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Pulmonary Artery Sling Causing Stridor in a Neonate

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Abstract Neonates with upper airway obstruction commonly present with stridor. We report a case of pulmonary artery sling and congenital tracheal stenosis in a neonate who presented with stridor and respiratory distress. Management of tracheal stenosis, in particular, the use of slide tracheoplasty, is discussed and reviewed.

Key words Congenital tracheal stenosis; Magnetic resonance imaging; Pulmonary artery sling; Slide tracheoplasty

Introduction

Stridor is a warning sign that suggests upper airway obstruction. Among the long list of causes of upper airway obstruction, one should particularly have a high index of suspicion for congenital causes if it presents in the early neonatal period. Early bronchoscopy and the appropriate use of radiological imaging examinations can assist clinicians to arrive at the diagnosis. We presented a case of pulmonary artery sling with tracheal stenosis that presented with stridor. Diagnosis of pulmonary artery sling and treatment of tracheal stenosis was discussed.

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Case Report

HKW, a previously well baby boy, born at 39 weeks of gestation with birth weight 2.57 kg to a non-consanguineous couple, presented on day 5 of life with stridor at rest. Antenatal history and perinatal course were unremarkable, and there was no evidence of sepsis. On physical examination, he had biphasic stridor with suprasternal insucking but no tachypnoea. Cardiovascular system and other parts of the examination were normal. Chest X-ray and lateral neck X-ray were normal.

Bronchoscopy showed an elongated and tubular epiglottis with collapse of arytenoid and cuneiform tubercles during inspiration, as well as an extramural compression at the mid-tracheal level, and there was no laryngomalacia. Magnetic resonance imaging (MRI) of thorax (Siemens Magnetom Vision Plus, 1.5T VB33G (Erlanger, Germany)) (Figure 1) revealed an anomalous left pulmonary artery, which branched from the right main pulmonary artery and passed behind the trachea to reach the left lung. Echocardiogram, cardiac catheterisation and pulmonary arteriogram (Figure 2) confirmed the anomalous

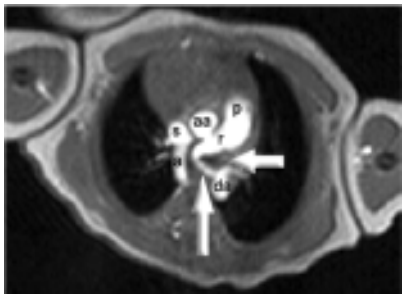

Figure 1 Magnetic resonance angiogram demonstrated the anomalous left pulmonary artery (vertical arrow) compressing the trachea (horizontal arrow). Pulmonary artery trunk (p), right main pulmonary artery (r), ascending aorta (aa), descending aorta (da), azygos vein (a) and superior vena cava (s) are labelled.

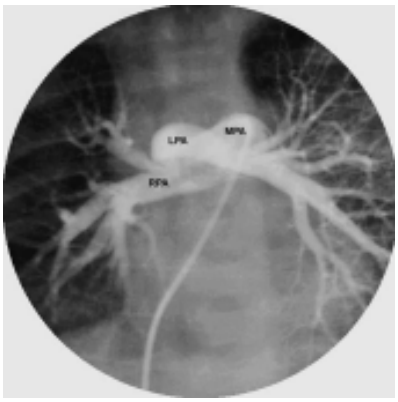

Figure 2 This pulmonary arteriogram in anteroposterior projection showing the left pulmonary (LPA) arises from the right pulmonary artery (RPA) instead of the main pulmonary artery (MPA).

left pulmonary artery arising from the right pulmonary artery, together with a small patent ductus arteriosus and a small patent foramen ovale. Bronchogram revealed a tapering tracheal stensosis of the middle and lower portions about 13.4 mm long, with the narrowest part being 1.5 mm in diameter.

The baby underwent correction of the left pulmonary artery sling and slide tracheoplasty with cardiopulmonary bypass at two months of life. Apart from sternotomy wound infection by methicillin-resistant staphylococcus aureus, which was treated with antibiotics, the post-operative recovery was uneventful. He was discharged 27 days after the operation. Echocardiogram on discharge was normal.

Discussion

Pulmonary sling, or anomalous left pulmonary artery, represents failure of development of the left sixth aortic arch, followed by the development of a collateral branch from the posterior aspect of the right pulmonary artery to supply the left lung. The anomalous left pulmonary artery courses over the right main stem bronchus, passing posterior to the trachea and anterior to the oesophagus, to reach the left hilum. The course of the anomalous left pulmonary artery to the right of the trachea produces deviation of the lower trachea to the left with resulting compression of the right mainstem bronchus and lower trachea. This causes airway obstruction that affects primarily the right lung, although compression of the lower trachea and left mainstem bronchus can result in bilateral obstruction. There may be associated tracheobronchial abnormalities which include complete tracheal rings, tracheomalacia, hypoplasia and stenosis of tracheal segments. Congenital heart defects are found in 50% of the cases, most commonly atrial septal defect, patent ductus arteriosus, ventricular septal defect and left superior vena cava. Other organ system abnormalities may occur, including imperforate anus, Hirschsprung disease, biliary atresia, and genitourinary defects.[1]

Potts, in 1954, reported the first description of surgical repair of this lesion.[2] Surgery involves division of the anomalous left pulmonary artery and re-anastomosis to the main pulmonary artery anterior to the trachea. Mortality is variable and the major determining factors are the associated bronchial and tracheal abnormalities. If airway stenosis is severe, patients may require surgical reconstruction of the obstructed bronchi or trachea. Patients should be monitored closely for the possibility of left pulmonary artery stenosis after reimplantation of the vessel. Morbidity includes recurrent pneumonia, residual airway obstruction with recurrence of stridor and respiratory distress with or without wheezing and cyanosis. Some degree of airway obstruction may persist in up to 45% of surgical survivors.[1]

MRI directly demonstrates the anomalous vessel and its course, and is able to exclude other vascular rings and other associated abnormalities. The best sequence would be gradient echoes cine acquisition so that the mediastinal vascular structures bright up and a sense of directional flow can be visualised in the cine loop. A transmit receive surface coil, best fit the size of the patient should be used to optimise the signal to noise ratio and to reduce the specific absorption

rate. In this case, an extremity coil was used. Gradient cine sequence, TR 30.0 ms, TE 4.8 ms, flip angle 30°, FOV 15.6 × 25 cm, image matrix 114 × 256 and 4 mm slice thickness with 10% gap, was acquired through the upper mediastinum. Siripornpitak et al[3] had shown that magnetic resonance (MR) flow study could demonstrate the decreased flow in the left pulmonary artery with tardus and parvus flow pattern. Faust et al[4] used direct coronal or sagittal cine MR over the airway to demonstrate tracheomalacia. These techniques had not been used in our patient because we would like to avoid prolonged sedation and oxygen desaturation. Multi-slice computerised tomography covers a greater volume during a simple breath hold. It scans with supreme longitudinal and in-plane spatial resolution, as well as temporal resolution. It shows the morbid anatomy well and reconstruction can be done in various projections, cross-sections, shaded surface and volume rendering. It was not employed in this patient due to the concern of radiation dose and iodinated contrast injection.

Congenital tracheal stenosis, a rare condition once considered lethal, presents in infancy with symptoms of respiratory distress like stridor, retractions, apnoea, cyanosis and occasionally life threatening events. It occurs in varied lengths, which are most often characterised by complete cartilaginous rings. The stenotic segment may be located in upper or lower trachea, may be funnel shaped, and may involve different percentages of the length of trachea. In 30-50% of cases, a left pulmonary artery sling may be found with either a short segmental stenosis or a more complex form of stenosis.[5] Concomitant reconstruction of congenital tracheal stenosis and pulmonary artery sling has been reported. Other associated cardiac lesions include Tetralogy of Fallot, ventricular septal defect, complete atrioventricular canal and others. In the event of a severe cardiovascular malformation on echocardiography, cardiac catheterisation is mandatory. Chest radiography may show a narrowed airway. However, chest radiograph may only show non-specific changes or even normal findings. CT and MRI are helpful in identifying the stenosis and are important in identifying associated cardiovascular anomalies. Recent advances in three-dimensional CT imaging make it a promising non-invasive technique for delineating the anatomy of the stenosis. However, the key to confirming the diagnosis relies mainly on bronchoscopy.

Few surgical options were available for congenital tracheal stenosis, including resection with end-to-end anastomosis, oesophageal wall patch graft, cartilaginous support graft and pericardial patch, until Tsang et al proposed a new technique called slide tracheoplasty in 1989.[6] The long segment of stenosis was divided transversely in its midpoint, a vertical anterior incision made through the full length of the upper half of the stenotic segment and a corresponding vertical slit made posteriorly in the distal half of the divided stenotic segment. The right-angled corners where the vertical incisions met the transverse incision were trimmed. The two ends were slid together, doubling the circumference of the trachea. This resulted in a fourfold increase in cross-sectional area of the airway and the stenotic segment was shortened by one half of its length. The traditional technique of slide tracheoplasty was adopted for the current case.

Grillo[7] modified the technique and made access for suturing easier by reversing the vertical incisions such that the promixal segment was incised posteriorly and the distal segment anteriorly. In a series of 11 patients with congenital tracheal stenosis, Grillo et al[8] demonstrated excellent short-term and long-term results with slide tracheoplasty. The advantages of slide tracheoplasty over various techniques of patch tracheoplasty lie in the immediate reconstruction of trachea with the patient's native tracheal tissues, providing a stable cartilaginous wall with normal epithelial lining. It does not lead to loss of viability from interference with tracheal blood supply or to failure of healing. Most of the lateral blood supply of the trachea is preserved. Excessive tension does not develop. Direct complications are limited to a few one-time granulomas and a transient injury to a recurrent laryngeal nerve, whereas in patch tracheoplasty, recurrent granulation tissue formations and necrosis or collapse of the patch sometimes occurred. Cardiopulmonary bypass is usually not needed, and is usually employed for accompanying cardiovascular procedures or borderline cardiac physiology. Routine postoperative endotracheal tube splinting is unnecessary, thus allowing early extubation.

Acosta et al[9] showed better results with slide tracheoplasty over resection and anastomosis, as tension is more effectively distributed over a longer anastomotic line, trachea ischaemia is avoided and there is a lower incidence of problems with the anastomosis such as leaks, mediastinitis, fibrosis and recurrent stenosis. The first costal cartilage graft procedure for long segment congenital tracheal stenosis was reported by Kimura et

al in Japan[10] who recently presented an analysis of 29 patients over two decades that slide tracheoplasty is preferrable to cartilage graft because it preserves native tracheal tissue with less frequent complications.[11]

Intratracheal stenting with expandable metallic wire stents was also advocated for tracheomalacia or tracheal stenosis. In a recent series of five infants with congenital tracheal stenosis, aged 7 days to 12 months, placement of tracheal stents after intraluminal balloon dilatation relieved four patients of airway obstruction and the remaining one died after palliation.[12] Apart from the development of granulation tissue over the stents which could be managed by scraping or balloon compression, the stents were in place for 9 to 36 months without any other complications. Although the short-term result is satisfactory, further clinical trials are required to evaluate the long-term effect of this procedure. Possible long-term risks include re-stenosis by granulation tissue, failure of tracheal growth with age, and tracheal erosion or penetration to the great vessels.

Summary

In this paper we present a case of pulmonary artery sling and tracheal stenosis causing respiratory distress in a neonate. Appropriate use of imaging techniques helped arrive at the diagnosis. Different modalities of surgical treatments are available for this rare but potentially lethal condition. Slide tracheoplasty is one of the newer techniques that offer promising result.

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