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# Multiple Lung Abscesses in a Patient with Kartagener's Syndrome

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ABSTRACT

*Kartagener's syndrome is a rare genetic disorder, which is mostly inherited as an autosomal recessive trait. There are 4 genes with a proven pathogenetic role in Kartagener's syndrome. It results from ciliary dysfunction and is commonly characterized by sinusitis, male infertility, hydrocephalus, and situs inversus. Since Kartagener's syndrome causes deficiency or even stasis of the transport of secretions throughout the respiratory tract, it favors the growth of viruses and bacteria. As a result, patients have lifelong chronic and recurrent infections, typically suffering from bronchitis, pneumonia, hemoptysis, sinusitis, and infertility. We present a 27-year-old woman, a case of Kartagene'sr syndrome, with multiple pulmonary abscesses. Evaluation of sputum and tracheal secretions revealed Pseudomonas aeroginosa. Antibiotics were started and respiratory symptoms resolved and the patient was discharged in good general condition. As a conclusion, prompt and appropriate treatment of respiratory infections can minimize irreversible lung damage in such cases.* ***(Tanaffos 2008; 7(4): 64-67)***

***Key words:*** *Kartagener's syndrome, Lung abscess, Pseudomonas aeroginosa*

## INTRODUCTION

Primary ciliary dyskinesia is a rare etiology of sterility in men with prevalence between 1/6000 and 1/40000. Kartagener's syndrome (KS) is an autosomal recessive disorder, characterized by total or partial dysfunction of the ciliary or flagellated cells. This syndrome is associated with situs inversus, sinusitis, bronchiectasis and occasionally sterility in males (1). Patients develop respiratory infections due to ciliary dysfunction and stasis of secretions in the respiratory tract. Although pneumonia is frequently seen, lung abscess is rare in Kartagener's syndrome.

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## CASE SUMMARIES

A 27-year-old woman, a known case of Kartagener's syndrome referred complaining of fever, productive cough and dyspnea started one week prior to admission. She was diagnosed as having Kartagener's clinically several years ago, as she had chronic sinusitis, recurrent pneumonia, bronchiectasis and dexrtocardia. The patient had

taken ceftriaxone and metronidazole without improvement and was referred to our hospital for further evaluation.

Right lower lobectomy had been done for her 12 years ago due to destructive changes in the right upper lobe discovered on chest CT-scan after recurrent episodes of pneumonia. Since then, she has been stable. About 8 months prior to admission she developed fever and productive coughs. With antibiotic therapy, the symptoms disappeared but returned several weeks later. Her recent condition started with fever, dyspnea and productive coughs and was more severe than previous attacks. She was treated with ceftriaxone, metronidazole, hydrocortisone and aminophylline with no significant improvement.

Family history revealed that her parents were cousins and both were healthy. They had 5 children; 2 of them were healthy, 2 had diabetes melitus, seizures and deafness, and one suffered from asthma.

On physical examination she was febrile; T=38.5˚C, RR=22/min, PR=90/min, BP=120/80. Tenderness over maxillary sinuses was detected bilaterally. She had post nasal discharge. There was no cervical lymphadenopathy. On chest auscultation, fine crackles were detected bilaterally. Heart sounds were normal and point of maximal intensity (PMI) was detected on the right side in favor of dextrocardia. Abdomen, extremities and neurologic exams were normal. Laboratory data were as follows :

WBC=10500 (PMN=85%), Hgb=13 g/dl, PLT=353000, Na=145 meq/dl, K=4.1 meq/dl, ESR=25 mm, BUN=12, Cr=0.67, ALT=13, AST=12.

Smears and cultures of the sputum and tracheal secretions taken three times showed *Pseudomonas aeroginosa*. Sputum smears showed more than 25 PMNs, less than 10 epithelial cells and a large number of gram-negative organism under low-power microscopy. Culture of sputum showed *Pseudomonas aeroginosa*. Culture of blind endotracheal secretion showed more than 100,000 colony count of *Pseudomonas aeroginosa*. Sputum smear was negative for acid-fast bacilli (AFB). Electron microscopic study and gene sequence analysis were not done for confirmation of diagnosis. Chest x-ray showed bilateral infiltration and multiple air-fluid levels. Dextrocardia was seen on the CXR (Figure 1).

Lung CT-scan showed ground glass infiltration in upper and middle zones of both lungs, more prominent on the left side, emphysematous bullae in the apical part of both lungs, severe tubulosacular bronchiectasis and multiple air-fluid levels (Figure 2, 3). CT-scan of the paranasal sinuses showed inflammatory changes in maxillary, ethmoid and sphenoid sinuses.

Echocardiography showed mild right atrial and ventricular enlargements (RAE, RVE) and mild tricuspid regurgitation (TR).

Ceftazidime, amikacin and clindamycin were started to treat *Pseudomonas aeroginosa*. Conservative treatment with hydration, mucolytic agents and chest physiotherapy was also started. After 48 hours, fever disappeared and the patient's general condition gradually improved. After completion of the course of treatment she was discharged in a good general condition.

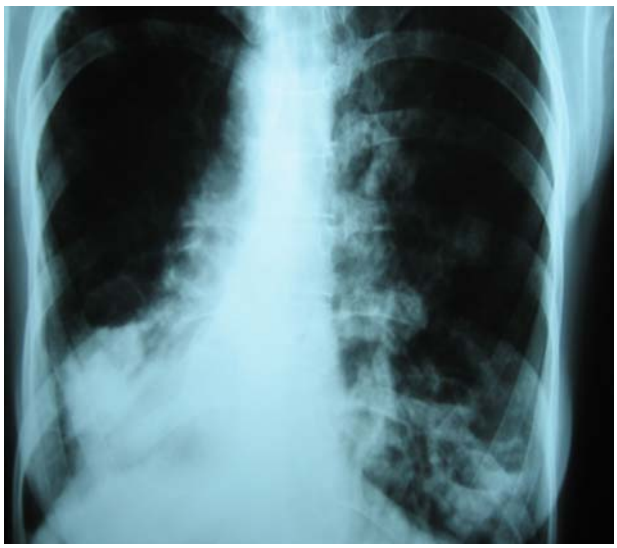

Figure 1. CXR of patient.

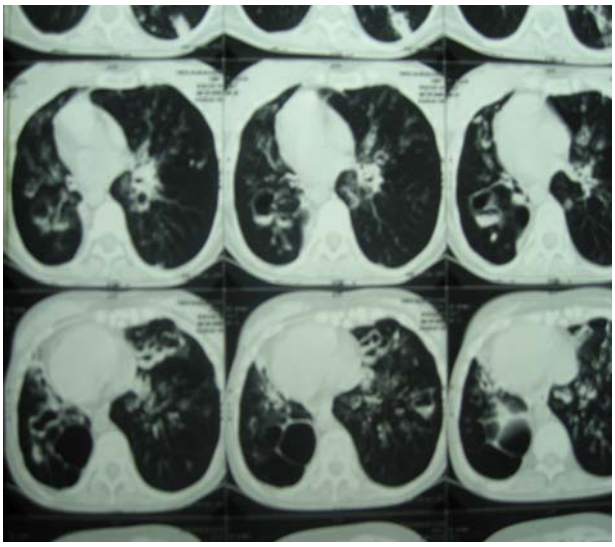

Figure 2. Chest CT scan.

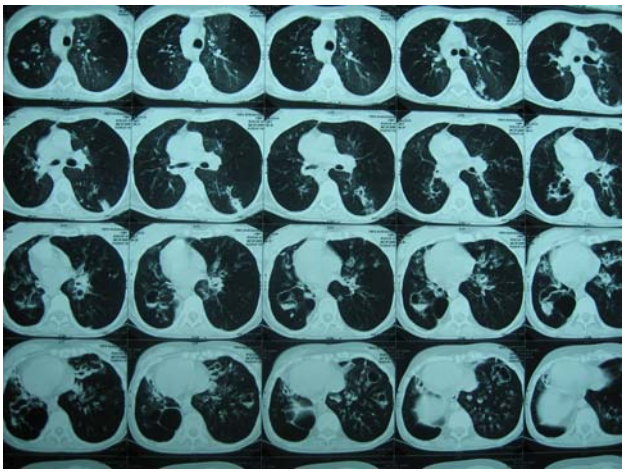

Figure 3. Chest CT scan.

## DISCUSSION

Primary ciliary dyskinesia (PCD) is a rare genetic disorder, which shows extensive genetic heterogeneity and is mostly inherited as an autosomal recessive trait. There are four genes with a proven pathogenetic role in PCD (2). This syndrome is commonly characterized by sinusitis, male infertility, hydrocephalus, and situs inversus (3). The incidence of PCD ranges from 1:20,000 to 1:60,000. Since PCD causes deficiency or even stasis of secretions throughout the respiratory tract, it promotes growth of viruses and bacteria. As a result, patients have lifelong chronic and recurrent infections, typically suffering from bronchitis, pneumonia, hemoptysis, sinusitis, and infertility. Bronchiectasis and other chronic infections can result from irreversible bronchial alterations, leading to chronic cor pulmonale and related complications. Only half of the patients affected by PCD present with all the symptoms (designated complete Kartagener's syndrome). Incomplete KS is defined as cases who do not have situs inversus (4). Our patient did not have situs inversus; only dextrocardia was detected.

Lung CT-scan demonstrates peribronchial thickening and bronchiectasis, which is mostly marked in the lower zones (5). High-resolution CT shows that pulmonary disease related to PCD predominantly involves the middle and lower lobes of the lungs (6). Although pneumonia is frequently seen in Kartagener's syndrome, lung abscess infrequently accompanies it (7). In our patient multiple air-fluid levels were detected on CXR and abscesses were confirmed by lung CT scan.

Cutaneous lesions in Kartagener's syndrome are nummular eczema, recurrent deep folliculitis and pyoderma gangraenosum (8). No cutaneous lesions were seen in our patient.

The diagnosis of KS is made clinically and confirmed through transmission electron microscopy (4). Screening tests for PCD include nasal nitric oxide and in vivo tests of ciliary motility such as the saccharin test. Specific diagnosis requires examination of cilia by light and electron microscopies, with epithelial culture in doubtful cases (9). Since there is no specific therapy for PCD, it is recommended that, upon diagnosis, secondary infections be treated with potent antibiotics and prophylactic interventions (10).

In a conclusion, prompt and appropriate treatment of respiratory infections can minimize irreversible lung damage in such cases.

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