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Case Report

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An Unusual Diagnosis of Kartagener Syndrome in an Adult

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Abstract

Kartagener's syndrome (KS) is a rare hereditary disease. It is a triad of chronic sinusitis, bronchiectasis and situs inversus. It poses a diagnostic difficulty owing to nonspecific clinical pictures. Sometimes, the diagnosis is suspected prenatally when situs inversus is documented in obstetric ultrasound; however, most cases are diagnosed later in life due to repeated respiratory infections or when the clinician listens to heart sounds in the right hemithorax and chest x-ray, complemented with abdominal and paranasal sinuses imaging, shows dextrocardia. An early and accurate diagnosis of this illness is crucial to avoid complications and improve the quality of life of patients.

Here, we highlight the case of a 29-year-old male with KS owing to the late diagnosis and rarity of clinical presentation.

Introduction

Primary ciliary dysfunction (PCD), first described in 1976, is a disorder of the structure and function of motile cilia that results in chronic oto-sinopulmonary disease [1].

Kartagener syndrome, which occurs in about 50% of PCD patients, is a triad of chronic sinusitis, bronchiectasis, and situs inversus resulting from embryonic (nodal) ciliary dyskinesia [2, 3]. Diagnosis can be made by tests to prove impaired cilia function, biopsy, and genetic studies. Treatment is supportive. In severe cases, the prognosis can be fatal if bilateral lung transplantation is delayed.

We report the case of a 29 -year- old man with chronic recurrent upper respiratory infections. He was diagnosed with Kartagener syndrome based on clinical presentation and radiological features.

Case Report

A 29-year-old male patient presented with the chief complaints of cough with copious purulent sputum and shortness of breath for 20 days. His previous records showed repeated clinic visits since the age of 2 years for recurrent episodes of nasal blockage sneezing, rhinorrhoea and productive cough for which he received treatment several times at various places. He was a casual alcohol consumer. There was no history of smoking or tuberculosis. He was born to non-consanguineous parents and the pregnancy was uneventful. There was no similar illness in his family. Physical examination was notable for an enlarged single cervical lymph node on right side which was non tender and firm in consistency. He had grade 2 clubbing of all digits. He was nourished, conscious, and oriented. His blood pressure was 100/70 mmHg, pulse rate 110 beats per minute, respiratory rate 20 breaths per minute, and temperature 39 °C. His arterial oxygen saturation was 93 % with room air.

A respiratory system examination revealed coarse crackles and scattered rhonchi on both basal lung fields. On cardiovascular examination, apex beat was felt on right fifth intercostal space along midclavicular line. Heart sounds were best audible on the right side of his chest. An abdominal examination revealed tympanitic note on percussion and no sign of fluid collection. A nervous system examination showed no abnormality.

A laboratory examination revealed haemoglobin 15 gm/dl, total leukocyte counts 14,500/µl granulocyte 81%, lymphocyte 17%), and platelet count 350,000/L. Sputum analysis revealed gram positive cocci consistent with staphylococcal pneumonia. Serum chemistries were normal. A chest X-ray revealed cardiac apex and aortic arch on right side (Figure 1)

Electrocardiogram also showed inverted P and T waves in lead I, negative deflection of QRS complex and poor progression of R wave in left side chest leads suggestive of dextrocardia. A chest computed tomography scan showed bronchiectatic changes prominent on both lower lung fields (Figure 2). CT scan of abdomen showed liver and inferior vena cava on left side, and spleen on right side, suggestive of situs inversus (Figure 3). Semen analysis revealed immotile and poorly motile sperms. Thence, a diagnosis of KS was made on the basis of clinical presentation and imaging features. He was started on supplemental oxygen with simple face mask, intravenous antibiotics, bronchodilators, mucolytic, and chest physiotherapy. He was symptomatically better with the above therapy and started on long-term low-dose prophylactic antibiotic. He was then referred for vaccination against Pneumococcus and influenza and to the medical chest clinic of our hospital for chest physiotherapy; advised micromanipulation techniques/ invitro fertilization for infertility. He is maintaining regular follow-up and now relatively symptom free.

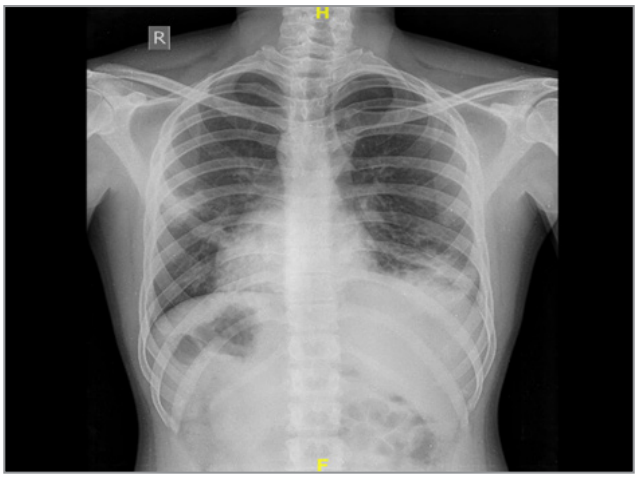

Figure 1: Chest x-ray of the patient showing dextrocardia.

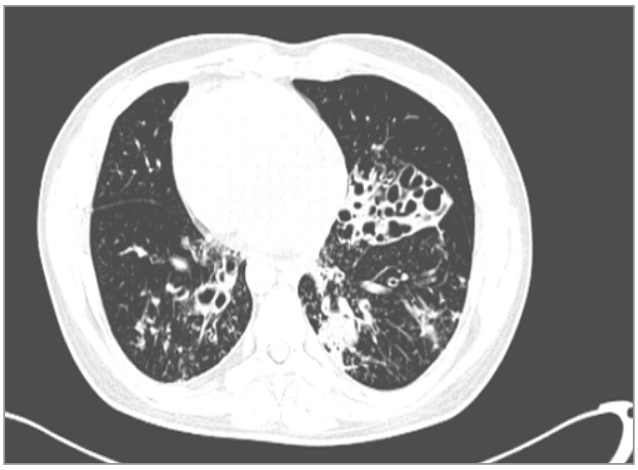

Figure 2: CT scan of the thorax showing bronchiectasis in bilateral lower lung fields.

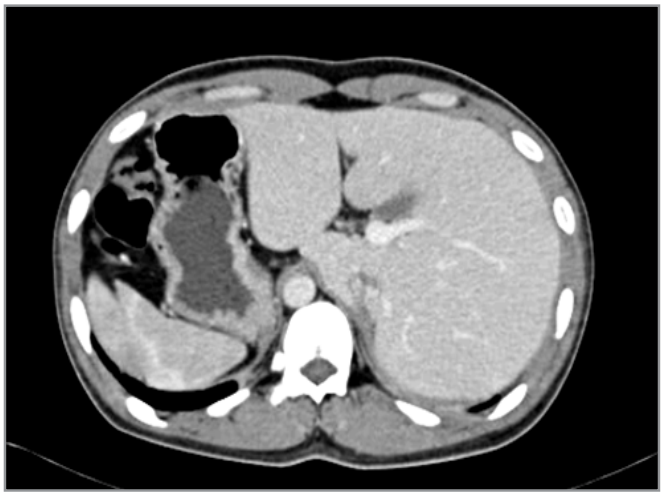

Figure 3: CT scan of the abdomen showing situs inversus

Discussion

KS is part of the PKD spectrum related to an autosomal recessive genetic disorder that affects ciliary motility and predisposes to problems of laterality, rhino pulmonary infections and impaired fertility [4].

PKD diagnosis is confirmed by the presence of one or more of the criteria proposed by the European Society of Pneumology: low levels of nasal nitric oxide, frequency of ciliary oscillation ≤-11Hz by high-speed video microscopy from ciliary biopsy or more than 20-30% of ciliary ultra-structural abnormalities by electron microscopy [5]. Imaging plays a key role in proving the anatomical findings that support the diagnosis of KS. Finally, prognosis depends on lung involvement. Although there is no specific treatment for this clinical entity, failure to diagnose this may subject the patient to unnecessary repeated admissions, investigations and inappropriate treatment.

Our case highlights the significance of early diagnosis and management of the condition to prevent irreversible lung damage and prevent chronic lifelong sequelae.

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