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Case Report

A CASE REPORT ON KARTAGENER'S SYNDROME**Mara Kurian^{*1}, Meekha Mary Varghese¹, Haritha B Nair¹, Navya Sunil¹,
Lubna Sulthana¹, Hemalatha S², Sivakumar T³.**¹Pharm D Interns, Nandha College of Pharmacy, Erode, Tamil Nadu, India.²Lecturer, Nandha College of Pharmacy, Erode, Tamil Nadu, India.³Principal, Nandha College of Pharmacy, Erode, Tamil Nadu, India.**Abstract:**

Kartagener's syndrome is a rare congenital disorder consisting of situs inversus, bronchiectasis and sinusitis. It is a subgroup of immotile cilia syndrome called primary ciliary dyskinesia. In this syndrome generally male patients show infertility and reduced fertility in females. Mutation in DNAH5 gene located on chromosome 5p is responsible for this Mendelian disorder. Clinical manifestation involves recurrent respiratory infections and variability with multisystem involvement. Routine radiological tests are usually used for diagnosis. Prompt diagnosis and ideal management can prevent complications.

Keywords: *primary ciliary dyskinesia, situs inversus, bronchiectasis, dextrocardia.*

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INTRODUCTION:

Kartagene's syndrome is an autosomal recessive genetic disorder described as a triad of situs inversus, bronchiectasis and sinusitis. It is recognized as a clean-cut congenital syndrome by Manes Kartagener an internist in 1933, so it bears his name. [1]

The primary problem is defective movement of cilia which causes immotile cilia syndrome (primary cilia dyskinesia), with an incidence of 1 in 20,000-30,000.[2] More than 19 causative genes have been associated with primary ciliary dyskinesia, in which mutation in DNAH5 gene located on chromosome 5p is responsible for Kartagener syndrome.[1] It was observed in consanguinity especially first cousin. [3]

Impaired mucociliary clearance leads to recurrent sinopulmonary infections with varying severity. [3] Nasal related problems can occur in the form of sinusitis and nasal-poliposis and ear involvement as otitis media. Usual chest complaints are chronic productive cough, recurrent pulmonary infections and dyspnoea or wheezing indicates bronchiectatic changes. Haemoptysis may also present in some cases. [6]

Generally, sperm of males with Kartagener syndrome are immotile, due to ciliary dysfunction but spermatozoa have fertility capacity. [3] However a few cases of semi-sterility have been reported in females. Prompt detection and regular treatment can prevent its various complications. [4]

CASE REPORT:

A 28 year old male patient presented with complaints of breathlessness, cough with expectoration, and headache. He had taken treatment for haemoptysis before 1week in another hospital. Previous records showed repeated hospital admissions for recurrent chest infections. Physical examination revealed patient is in respiratory distress and have grade-3 breathlessness.

On general examination his vitals were, respiratory rate-32 breaths/min, heart rate- 120 beats per minute, blood pressure 152/90 mmHg. On auscultation, bilateral wheeze and diffuse rhonchi were audible, more heard in right side of chest. Routine blood investigations were normal except ESR, which is 27mm/hr. Chest X-ray revealed cystic lesions and features suggestive of situs inversus. His abdominal ultrasonography showed situs inversus totalis. On sinus X-ray pansinusitis is evident. High resolution of computed tomography of thorax revealed bronchiectasis in inferior lingula and dextrocardia. Based on clinical presentation, laboratory evaluation

and radiological features, he was diagnosed with Kartagener Syndrome.

Clinician started the treatment with Ampicillin 500mg every 8 hourly intravenously for the eradication of infection. Theophylline (25.3mg), Etophyllin (84.7mg) intravenously and bromhexine syrup helped to relieve respiratory distress and complaints of cough. For headache oral paracetamol 500mg was advised to take whenever needed. Additionally, intravenous normal saline, ringer lactate with multivitamin infusion, vitamin B complex and vitamin C were recommended as supportive therapy. On the second day oral Amoxicillin 250mg was started to take in the interval of 8th hours. After 4 days of treatment, symptomatic improvement was observed. Thus intravenous Ampicillin was stopped and Ampicillin 250mg continued, which is taken every 8th hourly. On next day patient got discharged with the permission of physician. During discharge clinician ordered to continue the treatment with oral Ampicillin 250mg, Theophylline and Etophylline (23+77mg) combination PO daily. Besides this Chlorpheniramine maleate, vitamin B complex and paracetamol (whenever necessary) were advised.

DISCUSSION:

Kartagener's syndrome is a rare Mendelian disorder caused by a defect in dynein arm structure of cilia. As a result, ciliary movement and consequently its function are debilitated. Impaired ciliary function pave to incomplete rotation or malrotation of one or many internal organs, commonly the heart.[5] Sometimes malrotated heart is associated with severe anomalies in vessels connecting the heart have low survival. Generally, dextrocardia (right sided heart) exist along with situs inversus (malrotation of other internal organs namely: liver, lungs, spleen, kidney and intestines). [7] Nasopharynx, middle ear, larynx, paranasal sinus, trachea and bronchi is lined by pseudo stratified ciliated columnar cells. Uncoordinated infective ciliary movement in this syndrome affects mucociliary clearance which can progress to long standing sinonasal, aural and pulmonary problems. [8]

Routine radiological examinations are often used for diagnosis of Kartagener syndrome. Chest x ray reveals dextrocardia and situs inversus. Computed tomography of thorax gives clear picture of bronchiectatic changes. Situs inversus totalis is observed through ultrasonography of abdomen. Computed tomography of paranasal sinuses detects pansinusitis, polyposis or hypoplasia of sinuses. [8] Semen analysis for sperm motility is helpful to clear

out fertility issues. Majority of infertile patient with this syndrome have normal sperm count, but with structural defect. Invariable male patients are infertile, whereas female usually shows reduced fertility. [9]

Treatment is mandatory to relieve symptoms and prevent irreversible complications. A study conducted by Noone et.al reported that 94 patients from 68 family, cough was present in 100% of patients, bronchiectasis (98%), sinusitis (47%), otitis media (92%), and situs inversus (46%). [10] Treatment protocol encompasses daily chest physiotherapy, antibiotic with good pseudomonas coverage, supportive pulmonary care, and some adjunctive therapy. Surgical interventions can be beneficial when disease is localized. [4] For acute bacterial exacerbations antibiotic are intended. Long term antibiotic in low doses intended as prophylactic use. Early rational use of antibiotics can lessen the need of surgery in these patients. Inhaled corticosteroids, mucolytics, bronchodilators are beneficial for symptomatic relief. Surgical treatment in the form of FESS is helpful in patient with nasal polypoid and chronic sinusitis. For respiratory exacerbation with haemoptysis surgical interventions like lobectomy and pneumonectomy are advised. [11] Here in our patient, symptomatic treatment was given for recurrent chest infections, as the disease cannot be cured permanently. Since our patient is having haemoptysis, doctor advised to pay attention to sinopulmonary infections and to continue long term antibiotics to avoid further complications. In conclusion earlier goal directed treatment can prevent complications.

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