Kartagener’s Syndrome: A Case Report

PM Basak¹, MN Islam¹, BC Sarkar¹, MA Islam¹, HM Rashed², Jafor Islam³, HS Das⁴

Abstract

Key words: kartagener syndrome; respiratory tract infection; bronchiectasis; therapeutics; fertility.

Introduction

Kartagener’s syndrome is a recessive autosomal disease which is mainly seen to affect ciliary movement¹. The incidence of Kartagener’s syndrome is 1–2/30,000 births. Siewert first described the combination of situs inversus, chronic sinusitis and bronchiectasis in 1904². Manes Kartagener, a pulmonologist, first recognized this clinical triad as a distinct congenital syndrome in 1933. Kartagener described this syndrome in detail, so it bears his name³. The symptoms of the syndrome are the consequence of the defective motility of the cilia found in the respiratory tract and that results with recurrent lung infections caused by mucus stasis in the bronchi¹;³. In older children and adults with primary ciliary dyskinesia,³ diseases of the lower respiratory tract have been described: pneumonia, bronchiectasis and asthma⁴. Patients with Kartagener syndrome may have immotile spermatozoa as well⁵.⁶. Treatment for patients with this syndrome has not been established, but it is important to control chronic lung infections and prevent declining of lung function⁵;⁷.

Case Report

A 35-year-old female, married, mother of one child, presented with the history of productive cough, hemoptysis, wheezing, dyspnea, headache and occasional fever. At clinical examination, the patient is ill looking, anxious, emaciated. There is generalized clubbing present in both toes and fingers. Crepitations and rhonchi were evidenced in both lung field.

A chest X-ray showed dextrocardia, Chest CT revealed dextrocardia, bronchiectasis, left liver, and spleen on the right.

Figure-1

¹ Assistant Professor, Department of Medicine, Rajshahi Medical College.
² Assistant Registrar, Department of Medicine, Rajshahi Medical College Hospital.
³ Indoor Medical Officer, Department of Medicine, Rajshahi Medical College Hospital.
⁴ Lecturer, Department of Community Medicine, Rajshahi Medical College.
Fig. 1 – The Patient
Fig. 2 - X-ray PNS Show Maxillary Sinusitis.
Fig. 3 – Contrast-enhanced computed tomography images show dextrocardia and bronchiectasis in the lower lobes, dominantly on the left side.

Echocardiography confirmed dextrocardia, too. A PNS radiograph shows bilateral maxillary sinusitis. The patient is treated with a combination of inhaled bronchodilators (inhaled corticosteroids + long-acting beta-2 agonists), and occasional administration of antibiotics, oral prednisolone, mucolytics in episodes of exacerbations of disease over a period of 7–14 days.

Discussion
Kartagener’s syndrome is seen in 50% of patients with primary ciliary dyskinesia. Kartagener’s syndrome is characterized characterized by situs inversus, bronchiectasis, sinusitis and otitis media. In some cases it is inherited with bronchial asthma. Our case is unusual for one reason. The presented patient was fertile and had a child. Munro et al.⁹ show that 30% of their patients with primary ciliary dyskinesia have normal fertility. The presented clinical case demonstrated a progressive course of bronchiectasis, declining lung function, because of recurrent infections which were treated inappropriately in recent period.

Conclusion
We presented this case because the Kartagener’s syndrome is a very rare condition. The prognosis is generally considered favorable, and life expectancy is usually normal. An important part of the clinical visits at regular intervals should be monitoring the progression of the lung disease.

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All corresponds to

P M Basak
Assistant Professor
Department of Medicine
Rajshahi Medical College, Rajshahi, Bangladesh
e-mail: prabirbasak84@yahoo.com