

CASE REPORT**KARTAGENER'S SYNDROME: A CASE REPORT**Samuel Sisay Hailu¹, Eyoel Degafu Amerga¹, Yocabel Gorfu,¹ Daniel Zewedneh^{1*}**ABSTRACT**

Kartagener's syndrome is a rare autosomal recessive genetic disorder that causes a defect in the action of the cilia lining the respiratory tract. Patients usually present with chronic recurrent rhinosinusitis, otitis media, pneumonia, and bronchiectasis caused by Pseudomonas infection. In this case report, we present a 12-year old female with Kartagener's syndrome from Tikur Anbessa Teaching Hospital which to our knowledge is the first of it's kind to be reported in Ethiopia. The clinical and imaging findings are discussed.

Key words: *Kartagener's syndrome, Situs inversus, Dextrocardia, Allergy, Bronchiectasis, Sinusitis.*

INTRODUCTION

Kartagener Syndrome's (KS) is a rare autosomal recessive disorder characterized by dextrocardia, bronchiectasis and sinusitis. The condition was described for the first time by Siewert in 1904; therefore, some people have referred to it as Siewert Syndrome but the details of the condition were given by Manes Kartagener in 1933 and it is commonly known as Kartagener's syndrome. The basic defect is inefficient or unsynchronized movement of the cilia leading to impaired mucociliary clearance of respiratory secretions. Males are typically infertile due to dysfunctional sperm, though some males have completely normal spermatozoa. Cases of semi-sterility in females have been reported. Patients with KS occasionally present with anosmia (1).

CASE REPORT

A 12-year old female presented to Tikur Anbessa Teaching Hospital on June 30, 2015. The patient presented with productive cough, rhinorrhea and headache since childhood with episodic fever and worsening of symptoms.

Her record showed numerous laboratory as well as radiologic investigations including serial chest x-rays, a chest CT and AFB sputum examination. Her record also revealed that she had been treated with different courses of antibiotics. She also completed anti-tuberculosis treatment with good adherence three years prior to her current presentation but had no significant clinical improvement.

On current presentation her exam was remarkable for yellowish nasal discharge, decreased air entry in the left mid lung fields, cyanosis, digital clubbing (Fig 1), and normal S1 and S3 heart sounds best observed on the right side of the chest. All vital signs were in the normal range. All laboratory findings were in the normal range.

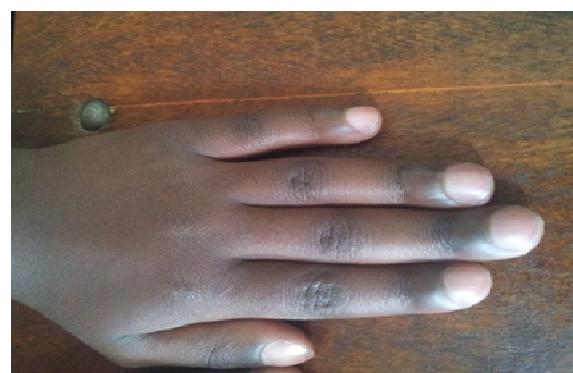


Figure 1: Photograph of the patient's hands showing clubbing of the fingers (right & left hands).

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The chest x-ray showed features suggestive of situs inversus, and left paracardiac bronchiectasis and fibrotic changes; these were persistent findings on serial chest x-rays. The cardiac apex and the aortic arch were located on the right side (Fig 2). Echocardiography was normal.

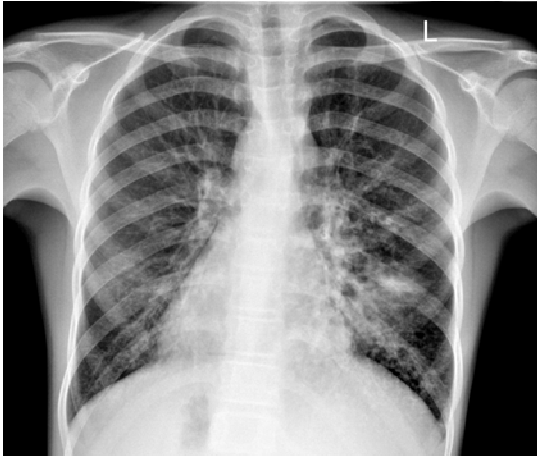


Figure 2: PA chest X-ray showing dextro-cardia, left paracardiac bronchiectasis and fibrotic change. Note the correct side marker on the right upper corner.

Paranasal sinus Water's view x-ray showed opacified maxillary sinus and non-visualized frontal sinuses



bilaterally (Fig 3) suggesting chronic sinusitis.

Chest CT showed bronchiectatic change with surrounding consolidation in the lingular segment of the left lung and situs inversustotalis with the aortic arch and cardiac apex projecting in the right hemithorax, homogeneously enhancing liver with patent vessels present in the left upper quadrant, and normal spleen in the right upper quadrant (Fig 4). Considering the clinical and radiological presentation of sinusitis, bronchiectasis and situs inversus the diagnosis of Kartagener's syndrome was made.

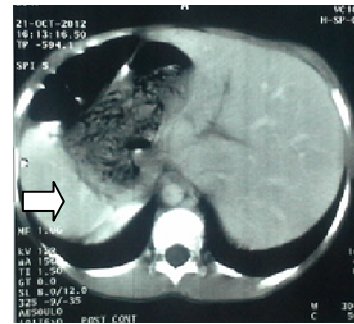
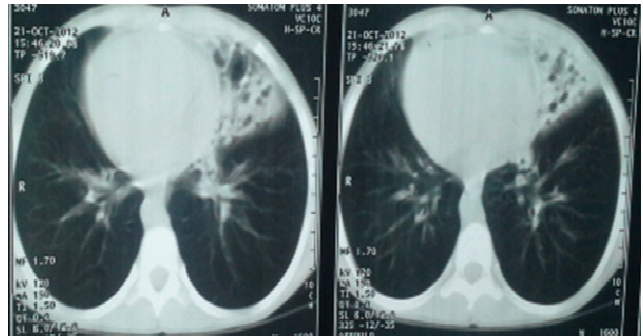


Figure 4: Axial post contrast CT showing (a) bronchiectatic change with surrounding consolidation and (b) situs inversustotalis of the liver (star) on the left and spleen (arrow) on the right side.

DISCUSSION

KS is an autosomal recessive inherited disease, with a prevalence of 1 case in 16,000 births. It consists of a triad of symptoms, situs inversus (mirror image organ placement), bronchiectasis and sinusitis. Patients usually present with high incidence of respiratory infections (2, 3). Our patient had all the components of the triad. The imaging findings on chest radiography, CT (both chest & abdomen) and paranasal sinus x-ray support the clinical diagnosis of KS (4).

Numerous defects have been described in components which comprise cilia, including lack of or dysfunction of inner and/or outer dynein arms, radial spokes, and microtubules. Consequently, the cilia inadequately clear bacteria from the upper and lower respiratory tract, leading to persistent infections (5, 6, 7).

The diagnosis should be considered in patients presenting with recurrent sinusitis and bronchiectasis, asthma like symptoms and signs responding poorly to conventional treatment, and recurrent lower respiratory tract infections causing fever, sweating and weight loss (1).

Bronchiectasis is much more prevalent in people with situs inversus than in the general population. Some investigations report an incidence of bronchiectasis in 0.3–0.5% of the general population, but the incidence rises to 12–25% in patients with situs inversus (1).

Sinusitis is the other component of the triad, which was quite marked in our patient. Abnormal ciliary movements lead to an accumulation of secretions inside the paranasal sinuses, and can become chronic leading to hypoplasia or even agenesis of the paranasal sinuses. Radiographs of the sinuses may show sinusitis, with opacification of the maxillary, ethmoid and frontal sinuses (6).

The imaging findings on simple radiography include: Situs inversus or dextrocardia (50%), paranasal sinusitis, bronchiectasis, bronchial wall thickening, segmental atelectasis and bronchiectasis (often involving the lower lobes), hyperinflation of the lungs and evidence of recurrent pneumonias. In addition to the findings depicted on radiographs chest CT may show the “signet ring sign”, dilation of bronchi with diameter greater than the accompanying pulmonary artery in which case the “ring” will be the dilated bronchus and the ‘pearl’ the accompanying pulmonary artery (3, 7).

Chest radiography is usually sufficient for confirmation of these features but high-resolution CT scanning may be useful to determine presence and extent of bronchiectasis. CT is more sensitive for recurrent pneumonias especially those involving lower lobes and the right middle lobe. Therefore a thin-section image with widths 1-2 mm at 10 mm intervals without IV contrast is recommended (7).

The differential diagnosis is broad, and includes Young Syndrome, Sinobronchial Allergic Mycosis, Cystic Fibrosis and others. With meticulous clinical evaluation and appropriate imaging findings it is possible to arrive at the right diagnosis (7, 9).

Formal diagnostic criteria for this condition include clinical picture suggestive of recurrent chest infections, bronchitis, and rhinitis since childhood, along with one or more of the following: situs inversus in the patient or sibling, viable but immotile spermatozoa, reduced or absent transbronchial mucociliary clearance, and cilia showing characteristic ultrastructural defects by electron microscopy (3,6).

Treatment of KS is much the same as that for bronchiectasis from other etiologies. The goal of therapy is to reduce symptoms and slow disease progression. Prophylactic measures such as appropriate immunizations, particularly influenza vaccine and pneumococcal vaccine, and vigorous pulmonary toilet are the mainstays of therapy. Acute bouts of bronchitis must be treated with antibiotics. The choice of drug should be based on findings from gram-stained sputum samples. Patients who develop recurrent pneumonia or hemoptysis and do not respond to antibiotics may benefit from segmental lung resection or lobectomy (2).

The long-term prognosis of patients with KS is good, with many patients living to an advanced age. Decreased quality of life is caused by chronic respiratory symptoms (8).

In conclusion, though KS is an uncommon disease entity, the case described here illustrates that it can occur in Ethiopia, and that it should be included in the differential diagnosis of patients presenting with sinusitis, recurrent chest infections, and situs inversus and other characteristic radiological findings. A correct early diagnosis of KS can avoid unnecessary investigations and inappropriate antibiotic treatment, and improve quality of life.

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