Vol 6 | Issue 2 | 2016 | 94-97.

e-ISSN 2248 – 9142
print-ISSN 2248 – 9134

International Journal of
Current Pharmaceutical & Clinical Research

www.ijcpcr.com

THE IMPORTANCE OF ADEQUATE BRONCHIECTASIS TREATMENT

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ABSTRACT

Kartagener syndrome is a rare autosomal recessive genetic disease, progressive damage to the respiratory system and situs inversus. Although patients with Kartagener syndrome management remains unclear, and the evidence is limited, but it is important to follow up these patients, appropriate and common supervisory framework to prevent future lung damage. In this case report, we announce a 25 year old woman with a diagnosis of Kartagener syndrome, committed 14 years ago. In 2015, patient was tested for cystic fibrosis and delF508 mutation in heterozygous state was found. The patient was under the supervision of the various areas of the respiratory clinic doctors. Daily physiotherapy, long-acting bronchodilators and appropriate antibiotic treatment has been used for treatment. After 7 years of treatment we have a good treatment results for lung function and radiological data is stable.

Key words: Bronchiectasis, Diagnosis, Health.

INTRODUCTION

Kartagener syndrome is an autosomal recessive disorder first manifests as ciliary dyskinesia. (1). Kartagener syndrome is part of a larger group of disorders known as primary ciliary dyskinesia (PCD). Although the condition is usually inherited as an autosomal recessive and some specific genetic defects have been recognized, it is clear that the syndrome shows considerable genetic heterogeneity (2). From a genetic disorder frequency is between about 1 in 15,000 and 1 in 30,000 (3). Symptoms appear defective cilia motility. Ciliated cell line from nasopharyngs, middle ear, paranasal sinuses, and upper respiratory tract respiratory tract from the terminal respiratory tract (2.3) trachea. Recurring lung infections, caused by a compromised mucociliary transport respiratory tract caused by stasis of mucus through the bronchial tubes (1, 4). The progressive and significant lung damage occurs to the time of diagnosis (3,4). These three lower respiratory tract disease has been described in older children and adults with primary ciliary dyskinesia pneumonia; bronchiectasis; and asthma (5). Although the management of patients with Kartagener syndrome remains unclear, it remains important Controle chronic lung infections and lung function deteriation.

Case report

We present 25-year-old woman with Kartagener syndrome case. The patient complained of cough and sputum. The current general physical examination revealed pulse of 80 / min, blood pressure was 110/70 mm Hg, respiratory rate was 18 / min. An examination of the airways revealed coarse crepitations with scattered rhonchi all over the chest. Apex beating was palpable on the right side to the right side of the chest. Liver was palpable left hypochondrium. Chest computed tomography (CT) showed dextrocardia and bilateral bronchiectasis (Figure 1). Spirometry showed a stable first degree of bronchial obstruction: FEV1 -pred. 93% (3.27l) FTC - 110% (4.40l), FEV1 / VC - (. 71% pred) 83%. It was very suprising that at 2015, patient was tested for cystic fibrosis and delF508 mutation in heterozygous state was found.

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Disease history

When she was 14 years old Kartagener syndrome was diagnosed. At the time, spirometry showed first degree of bronchial obstruction: FEV1 - 88% (3.08l), FVC - 111% (4.45l), FEV1 / VC - 82% (69% pred.). Chest X ray showed bilateral lower zone peribronchial cuffing with dextrocardia. At the end of 18 years of age was under adult pulmonologists and geneticists Kaunas Medical University Hospital. Here, computed tomography (CT) chest identification of dextrocardia, pulmonary fibrosis, multiple bronchiectasis upper lobes (Figure 2). Fixed inhaled long-acting bronchodilators (b2-agonists) have been used in the treatment of bronchial obstruction. Treatment of severe or persistent exacerbations of bronchiectasis with short antibiotic and mucolytics (bromhexine) courses of 10-14 days has been effective. Physiotherapy and exercise was daily, optimized nutrition, environmental pollutants (including tobacco smoke) to avoid. All of these methods to reduce symptoms and levels of inflammatory markers and improve quality of life. Subsequently Spirometry was measured several times once a year. Lung function did not deteriorate during the observation period of 7 years: Chest CT was used to monitor the progression of the disease within two years. Radiological findings are stable even with mild regression of bronchiectasis (Figure 1).

Figure 1. Chest CT at the age of 25.

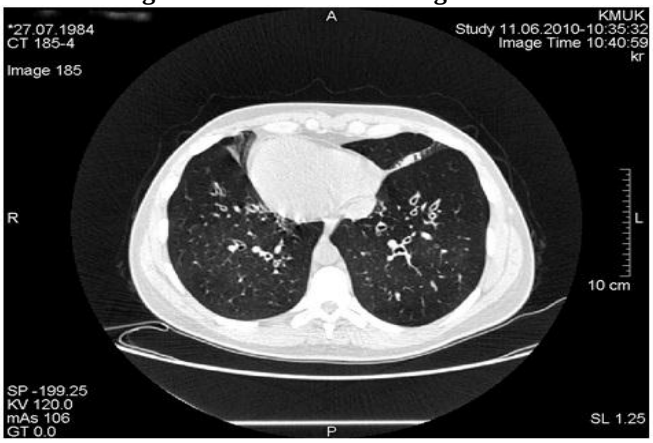
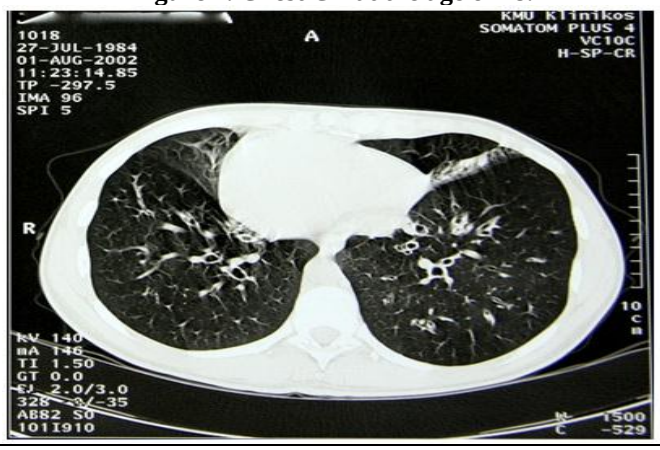


Figure 2. Chest CT at the age of 18.



DISCUSSION

Mucociliary escalator is the primary defense mechanism of the respiratory tract and any functional impairment, congenital or acquired, can lead to chronic sino-pulmonary symptoms (3). Clinical symptoms of Kartagener syndrome is a productive cough, respiratory tract infections, sinusitis, middle ear inflammation and infertility (6, 7). Aristocardia generally not accompanied heart abnormality when it occurs with situs inversus (6). The clinical phenotype of PCD is extensive and coincide with other chronic respiratory diseases (7). The defect is inherited and symptoms of early life, which highlights the importance of pediatricians know how this disorder although rarely appropriate differential diagnosis in children with recurrent symptoms of upper and lower respiratory tract. Especially if early diagnosis and intensive treatment is available, forecast to maintain lung function seems well (4). Kartagener syndrome in our patient was diagnosed at 14 years of PCD diagnosis is often delayed until late childhood or adulthood as a consequence of the heterogeneous nature of the disease, the disease characteristics of medical knowledge and technical expertise required for the lack of accurate diagnosis (3, 7). Also, the PCD diagnosis may be delayed because of bronchitis, sinusitis and otitis syndrome is easily mistaken for common infections. The delay in recognizing the disease can lead to negative consequences for patients in terms of inappropriate or improper maintenance programs of treatment (7). It should be taken into consideration in the differential diagnosis of PCD patients with chronic infections of the respiratory tract. Diagnostic suspicion of PCD increases the likelihood of some patients, chronic respiratory infections since birth and present situs inversus (8).

During airway pharmacotherapy such components are very important:

1) Bronchodilators

Although a large proportion of subjects with bronchiectasis have airflow obstruction with airway hyperreactivity and a significant bronchodilator response, there are no randomized, controlled trials investigating the effects of long-acting beta-agonists or anticholinergics in the management of patients with bronchiectasis.

Tiotropium bromide, a long-acting muscarinic receptor antagonist, relaxes airway smooth muscle cells and suppresses airway submucosal gland secretions. Nowadays, it is widely used as one of the important medications in patients with chronic obstructive pulmonary

disease, but the efficacy in bronchiectasis has not been studied adequately. In a small open label Japanese study, tiotropium improved symptoms of cough, sputum, and dyspnea in patients with chronic mucus hypersecretion, but too small numbers of patients with bronchiectasis were included. A recent trial in Chinese showed that one month of inhalation of tiotropium improved the clinical symptoms and body mass index, airflow obstruction, dyspnea, and exercise capacity (BODE) index of the patients with bronchiectasis. Randomized, controlled trials will be needed to evaluate the effectiveness of long-acting beta-agonists and anticholinergics in the treatment of bronchiectasis.

2) Inhaled corticosteroids

Inhaled corticosteroids have proved undeniable benefits in patients with asthma or chronic obstructive pulmonary disease, but very little is known of its anti-inflammatory effects in bronchiectasis. Several small scale studies have observed that in patients with non-cystic fibrosis bronchiectasis, high doses of inhaled corticosteroids can positively influence several bronchial inflammatory parameters and certain key symptoms, such as dyspnea or sputum volume, while improving patients' health-related quality of life, without affecting either the number of exacerbations or lung function. However, a Cochrane review concluded that there was insufficient evidence to recommend the routine use of inhaled corticosteroids in adults with stable-state bronchiectasis, and that a therapeutic trial may be justified in adults with difficult to control symptoms and in a certain subgroups.

Meanwhile, the addition of a long-acting beta-agonist to inhaled corticosteroids proved to reduce the dose of inhaled corticosteroids without effects of treatment in chronic obstructive pulmonary disease or asthma39. In actual practices, it is prescribed to a high percentage of patients with bronchiectasis without any clear scientific evidence. A recent, small-sized, 12-month randomized trial revealed that inhaled medium-dose formoterol-budesonide combined treatment is more effective in symptom control and health-related quality of life compared with high-dose budesonide treatment in patients with non-cystic fibrosis bronchiectasis. Largerscale studies of longer duration are needed to confirm this result.

3) Inhaled antibiotics

There are a substantial number of literatures on the use of prolonged antibiotics in patients with bronchiectasis, because reducing the bacterial burden in the airways may decrease inflammation and promote healing of the bronchial tree. These studies showed small benefit in response rates and sputum scores, but did not show any differences in exacerbation rates, lung function, or quality of life scores. Similarly, there are also a large number of publications concerning the use of inhaled antibiotics (mainly tobramycin and gentamicin) in patients with bronchiectasis, particularly in the setting of Pseudomonas aeruginosa infection. Some benefits have been documented in these studies, including a decrease in bacterial density, airway inflammation, and exacerbation frequency. However, the benefits appear to be less than in cystic fibrosis cases, and bronchospasm appear to be more common in adults with non-cystic fibrosis bronchiectasis than reported in the cystic fibrosis population, and treatment needs to be continuous for its ongoing efficacy44. More recently, new inhaled agents such as liposomal ciprofloxacin and liposomal amikacin are also being investigated for potential use in bronchiectasis.

Pulmonary disease progression varies and affects the diagnosis, treatment ability to control the symptoms and complications that affect the life (7) the quality controls. Lung damage is caused by undertreatment of recurrent pulmonary infections and seems to be preventing an accurate diagnosis and appropriate clinical care. Management is not evidence-based. Respiratory management consists of regular respiratory monitoring, airway clearance to a combination of physiotherapy and physical exercise, and aggressive behavior of the upper and lower respiratory tract infections (9). There are currently no treatments that have been tested sufficiently de fi nitively prove their ef fi cacy in the treatment of PCD. Our patient received a wide range of healing techniques: Regular courses of antibiotics, mucolitics, long bronchial and dayly physiotherapy. In general, antibiotics are used for acute exacerbations of the disease and is found by sputum culture grown in the last bacteria. The goal of treatment should be chronic lung injury and bronchiectasis. (10) Two pillars are used to treat respiratory therapy with antibiotics and chest physiotherapy. For a long time, high-dose oral antibiotics should be administered at the first sign of any respiratory symptoms increase or worsening of lung function (11). Physiotherapy is therefore imperative to develop airway clearance in order to delay the onset and progression of obstructive airways disease (10, 11). Physical exercise can help sputum clearance. Exercise has been shown to be better than bronchodilation bronchodilator use PCD (9). Prognosis is generally considered good, with usually a normal life expectancy.

Also because of disease low incidence, we can assume that there is only a slight chance of these two rare diseases (Kartagener syndrome and cystic fibrosis carrier) occurs together in the same patient.

CONCLUSIONS

Monitoring the progression of lung disease should be an important part of a regular clinic visit (3). Our patient was followed for regular visits every 6 months, despite additional visits during exacerbations. Early diagnosis and management of chest infections can prevent permanent damage to the lungs and to avoid possible complications. In our paper we wanted to draw the proper management of even non-specific innate Kartagener syndrome can be

effective, with a positive impact on the course of the disease.

ACKNOWLEDGEMENT: None

CONFLICT OF INTEREST:
The authors declare that they have no conflict of interest.

STATEMENT OF HUMAN AND ANIMAL RIGHTS

All procedures performed in human participants were in accordance with the ethical standards of the institutional research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. This article does not contain any studies with animals performed by any of the authors.

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