an important reason for respiratory insufficiency, largely because both conditions may be mutually exacerbating. Most cases of significant airway obstruction in these children result from altered hemodynamics or anomalous relationship between the tracheobronchial tree and vascular structures [1–3]. The respiratory distress in our case was due to a right mainstem bronchial compression by an atypical vertical vascular ring, a non-functioning left lung, secondary tracheomalacia, and a cellular immunodeficiency due to thymic hypoplasia. Bilateral arterial ducts are most often found in the context of discontinuous pulmonary arteries with one duct giving origin to the ‘excluded’ pulmonary artery and the other running between the central pulmonary and systemic artery [4]. Nevertheless, a right duct running under the right main bronchus and hereby completing a vertical vascular ring, has never been reported. Although concomitant cardiac malformations were described such as tetralogy of Fallot, atrial septal defect, or coarctation of the aorta, this was neither the case in our patient [4]. As expected, the right pulmonary artery was disproportionally dilated with pulmonary hypertensive changes because of the passage of the full cardiac output through the right lung. The optimal strategy to treat this case represented a difficult issue mainly because the patient’s respiratory symptoms were caused by bronchial compression of vascular origin and tracheomalacia. Undoubtedly, the best treatment would have been neonatal stenting of the left arterial duct and later surgical reconstruction of the ‘excluded’ pulmonary artery concomitantly with transection of the right ligamentum arteriosum [5]. Nevertheless, in most cases, at the time of diagnosis the ductus has already closed and fibrosed with none or minimal atypical symptoms. The only approach to ensure pulmonary artery branch
development and to allow normal lung growth is by implantation of a modified Blalock-Taussig shunt. However, in the worst case, as reported here, the pulmonary artery becomes thrombosed and salvage of the ipsilateral lung is almost impossible [6]. In our patient, there might have been a role for prophylactic division of the right ligamentum arteriosum which by itself contributed for a large part to compression of the right main bronchus. Unlike in our case, recurrent infections of the underdeveloped lung may necessitate pneumonectomy. Another dilemma that we encountered was the surgical approach to address this anomaly. We chose a right thoracotomy and used fiberoptic endoscopy as complementary tool to direct the position of aortopexy. However, through this approach, it was technically impossible to plico-suspend the distended pulmonary artery. Bronchoscopic guided silicone stent implantation offered a valuable alternative to treat residual tracheobronchial narrowing. This case may be considered as another not yet described anatomic variation of vascular ring in conjunction with discontinuous pulmonary arteries. In the setting of discontinuous pulmonary arteries with bilateral ducts, the clinician should be aware of the possibility of a vertical vascular ring around the right mainstem bronchus.

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References