Mounier Kuhn Syndrome Presenting with Recurrent Atelectasis

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Abstract

Context: Mounier Kuhn syndrome is usually diagnosed in adulthood, and only a few cases have been described in childhood.

Case report: We present the case of a seven-year old boy suffering from recurrent pneumonia and atelectasis. Previously performed chest X-rays showed bilateral hyperinflation and tracheobronchomegaly. Chest computed tomography (CT) confirmed the presence of distal enlargement of trachea and bronchi. Tracheobronchomegaly associated with recurrent respiratory tract infections is consistent with Mounier Kuhn syndrome. Pseudomonas aeruginosa was isolated from the sputum of the patient. He was then treated according to the guidelines for Pseudomonas aeruginosa management in cystic fibrosis patients considering the similarities in clinical presentations and pathophysiology of both diseases. Antibiotic treatment resulted in a remarkable reduction of events of pulmonary exacerbation and hospitalizations. There are no specific guidelines for treatment options in case of pulmonary exacerbation of Mounier Kuhn syndrome. Case reports discussing the choice and efficiency of antibiotic treatment are random.

Conclusion: We share our experience of treating pulmonary exacerbation caused by Pseudomonas aeruginosa in a patient with Mounier Kuhn syndrome suggesting a possible treatment option of pseudomonas infections in this syndrome.

Keywords: Mounier Kuhn Syndrome; Pseudomonas aeruginosa; Tracheobronchomegaly

Introduction

Enlargement of the proximal airways, mainly trachea and large bronchi, was first described in 1932 by Mounier Kuhn [1]. This tracheobronchomegaly, associated with recurrent airway infections, is due to a thinning of the muscular mucosa, as well as an atrophy of longitudinal muscles and elastic fibers [2]. Three different types are defined [3]: type 1, the most common, with symmetrical diffuse enlargement of both trachea and main bronchi; type 2 with eccentric enlargements, diverticula and abrupt changes of the bronchial diameter; and type 3, the least frequent, with diverticula extending to the more distal bronchi. Etiology of these modifications is not yet known. Prevalence is estimated between 0.4-1.6% in patients with respiratory symptoms, and there is a strong male predominance with a ratio of 8 to 1. It is usually diagnosed in adulthood [2,4-6], and only a few cases have been described in childhood. We report a case of Mounier Kuhn syndrome in a young child with early respiratory symptoms. In addition, we would like to highlight the fact that large airway disease is usually overlooked initially at chest X-rays.

Case Report

A seven-year old boy was referred to our hospital for recurrent respiratory infections with persistent atelectasis. From the age of 5 months on he presented with wheezing and chronic cough. He was diagnosed with asthma based on clinical manifestations and positive skin prick test to house dust mite. The lung function test...
did not show any sign of obstructive or restrictive disease but a
discrete notch in the flow volume loop. A treatment of inhaled
bronchodilators and corticosteroids in addition to leukotriene
antagonists was initiated. Nevertheless, the boy suffered
persistent chronic cough and respiratory infections associated
with recurrent atelectasis of the right middle and left lingual
lobe. He was hospitalized 4 times in two years for exacerbation
requiring non-invasive respiratory support. Complementary
investigation included normal values of immunoglobulins A,
M, G, total hemolytic complement (CH50) and its fractions C3
and C4, immunophenotyping, and pneumococcal antibodies.
There was no sign of reflux or cardiac disease. Sweat chloride
level was normal and the 35 most frequent mutations of the
cystic fibrosis transmembrane conductance regulator (CFTR)
gene were excluded. At the time he was admitted to our center,
physical examination was normal. Reviewing the previously
performed chest X-rays we noted bilateral hyperinflation and
physical examination was normal. Reviewing the previously
performed chest X-rays we noted bilateral hyperinflation and
tracheobronchomegaly (Figure 1). Chest computed tomography
(CT) (Figure 2) confirmed the presence of distal enlargement
of trachea and bronchi. The sagittal diameter of the carina
measured 20 mm and the left and right bronchi measured 13 mm.
There was evidence of middle lobe atelectasis and segmental
atelectasis of the anterior upper left lobe. No bronchiectasis was
noted. Bronchoscopy confirmed tracheobronchomegaly of the middle
and lower third of the trachea in association with bronchomegaly
of the main bronchi. There was evidence of tracheal and bronchial malacia as well as presence of purulent secretions.
Bronchoalveolar lavage revealed Staphylococcus aureus.
Tracheobronchomegaly in association with recurrent respiratory
trace infection is consistent with Mounier Kuhn Syndrome.
Treatment with physiotherapy and inhaled hypertonic saline
was initiated to improve airway clearance. Clinical evolution
was characterized by several pulmonary exacerbations treated
by oral antibiotics. Azithromycin treatment three times a week
was added to his treatment plan for 1 year to reduce pulmonary
inflammation. Still, there was no significant reduction in
frequency of pulmonary exacerbations and hospitalization stays.
Pseudomonas aeruginosa was isolated in sputum and treated with
oral ciprofloxacin during 3 weeks at a dose of 30mg/kg/day. During
one year follow-ups repeated sputum analysis was negative
for Pseudomonas aeruginosa. The patient is considered to be eradicated until present. The year after eradication the patient
presented with significantly less pulmonary exacerbations and
did not need any hospitalization. Therefore, we consider the
applied Pseudomonas aeruginosa eradication to be causative for
this remarkable clinical improvement.

Discussion

Mounier Kuhn syndrome is characterized by abnormal
enlargement of the major airways. The primary respiratory
symptoms are usually non-specific such as cough, dyspnea,
or recurrent respiratory infections [5]. Radiologic features
might be seen on chest X-rays but are very often overlooked as
described in our case. Since 1988 chest CT is the gold standard
to confirm the diagnosis [7]. Magnetic resonance imaging (MRI)
does not have any supplementary advantage in diagnosis.
Bronchoscopy can be difficult to perform because of important
airway obstruction due to tracheomalacia. Spirometry can show
different degrees of obstruction or increased residual
function. Differential diagnosis should consider William Campbell
syndrome, Ehlers-Danlos syndrome, or Marfan syndrome. Most
frequent pulmonary complications are bullous emphysema,
aspergillosis and pneumothorax. Management is based on
supportive treatment with physiotherapy and inhaled mucoytic
therapy [5]. Pneumococcal and seasonal influenza vaccination
is useful. Appropriate antibiotic therapy in case of pulmonary
exacerbation might be guided by guidelines of non-cystic fibrosis
bronchiectasis [8]. Non-invasive ventilator support can help to
cope with the malacia. Surgical treatment of the tracheomalacia
by tracheobronchoplasty or the use of airway stents has been
described. Cases of Mounier Kuhn syndrome with a wide variety
of clinical presentations and treatment options have been
reported – including patients with Pseudomonas aeruginosa [9].
However, in our case, the patient did not show any bronchiectasis.
Impaired airway clearance in Mounier Kuhn syndrome caused by expiratory airway-collapse due to tracheobronchomegaly in association with chronic inflammation might promote this colonization. As no specific guidelines for the treatment of pulmonary exacerbation in Mounier Kuhn syndrome exist, we decided to treat our patient with oral antibiotic therapy according to guidelines of *Pseudomonas aeruginosa* management in cystic fibrosis patients because of its similar pathophysiology. We were not able to add inhaled antibiotics because of its high cost and lack of mutual refund as recommended by *Pseudomonas aeruginosa* guidelines [10,11]. Our case highlights the value of standard chest X-ray which can still offer valuable information in many diseases and, in some cases, avoid numerous and unnecessary complicated investigations. This case report of a rare disease also gives us the opportunity to share our experience of treating a patient with Mounier Kuhn syndrome and in particular a pulmonary exacerbation caused by *Pseudomonas aeruginosa*.

**Take home message**
- Clinical diagnosis of Mounier Kuhn syndrome is still complex and is frequently overlooked in chest X-rays.
- Patients with Mounier Kuhn syndrome might be colonized by *Pseudomonas aeruginosa*.
- Establishing treatment options especially during exacerbation with *Pseudomonas aeruginosa*. 
References