PD003

Paediatric Cardiology & Congenital Heart Disease • Poster Presentation

KARTAGENER SYNDROME IN A 6-YEAR-OLD GIRL: A CASE REPORT

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Introduction Kartagener syndrome (KS) is a rare autosomal recessive disorder and a subset of primary ciliary dyskinesia (PCD), which manifests clinically as a triad of situs inversus, chronic sinusitis, and chronic bronchiectasis. Early diagnosis and appropriate management are essential to improve outcomes.

Case Report A 6-year-old girl presented with recurrent cough and fever since infancy, progressive weight loss of a month's duration, difficulty breathing of two weeks' duration, and abdominal and leg swelling of a week's duration. On examination, she was wasted and chronically ill-looking, febrile, tachycardic, tachypnoeic, dyspnoeic, had grade 3 finger clubbing and bilateral pitting pedal oedema up to the lower one-third of the legs. She had pectus carinatum and coarse crepitations heard across most lung zones. She had a distended abdomen with visible abdominal wall veins, ascites and tender hepatomegaly. She was in class IV heart failure according to the New York Heart Association (NYHA) classification. Chest radiograph revealed dextrocardia with pneumonic lung changes. Echocardiography revealed a structurally normal heart with dextrocardia and situs inversus, and pulmonary hypertension. Chest computed tomography revealed dextrocardia, situs inversus, and widespread bronchiectatic lung changes. She was managed with supplemental oxygen, antibiotics, diuretics, an angiotensin-converting enzyme inhibitor, and oral sildenafil. However, she developed a sudden onset of stridor, wheezing and worsening respiratory distress 12 days into admission and died on the 13th day of admission.

Conclusion This case highlights the importance of newborn screening for prompt diagnosis and treatment of Kartagener syndrome to reduce morbidity and prevent mortality.

Keywords: Kartagener Syndrome, Primary ciliary dyskinesia, dextrocardia, bronchiectasis