

Two Major Respiratory Disorders in One Unlucky Child, Operated Congenital Lobar Emphysema Turns Out to be Asthmatic

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Abstract

Congenital lobar emphysema (CLE) is a rare congenital abnormality characterized by over inflation of a pulmonary lobe. It often presents a diagnostic and therapeutic dilemma. On the other hand, Bronchial asthma is a chronic inflammatory disorder of airways that has both genetic and environmental factors.

We describe here a case of 2 years old child who developed respiratory problems soon after birth diagnosed and operated as congenital lobar emphysema, both his vascular anatomy and the bronchial wall were normal. He improved dramatically after surgery. The SaO₂ normalized within a week of Surgery (lobectomy) and later he suffered two attacks of bacterial pneumonia. After recovery from surgery and gradual reinflation of the lung a whole new respiratory problem emerged to the surface, by 6 months the child started to develop wheezy chest with each upper respiratory viral infection and not controlled until he received the regular treatment of asthma. Up to our knowledge this cannot be as a complication of congenital lobar emphysema.

Keywords: *Congenital Lobar Emphysema; Asthma*

Abbreviations

CLE: Congenital Lobar Emphysema; NVD: Normal Vaginal Delivery; CT: Computed Tomography; FEV₁: Forced Expiratory Volume in First Second

Introduction

We describe here a child that was unlucky enough to have two major separate diseases in one system, strange enough these two disorders are completely separate in pathophysiology and treatment but this child happens to have them both.

This child had congenital lobar emphysema which has been operated (surgically removed the emphysematous lobe) and as he recovers with gradual inflation of the lungs and gradual improvement of pulmonary hypertension then he seems to develop the typical asthma along with some skin atopy. His asthma attacks were mostly following upper respiratory tract infection, we could hear bilateral expiratory wheeze with some crepitations. Those attacks were not relieved except with bronchodilators and steroids and they were so frequent that we had to start controllers according to asthma guidelines.

Case Report and Discussion

This is a case of a 2 years old child who was delivered NVD and went home not incubated within few days he started developing respiratory distress and difficult feeding, as parents sought medical advice he was admitted and chest X-ray performed then CT which revealed congenital lobar emphysema.

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At the age of 15 days the newborn was operated and stayed after surgery on oxygen for almost 10 days, as gradually his respiratory distress was improving he was discharged home with intermittent oxygen therapy. His echo examination at that time revealed mild pulmonary hypertension of 45 mmhg.

Later on, He developed 2 attacks of bacterial pneumonia in which he was admitted and received 2 courses of parental antibiotic but mechanical ventilation was not needed in either attack. Other than this his lung seemed to be reinflating, he was gaining weight and his condition was getting more stable with each day.

By the age of 6 months the child started experiencing attacks of wheezy chest, mostly following viral upper respiratory tract infection and sometimes not. Bilateral expiratory wheeze with medium sized crepitations could be heard. He has skin atopy following insect bite and later to some forms of food. Those attacks required nebulizers with bronchodilators and steroids to be controlled and later as the child grow and his air entry on the left lung where he had the emphysema improved it was obvious that his asthma was a separate entity than his emphysema.

As the attacks were so frequent requiring systemic steroids almost every 15 days we started him on controllers.

Now the child is on budesonide inhaler 200mic twice daily and montelukast 4 mg once daily with good control of his asthma.

Those two conditions are very different regarding the cause, pathophysiology and treatment. Though congenital lobar emphysema can be attributed to defect in bronchial tree anatomy and wheezes can be heard along the disease, cases usually improve dramatically following the surgery, also the type of attacks in this child, its triggers, its reversibility and its control on antiasthma drugs along with history of skin atopy in this child all that points to the atopic form of bronchial asthma as a separate entity in this child. Though tests of pulmonary function and tests of allergy could not be performed on this child regarding his age and the inconclusive nature of those tests clinical evaluation of this child left no doubt regarding the nature of his asthma.

Congenital lobar emphysema is a rare respiratory disorder in which air can enter the lungs but cannot escape, causing over inflation (hyperinflation) of the lobes of the lung. It is most often detected in newborns or young infants, but some cases do not become apparent until adulthood. This disorder may be severe enough to cause associated heart problems (15% of cases) or so mild as to never become apparent. Some cases of congenital lobar emphysema may be caused by autosomal dominant inheritance while others occur for no apparent reason (sporadic).

In 1954 Gross and Lewis, published the first case report of CLE [1]. Male babies are affected more often than female in the ratio of 3:2 [2]. The incidence of left upper lobe involvement is 43%, right middle lobe 32%, right upper lobe 20%, and bilateral involvement 20% [3].

In these infants, there is increased intrathoracic pressure because of hyperinflation of one or more pulmonary lobes, leading to mediastinal shift and atelectasis of the ipsilateral or contra lateral lobes of the lung. This causes displacement of heart sounds, decreased venous return, and varying degrees of ventilation-perfusion mismatch, which leads to hypoxia. Chest radiographs help to diagnose but is not definitive [4]. A CT scan confirms the diagnosis and may rule out associated anomalous vascular slings.

Several factors have been associated with the development of congenital lobar emphysema. In 50% of cases there is decreased bronchial cartilage tissue. This defect produces a ball valve effect with consequent over inflation [5]. Vascular abnormalities that produce compression [7] bronchial stenosis [6] bronchogenic cysts [8,9] and congenital cytomegaloviral infection have also been associated [5]. Congenital lobar emphysema has been described in twins [6] but in up to 40% of cases, the cause is unclear [6,10]. Our patient had a normal vascular anatomy and bronchial cartilage on gross examination nor were there any other lung abnormalities. Concurrent congenital heart disease (usually patent ductus arteriosus, or ventricular septal defect, occurs in 15% of cases [11]. Our patients had neither of these on echocardiography.

Congenital lobar emphysema is characterized by (1) difficulty in breathing or very rapid respiration (respiratory distress) in infancy, (2) an enlarged chest due to over inflation of at least one lobe of the lung, (3) compressed normal lung tissue in the section of the lung nearest to the diseased lobe, (4) bluish color of the skin due to a lack of oxygen in the blood (cyanosis), and (5) underdevelopment of the cartilage that supports the bronchial tube (bronchial hypoplasia).

Asthma is a chronic inflammatory disorder of the airways characterized by an obstruction of airflow.

Airway inflammation is the result of interactions between various cells, cellular elements, and cytokines. In susceptible individuals, airway inflammation may cause recurrent or persistent bronchospasm, which causes symptoms that include wheezing, breathlessness, chest tightness, and cough, particularly at night (early morning hours) or after exercise [12].

Airway inflammation is associated with airway hyperreactivity or bronchial hyperresponsiveness (BHR), which is defined as the inherent tendency of the airways to narrow in response to various stimuli (e.g. environmental allergens and irritants) [13].

Interactions between environmental and genetic factors result in airway inflammation, which limits airflow and leads to functional and structural changes in the airways in the form of bronchospasm, mucosal edema, and mucus plugs [14].

According to the National Asthma Education and Prevention Program guidelines, spirometry is an essential objective measure for establishing the diagnosis of asthma. Additional studies are not routinely necessary, but they may be useful when the clinician is considering alternative diagnoses. Eosinophil counts and IgE levels may be useful when allergic factors are suspected [15].

Bronchial provocation tests may be performed to diagnose bronchial hyperresponsiveness (BHR). These tests are performed in specialized laboratories by specially trained personnel to document airway hyperresponsiveness to substances (e.g. methacholine, histamine). Increasing doses of provocation agents are given, and FEV_1 is measured. The endpoint is a 20% decrease in FEV_1 (PC_{20}) [16].

Results of pulmonary function testing are not reliable in patients younger than 5 years. In young children (3 - 6 y) and older children who are unable to perform the conventional spirometry maneuver, newer techniques, such as measurement of airway resistance using impulse oscillometry system, are used. Measurement of airway resistance before and after a dose of inhaled bronchodilator may help to diagnose bronchodilator-responsive airway obstruction [17].



Figure 1, 2: Show postoperative hypovoluminous left lung after lobectomy with gradual re-inflation of left lung.

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