Original Research Paper

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Respiratory Medicine

A RARE CASE OF KARTAGENER'S SYNDROME PRESENTING AT TERTIARY CARE CENTER.

Dr. Jignesh Gengadiya	Resident Doctor, Department of Respiratory Medicine, Government Medical College, Surat.
Dr. Parul Vadgama*	(M.B.B.S., M.D. Respiratory Medicine) Associate Professor, Department of Respiratory Medicine, Government Medical College, Surat. *Corresponding Author
Dr. Khyati Shamaliya	(M.B.B.S., M.D. Respiratory Medicine) Assistant Professor, Department of Respiratory Medicine, Government Medical College, Surat.
Dr. Sreerag Varior	(M.B.B.S., M.D. Respiratory Medicine) Resident Doctor, Department of Respiratory Medicine, Government Medical College, Surat.
Dr. Keyur Patel	(M.B.B.S., M.D. Emergency Medicine) Resident doctor, Department Of Emergency Medicine, Government Medical College, Surat.

ABSTRACT Kartagener's syndrome is inherited autosomal recessive disorder characterized by impaired ciliary dysfunction. It is clinical triad of chronic sinusitis, bronchiectasis and situs inversus. A 24 year male patient present with past history of repeated nasal blockage, thick nasal secretion and headache since childhood, presenting to us with complain of cough with expectoration and fever since 4 days. 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. Management of bronchiectasis and sinusitis should be optimum and recurrence of infection should be prevented. Early diagnosis and management of kartagener's are critical to prevent irreversible damage and chronic lifelong sequale.

KEYWORDS :

INTRODUCTION:

Kartagener's syndrome named after German physician, Manes Kartagener's in 1933.Kartageners syndrome is rare autosomal recessive disorder which was first describe by Siewert in 1904.Kartagener syndrome is a subset of primary ciliary dyskinesia. It is clinical triad of chronic sinusitis, bronchiectasis and situs inversus. Also called as 'Immotile Cilia Syndrome'. Normal ciliary function is critical for respiratory host defense and motility of sperms, and ensure proper visceral orientation during embryogenesis. Any defect in DYNEIN gene will cause impaired movement of cilia, DNAI1 and DNAH5 gene mutation leads to impaired ciliary motility which predisposes to recurrent respiratory and sinus infection.

Case Report:

A 24 year male patient present with past history of repeated nasal blockage, thick nasal secretion and headache since childhood, presenting to us with complain of cough with expectoration and fever since 4 days. On examination apex beat was palpated on right side and liver was palpable on left side. On Auscultation he had bilateral infrascapular crackles. Routine blood investigation was normal. Sputum AFB and Gene expert report was negative for mycobacterium tuberculosis. Semen analysis was suggesting of azoospermia.2D ECHO report suggest mild RA and RV dilated. Chest X-ray was suggesting of dextrocardia with bilateral infiltration in lower lungs. USG Abdomen findings shows liver on left side and spleen on right side which indicate feature of situs inversus. HRCT was suggestive of bronchiectasis with secondary infection of bilateral lower lungs. CT PNS suggestive of Pansinusitis. Based on clinical presentation and radiological findings, kartagener's syndrome was diagnosed. Initially Patient was treated with empirical antibiotics therapy and then with antibiotics according to culture report, chest physiotherapy with cough expectoration was given. Patient was advice for get pneumococcal vaccine and influenza vaccine on follow-up.

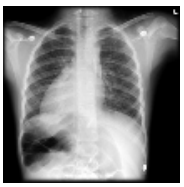

Figure 1: Chest X-ray shows dextrocardia

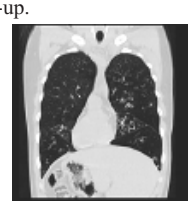

figure 2: HRCT showing bronchiectasis

DISCUSSION:

Kartagener's syndrome is a rare, autosomal recessive disorder characterized by the clinical triad of chronic sinusitis, bronchiectasis, and situs inversus. Its estimated incidence is approximately 1 in 30,000 live births. 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 ultra-structural defect on electron microscopy. 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. Abnormal laboratory findings in KS include reduced nasal nitric oxide level (~10% of normal), prolonged saccharin clearance time (>1 hour), reduced ciliary beat frequency (<11 Hz/second), absent ciliary ultrastructure (dynein arms), and mutated *DNAI1* and *DNAH5* genes. Standard treatment for sinopulmonary problems in people with KS includes chest physiotherapy, mucolytic, 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.

CONCLUSION:

A young male patient presenting with chronic sinusitis and bronchiectasis should be fully investigate for kartagener's syndrome as it is a rare condition. Other Systems involvement should be evaluated. Management of bronchiectasis and sinusitis should be optimum and recurrence of infection should be prevented. Early diagnosis and management of kartagener's are useful to prevent irreversible damage and chronic lifelong sequale.

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