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Clinical Case Seminar **A6 (1-5)**

A clinical case of intricate tracheal disease and asthma

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Abstract

Context: Asthma is the most common inflammatory disease of the airways in children. The term problematic asthma is used to describe children with chronic symptoms or acute severe exacerbations, or both, not responding to standard asthma therapy.

Case Presentation: We describe the case of a 5 years old girl with a history of recurrent respiratory infections and asthma since the first months of life, with a poor response to conventional therapies (antibiotics, high dose corticosteroids combined with long-acting β2 agonists and oral leukotriene-receptor antagonists). In some cases she needed hospitalization for the important respiratory engagement and radiological findings of pulmonary consolidations, predominantly localized to the right lung. Computed tomography angiography (CTA), described a mild tracheobronchomalacia caused by the right innominate artery compression and a dense tissue mediastinal extrinsic compression of the main bronchus of the middle lobe, defined a Middle Lobe Syndrome (MLS).

Evidence Acquisition: MLS is defined as a recurrent or chronic collapse or infection of the middle lobe of the right lung. There is often a history of multiple treatments with antibiotics and anti-asthmatic drugs for ''recurrent pneumonia'' or ''asthma'', as in our patient. Chest X-ray is the first-line diagnostic tool, especially on the lateral view. CT-scan and bronchoscopy are considered useful for diagnosis or treatment. Treatment depends on etiology of MLS.

Conclusion: In case of severe asthma symptoms co-morbidities must be evaluated. MLS is frequently unrecognized in children and thinking of it is a prerequisite for diagnosis, especially when recurrent respiratory infections or asthma symptoms are predominantly localized to the right lung.

Key-Words: Respiratory tract infections, severe asthma, tracheobronchomalacia, Middle Lobe syndrome

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Introduction

Asthma is the most common inflammatory disease of the airways in children characterized by period of reversible airflow obstruction and increased bronchial reactivity to nonspecific stimuli. Among children current asthma prevalence is about 10%. Asthma episodes may be mild or moderate and can be controlled by traditional asthma therapies according to International Guidelines. However, less than 5% of patients has a severe asthmatic disease.(1).

Severe asthma can be defined as the American Thoracic Society stated in 2000 when one of two major criteria (1. Continuous intake of high-dose inhaled steroids; 2.taking oral steroids -OCS- for more than six months during the previous year) and at least two of seven minor criteria

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pulmonary, function tests, frequency of exacerbations, disease stability at reductions of therapy, use of health facilities, are fulfilled.(1)

The term of problematic asthma was first used by Bush et al. (2) in 2008 to describe children with chronic symptoms or acute severe exacerbations, or both, not responding to standard asthma therapy. The exact prevalence of this population is difficult to estimate, but the atopy is almost always present.(3, 4) The assessment of these children should be obtained through a systematic protocol. The initial step is to ensure that a wrong diagnosis has not been made ("not asthma at all"); and next to evaluate co-morbidities ("asthma plus"). The next step is a detailed multidisciplinary assessment to develop the best individualized treatment.

Case Report.

We describe the case of a 5 years old girl. She had been born at full-term gestation, without complications. Weight and length were adequate for gestational age at birth. The patient was breast-fed exclusively for one month. Her mother and brother had a history of asthma and allergic rhinitis during childhood. Since the early months of life she suffered from recurrent airway infections and asthma. The episodes often have a severe course and in some cases she needed hospitalization for the important respiratory engagement. In these occasions we underlined high inflammatory markers (neutrophilic leukocytosis and high values of c-reactive protein) on hematological tests and radiological findings of pulmonary consolidations, predominantly localized to the right lung. . She was treated with short-acting β2 agonists and antibiotics, in cases of acute severe asthma or suspected pneumonia, and traditional maintenance therapy asthma using an anti-static valved holding chamber (CSI combined with long-acting β2 agonists and oral leukotriene-receptor antagonists) with poor benefit. Therefore our patient had undergone careful systematic protocol to exclude other diagnostic hypotheses. Skin Prick Test and IgEs were mild positive for house dust mites and Grass pollen. Throat swab, blood culture, Mantoux test,, QuantiFERON test and viruses, Mycoplasma and Chlamidia Pneumonia serological tests were normal.

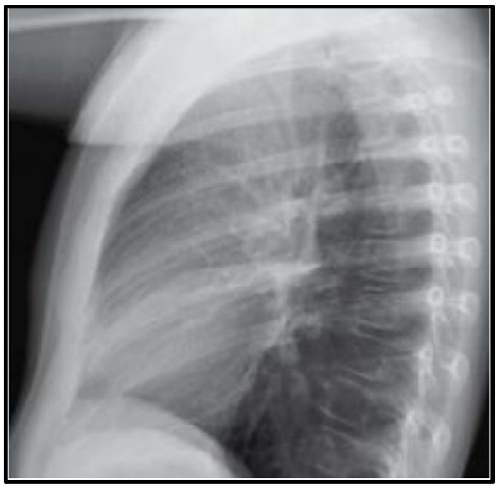

Figure 1. Chest X-ray on the lateral view. Wedge-shaped opacity bordered by the horizontal and oblique fissures.

Immunodeficiency disorders, Cystic Fibrosis and Ciliary Dyskinesia were excluded.

An esophago-gastro-duodenal transit study was normal. During exacerbations chest X-ray on the lateral view showed a wedge-shaped opacity bordered by the horizontal and oblique fissures (Figure1). Finally Computed Tomography Angiography (CTA) described a mild tracheobronchomalacia caused by the right innominate artery compression and the bronchomalacia

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and stenosis of the main bronchus of the middle lobe determined by a dense tissue mediastinal extrinsic compression, associated with right middle lobe atelectasis. (Figure2)

Radiological findings described a condition of tracheobronchomalacia and Middle Lobe Syndrome. In clinical health condition the airway endoscopy confirmed the anatomical malformations CT-scan revealed. The cytological and culture study of BAL-fluid showed inflammatory cellularity with few fat macrophages and positivity for H. Influenzae type B (1.000.000 UFC/mL).

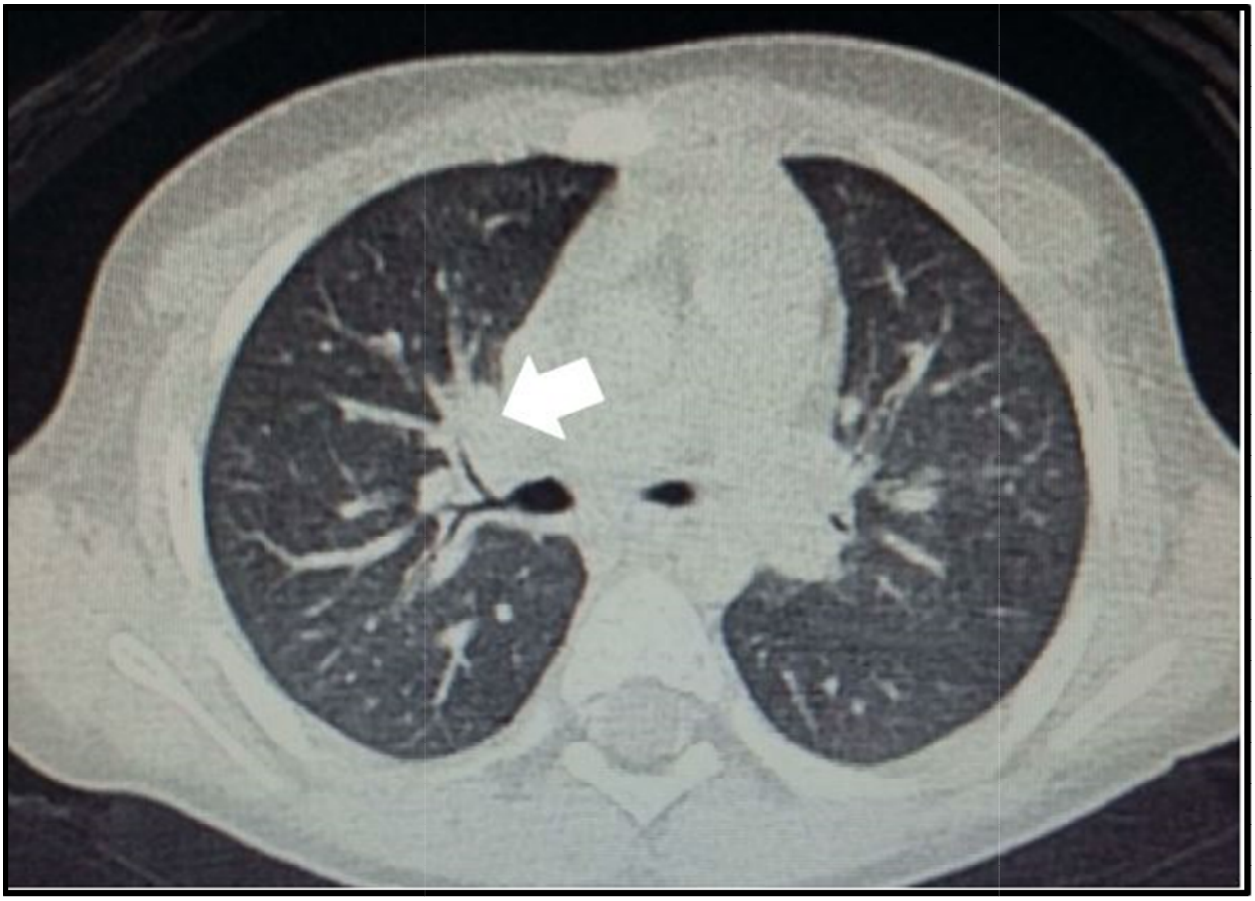

Figure 2. Computed Tomography Angiography (CTA)- Bronchomalacia and stenosis of the main bronchus of the middle lobe determined by a dense tissue mediastinal extrinsic compression, associated with right middle lobe atelectasis.

Discussion

Middle Lobe Syndrome (MLS) is long been recognized in literature as a specific clinical and radiographic entity, defined as a recurrent or chronic collapse or infection of the middle lobe of the right lung. The process can also occur in other areas of the lung, particularly in the lingula.(5) The exact prevalence of MLS is unknown, it can occur in all age groups, but it is more frequent in preschool years and girls are interest twice as often than boys. (5, 7). The etiologies are traditionally classified into obstructive and nonobstructive, but pathophysiologic mechanisms can occasionally interact. (Table 1).

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Obstructive MLS can be caused by extrinsic compression of the middle lobe bronchus or by endobronchial lesions.

In the nonobstructive type of MLS, the middle lobe bronchus is susceptible to collapse for several anatomical as well as functional factors: the narrow diameter and long length of the middle lobe bronchus, the deep fissures of the middle lobe with only scanty parenchymal bridges which determine a relative anatomical isolation, the crown arrangement of lymph nodes around the middle bronchus, and finally the poor development of the inter-alveolar pores of Kohn in early life.(5, 7). The MLS clinical presentation is characterized by nonspecific respiratory symptoms, such as chronic or recurrent cough, intermittent wheezing, and recurrent or persistent pneumonia.

Table 1 Romagnoli et al. (2014) Middle lobe syndrome in children today. PaediatrRespirRev;15.1883. doi:10.1016/j.prrv.2014. 01.002.Epub2014Jan31

Causes of atelectasis in childhood

1. Obstructive
Bronchial compression
Lymph nodes
Tumors
Cardiomegaly
Endobronchial obstruction
Exogenous
Foreign body
Recurrent aspiration
Histoplasmosis
Endogenous
Polyps
Papillomas
Adenomas
Granulomas
2. Nonobstructive
Mucus plugs
Asthma
Cystic fibrosis
Immotile cilia syndrome
Bronchiectasis
Pneumonia
Bronchopulmonary dysplasia
Surfactant deficiency or dysfunction
Hyaline membrane disease
Pulmonary edema
Near-drowning
Respiratory distress syndrome
Chest wall defects and neuromuscular diseases
Abnormalities of diaphragm
Spinal muscular atrophy
Werding-Hoffman disease
Muscular dystrophies
Guillan-Barrè syndrome
Intrathoracic compression
Pleural effusion
Chylothorax
Hemothorax
Pneumothorax

It also may present as asymptomatic, so it leads to delayed or frequently unrecognized diagnosis in children. There is often a history of multiple treatments with antibiotics and anti-asthmatic drugs for ‘‘recurrent pneumonia’’ or ‘‘asthma”, as in our patient. Indeed asthma may be frequently associated with MLS and when a viral infection occurs in an asthmatic patient, inflammation can occlude the lumen of the right middle bronchus, causing partial or complete obstruction. Chest X-ray is the first-line diagnostic tool, especially on the lateral view. In recent years CT-scan and bronchoscopy are considered useful for diagnosis or treatment, the first to identify endobronchial abnormalities, bronchiectasis and other parenchymal abnormalities, the second is often therapeutic in acute or chronic phase (foreign body aspiration or mucus plugs removal). Bronchoalveolar lavage can be concurrently performed to determine cellular elements and infection signs. Treatment depends on causes of MLS.

Conservative intervention should be the first-line, using anti-asthmatic therapy and antibiotics agents, only if bacterial infection is present or suspected. Chest-physiotherapy adds on aerosol-therapy by inhalation of jet-nebulized saline increases sputum volume and clearance. There is no consensus regarding the indication for surgical intervention in MLS, especially in children.(5)

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Children with right middle lobe syndrome unresponsive to medical treatment should undergo early lobe resection to avoid serious complications and the progression of the disease to other segments or lobes.(8)

In conclusion, we describe a case of severe asthma worsen by the presence of tracheobronchomalacia and Middle Lobe Syndrome. Actually she is treated with maintenance therapy (CSI and oral leukotriene-receptor antagonists) and short-acting β2 agonists and antibiotics, only if acute asthma or bacterial infection are present or suspected respectively. She also practices physiotherapy with PEEP-mask twice a day and Continuous-Positive-Airway-Pressure ventilation therapy during the night for the major risk of the airway collapse and sleep apnea during the night. She is monitored in clinic every three months.

Thinking of MLS is a prerequisite for diagnosis, especially when there is a history of recurrent respiratory infections or asthma, and when clinical or radiological findings are predominantly localized to the right lung. Chest X-ray is the first-line diagnostic tool, and the lateral view should be requested.

Conflicts of Interest: There is no potential conflict of interest, and the authors have nothing to disclose. This work was not supported by any grant.

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