CASE REPORT

Severe and Fatal Postoperative Bronchospasm in a Child with a Pulmonary Artery Sling

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Summary

We report a case of left pulmonary artery sling in a child who also had duodenal atresia. He was admitted for respiratory failure requiring prolonged respiratory support due to a bronchiolitis-like illness at two months of age. Diagnostic procedures confirmed the presence of left pulmonary artery sling. He had a corrective procedure which relieved the compression. However postoperatively he had intermittent episodes of severe bronchospasm in addition to a persistent airway obstruction. Finally one such episode of severe bronchospasm did not respond to medical and resuscitative therapy and the baby succumbed. To our knowledge no case of left pulmonary artery sling has been described previously in a Malaysian child. This case also highlights the postoperative airway problems that may be encountered.

Key Words: Pulmonary artery sling, Postoperative airway problems

Introduction

An aberrant left pulmonary artery sling represents a developmental anomaly associated with respiratory distress in infancy. Symptoms of airway obstruction are secondary to the compression by the aberrant pulmonary vessel on the tracheo-bronchial tree as well as the maldevelopment of the tracheo-bronchial cartilage leading to tracheomalacia. We present a case of left pulmonary artery sling admitted for severe acute respiratory distress at two months of age. This child also had duodenal atresia which had been operated at day four of life. He was successfully operated on but continued to wheeze with intermittent severe bronchospasms and finally died.

Case Report

M.A. was born at term, to a healthy 34-year-old primigravida. The Apgar score was 9 at one

minute and the birth weight was 3.0kg. He was admitted to the special care nursery at two hours of life for congenital pneumonia. He was operated at day four of life for a duodenal atresia. He was discharged well at two weeks old. At home mother noticed that he had noisy breathing which was dismissed by the family doctor. At two months old he was readmitted to the paediatric unit with noisy breathing of four days duration. On admission he was tachypnoeic with subcostal and intercostal recessions. There was an audible wheeze and a suggestion of a stridor. Auscultation of the lungs' fields revealed equal breath sounds with bilateral rhonchi and widespread crepitations. Other systems were normal.

Three hours after admission he was more tachypnoeic and cyanosed (type two respiratory failure) requiring ventilation. He required frequent inhalational bronchodilator with intravenous steroids. Weaning from the ventilator was difficult and he was ventilated for a month. During this period he had recurrent pneumonias and bilateral pneumothoraces which was drained. Although there was no proof of gastroesophageal reflux, intravenous metaclopropamide and cimetidine were added.

With these measures he was finally extubated. Post extubation he continued to wheeze with marked audible stridor. He was tachypnoeic with subcostal and intercostal recession requiring oxygen support and inhalational bronchodilator and steroids.

Investigations done showed a normal milk scan. An oesophagogram showed an anterior indentation of the oesophagus suggesting a vascular anomaly (Fig. 1). This explains the persistent wheeze he had. Echocardiography followed by an angiogram confirmed that he had left pulmonary artery sling (Fig. 2).

A direct laryngoscopy and bronchoscopy showed that the lower part of the right bronchus was narrowed 0.5 cm distal to the right carina with normal mucosa.

The larynx and trachea were normal with no compression seen. The left bronchus was normal. He was successfully operated at the National Heart Institute.

Fig. 1: Contrast oesophagogram showing anterior indentation of the oesophagus caused by a left pulmonary artery sling

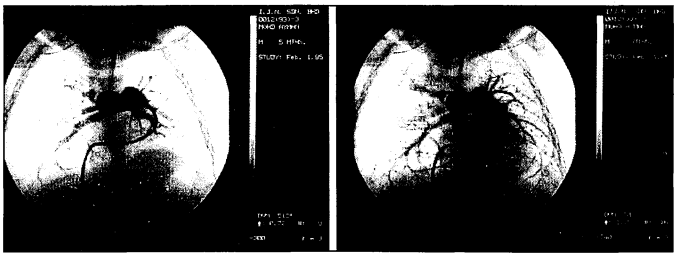

Fig. 2: Cardiac angiogram showing left pulmonary artery sling

Postoperatively he was ventilator dependent with frequent episodic and severe bronchospasms. Finally he succumbed to irreversible severe attacks of bronchospasms despite all resuscitative measures.

Discussion

Pulmonary artery sling was first described in 1897 as a post mortem finding by Glaevecke and Doehle. It is a vascular abnormality where the left pulmonary artery arises from the right pulmonary artery and transverses between the oesophagus and trachea towards the hilum of the left lung. This produces a sling around the distal trachea and the proximal main bronchus.

The extent of symptoms correspond to the degree of compression of the trachebronchial structures by the aberrant vessels which include wheezing and stridor. Tracheal anomalies occur with equal frequency, the most common being hypoplasia of the distal trachea. However there is no literature describing gastrointestinal anomalies with pulmonary artery sling as seen in this patient.

Associated cardiac anomalies are seen in 50 per cent of these patients[1] and most commonly include persistent left superior venacava and ventricular septal defect.

The plain chest radiograph may show hyperlucency and/or atelactasis of the right lung secondary to expiratory obstruction of the right main stem bronchus.

The pulmonary artery sling is the only vascular ring that produces an anterior indentation[2] of the oesophagus on the oesophagogram, and consequently this is an excellent diagnostic tool.

Pulmonary artery sling is treated surgically with a low mortality and morbidity[3]. This patient had a successful operation. Although the compression was relieved he continued to have persistent intermittent severe airway obstruction and was not able to be extubated postoperatively. The persistent obstruction could be due to localised tracheomalacia at the site of compression. The severe intermittent bronchospasms which finally proved fatal could have several possible contributing factors. The bronchiolitic illness which precipitated the initial respiratory failure could have resulted in severe bronchial hyperactivity. Additionally, there could have been an associated gastroesophageal reflux which predisposed to bronchospasms. There is no direct evidence for this, but we noted earlier that there was worsening respiratory distress with increasing feeds and that an antireflux regime improved the respiratory status significantly. It might be postulated also that the repaired duodenal atresia could have interfered with gastric motility predisposing to reflux.

This case illustrates that the course of the respiratory illness in a child with pulmonary artery sling may be fatal and severe.

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