Kartagener Syndrome In A 10-Year-Old Girl

Oduro-Boatey C[1]*, Asiamah B[2], Jackson E[3]*
[1]Department of Child Health, University of Ghana Medical Centre, Accra, Ghana
[2]Department of Child Health, Korle-Bu Teaching Hospital, Accra, Ghana
[3]Department of Radiology, Korle-Bu Teaching Hospital, Accra, Ghana

***Corresponding Author: Collins Oduro-Boatey**
Address: Department of Child Health, University of Ghana Medical Centre, Accra, Ghana
Phone: 0208179270 Email: coduroboatey@yahoo.com

Background

Kartagener Syndrome is a subset of Primary Ciliary Dyskinesia (PCD), an autosomal recessive inherited disorder. The clinical triad of sinusitis, bronchiectasis, and situs inversus is diagnostic of the syndrome. Genetic testing is crucial for confirming the diagnosis of primary ciliary dyskinesia.

Keywords: Kartagener syndrome, bronchiectasis, primary ciliary dyskinesia

Case Report

A 4-year-old girl was first seen in our clinic on account of failure to thrive and recurrent cough since birth. She was born preterm by caesarian section on account of antepartum haemorrhage. She was diagnosed with dextrocardia soon after birth.

On examination, she was not cyanosed and not clubbed. Her anthropometric indices were all < 3rd centile. The cardiovascular system was standard, except for the presence of dextrocardia. The abdomen was whole but soft and non-tender, with no masses palpable. There were coarse crepitations in both lung fields. A diagnosis of Kartagener syndrome was made based on the dextrocardia, recurrent cough and failure to thrive.

Echocardiography confirmed dextrocardia as the only abnormality. A Computed Tomography (CT) angiogram of the chest and abdomen showed bronchiectasis of the left lung apex, situs inversus of the abdominal viscera, and dextrocardia, all consistent with the diagnosis of Kartagener syndrome.[1] She began chest physiotherapy to help clear secretions from the airways. She has since been seen on numerous occasions in the outpatient department with recurrent chest infections, sinusitis and otitis media, all managed with oral antibiotics and sometimes expectorants.

Currently, she is in school, but looks small for her age. She is on her weekly chest physiotherapy.

Discussion

Kartagener syndrome is a subset of primary ciliary dyskinesia, an autosomal recessive inherited disease also known as immotile cilia syndrome.[1,2] Immotile syndrome, also called primary ciliary dyskinesia, is due to ciliary abnormality due to inherited primary structural defects in the cilia. This leads to repeated and chronic lung infections.[2,3]

Cilia are located on the epithelial surfaces of the respiratory epithelium and in the fallopian tubes and semen. Cilia are more prevalent in the larger airways. The larger airways also have a higher frequency of ciliary beating, known as the beat frequency, which ranges from 1 to 20 Hz. Cilia are finger-like structures extending from the surface of cells into the lumen of airways. Each cell has about 200 cilia on its surface. Cilia beat and move by the sliding motion of microtubules.[4] This leads to bending and rotating movements which transport mucus towards the mouth. Structural abnormalities of the cilia lead to abnormal beat frequency or immotility, which in turn result in defective mucociliary clearance of airway secretions.

About 50% of patients with ciliary dyskinesia have Kartagener syndrome. The course is variable. In one study, 100% of the children had a productive cough, a hallmark symptom. Lower respiratory tract disease can progress to weight loss and bronchiectasis[3]. Kartagener syndrome is often undiagnosed in children. The presence of dextrocardia in a child with chronic respiratory tract symptoms is diagnostic of Kartagener syndrome.[4] Treatment for Kartagener syndrome is symptomatic. Coughing should be encouraged. Chest physiotherapy helps clear mucus from the airways. Antibiotics are prescribed for evidence of infection of the sinuses or lower airways. Oral antibiotics are usually effective, as in our case. Bronchodilators are helpful for symptomatic wheezing.

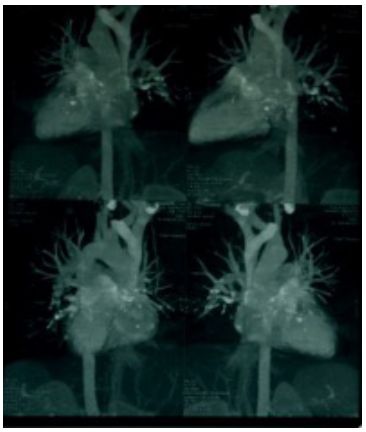

Figure 1 Radiological Scan of condition

References

1. Takeuchi K, Kitano M, Ishinaga H, Kobayashi M, Ogawa S, Nakatani K, Masuda S, Nagao M, Fujisawa T. Recent advances in primary ciliary dyskinesia. Auris Nasus Larynx. 2016 Jun 1;43(3):229-36.6/j.anl. 2015.09.012
2. Rutland J, De Iongh RU. Random ciliary orientation: a cause of respiratory tract disease. New England Journal of Medicine. 1990 Dec 13;323(24):1681-4.
3. Narayan D, Krishnan SN, Upender M, Ravikumar TS, Mahoney MJ, Dolan TF, Teebi AS, Haddad GG. Unusual inheritance of primary ciliary dyskinesia (Kartagener's syndrome). Journal of medical genetics. 1994 Jun 1;31(6):493-6..
4. Hosie P, Fitzgerald DA, Jaffe A, Birman CS, Morgan L. P rimary ciliary dyskinesia: Overlooked and undertreated in children. Journal of paediatrics and child health. 2014 Dec;50(12):952-8.