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Case Study

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KARTAGENER'S SYNDROME:- A CASE REPORT

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ABSTRACT

Background: Kartagener's Syndrome Is Subset Of Primary Ciliary Dyskinesia, An Autosomal Recessive Inherited Disorder Characterized By The Clinical Triad Of Chronic Sinusitis, Bronchiectasis And Situs Inversus. Abnormal Ciliary Structure Or Function Leading To Impaired Ciliary Motility Is The Main Pathophysiologic Problem In Kartegener's Syndrome. **Case Presentation:** A 80 Year Old Man From Meerut City, Up; Presented To Lokpriya Hospital With Recurrent High Grade Fever, Shortness Of Breath, Productive Cough, Multiple Loose Stools, Abdominal Pain More Than Decade. Clinical And Imaging Findings Revealed Chronic Sinusitis, Bronchiectasis, Dextrocardia And Situs Inversus. He Was Treated With Orally Administered Antibiotics, Mucolytic And Chest Physiotherapy. He Was Symptomatically Better With The Above Therapy And Started On A Long Term Low Dose Prophylactic Antibiotics. **Conclusions:** Patients With Kartegener's Syndrome Exist In India As Cases Of Chronic Recurrent Sinopulmonary Infections. As There Is No Easy, Reliable Non-Invasive Diagnostic Test For Kartegener's Syndrome And The Correct Diagnosis Is Often Delayed By Years, It May Cause Chronic Respiratory Problems With Reduced Quality Of Life. Genetic Counseling And Fertility Issues Should Be Addressed Once Kartegener's Syndrome Is Diagnosed.

KEYWORDS: Kartagener's Syndrome, Primary Ciliary Dyskinesia, Chronic Sinusitis, Bronchiectasis, Situs Inversus.

CASE PRESENTATION

80 Year Old Male, Bihari, Non-Smoker, Non-Alcoholic Presented With High Grade Fever, Loose Stools, Abdominal Pain, Shortness Of Breath And Productive Cough Since 10 Days. Patient Works In A Factory Since 1977.

Shortness Of Breath And Productive Cough Since 15 Years With Episodic Fever And Worsening Of Symptoms. History Of Old Treated Pulmonary Koch Since 1977. History Of Hypertension Since 20 Years, History Of Copd Since 13 Years. He Had Been Previously Treated With Antibiotics, Antihistaminics, Bronchodilators, Inhaled /Oral Steroids, Took Att On 2011 For 12 Months, But The Response Was Only Partial And Temporary. He Also Had Similar Complains On And Off, Since 1977. On Examination, He Was Febrile With Nasal Discharge, Wheezy Chest And B/L Coarse Crackles. His Heart Sounds Were Heard Best On The Right Side Of The Chest. There Was No Digital Clubbing. Chest X–Ray Shows Copd Changes With Dextrocardia. Hrct Chest Shows Copd Changes With Multiple Fibrotic Bands With Bronchiectatic Changes Seen In B/L Upper Lobes, Right Middle Lobe And B/L Lower Lobes. Ultrasound Of The Abdomen Showed Spleen On The Right Side Of Abdomen, While Liver On The Left Was Suggestive Of Complete Situs Inversus.2d Echo Shows Severe Tr, Moderate Pah And Abnormal Motion Of The Septum.

DISCUSSION

Ks Is A Rare, Autosomal Recessive Ciliopathic Disorder Characterized By The Clinical Triad Of Chronic Sinusitis, Bronchiectasis And Situs Inversus. Its Estimated Incidence Is Approximately 1 In 30,000 Live Births. Normal Ciliary Function Is Critical For Respiratory Tract Host Defence, Sperm Motility, And Normal Visceral Orientation During Embryogenesis. Lack Or Dysfunction Of Dynein Arms, Radial Spokes, And Microtubules Of Cilia Are Recognized Structural And Functional Abnormalities Of Ciliary Ultrastructures, Encoded By The Mutated Genes Dnai1 And Dnah5. These Faulty Genes Cause The Cilia To Be The Wrong Size Or Shape Or Move In The Wrong Way, Making Ciliary Motility Defective.[5,6] Abnormal Ciliary Motility At Sites Leads To Chronic Recurrent Sinopulmonary Infections And Infertility. Impaired Ciliary Motility During Embryogenesis Predisposes To Left– Right Laterality Defects Like Situs Solitus (That Is, Dextrocardia Only) Or Situs Inversus Totalis Where Transpositions Of Thoracic And Abdominal Organs Are Noticed.[5–7] The Diagnostic Criteria Recommended For

This Syndrome Include History Of Chronic Bronchial Infection And Rhinitis From Early Childhood, Combined With One Or More Of Following Features: (A) Situs Inversus Or Dextrocardia In A Patient Or A Sibling, (B) Alive But Immotile Spermatozoa, (C) Absent Or Impaired Tracheobronchial Clearance, And (D) Cilia Showing Characteristic Ultrastructural Defect On Electron Microscopy.[7, 8]

X-Ray Chest- Dextrocardia With Copd.
Hrct Chest- Cystic Bronchiectasis With Copd.

Laboratory Screening Tests Include Exhaled Nasal Nitric Oxide Level Determination And Saccharin Test For Assessing Nasal Epithelial Mucociliary Function. High-Speed Video Microscopy For Assessing Ciliary Beat Frequency And Pattern, Transmission Electron Microscopic For Detecting Ultrastructural Ciliary Defect, And Genetic Testing For Dnai1 And Dnah5 Mutations Are Confirmatory Laboratory Tests. Abnormal Laboratory Findings In Ks Include Reduced Nasal Nitric Oxide Level (~10% Of Normal), Prolonged Saccharin Clearance Time (>1 Hour), Reduced Ciliary Beat Frequency (Absent Ciliary Ultrastructure (Dynein Arms), And Mutated Dnai1 And Dnah5 Genes.[6,7] Our Patient Presented With Recurrent Episodes Of Sinopulmonary Infections. Imaging Findings Revealed Bronchiectasis, Dextrocardia, And Situs Inversus, Which Met The Diagnostic Criteria For Ks. Laboratory Screening And Confirmatory Tests, Which Required A Better Clinical Setup, Were Not Done. As There Is No Easy, Reliable Non-Invasive Diagnostic Test For Ks And The Correct Diagnosis Is Often Delayed By Years, It May Cause Chronic Respiratory Problems With Reduced Quality Of Life.[7,9,10] Standard Treatment For Sinopulmonary Problems In People With Ks Includes Chest Physiotherapy, Mucolytics, And Antibiotics. A Long-Term Low-Dose Prophylactic Antibiotic Is Required In Those With Frequent Exacerbation Of Bronchiectasis (≥3 Times/Year). Influenza And Pneumococcal Vaccination Should Be Routinely Given.[7,9,10]

CONCLUSIONS

Patients With Ks Exist In Ethiopia As Cases Of Chronic Recurrent Sinopulmonary Infections. As There Is No Easy, Reliable Non-Invasive Diagnostic Test For Ks And The Correct Diagnosis Is Often Delayed By Years, It May Cause Chronic Respiratory Problems With Reduced Quality Of Life. Genetic Counseling And Fertility Issues Should Be Addressed Once Ks Is Diagnosed.

Abbreviations

Afb: Acid-Fast Bacilli; Bp: Blood Pressure; Ct: Computed Tomography; Ent: Ear, Nose, And Throat; Ks: Kartagener's Syndrome; Pr: Pulse Rate; Rr: Respiratory Rate; Sao2: Arterial Oxygen Saturation; T°: Temperature.

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Availability of data and materials

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Ethics approval and consent to participate

The authors declare that ethics approval was not required for this case report as we did not use any new procedures or any treatment, which was not approved for clinical use in our institution.

Consent for publication

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Competing interests

The authors declare that they have no competing interests.

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