

CASE REPORT

KARTAGENER'S SYNDROME: A RARE CAUSE FOR RECURRENT CHRONIC SINUSITIS

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ABSTRACT: Kartagener's syndrome is a rare condition with systemic manifestation. Chronic rhinosinusitis is a common condition seen in ENT OPD routinely. Of the several causes for sinusitis, Kartagener's syndrome is a very rare, unusual cause, which can remain undetected for a long time. This case highlights the ENT and systemic manifestation of Kartagener's syndrome and its rarity in ENT practice. The multi disciplinary approach in its management is also discussed.

KEYWORDS: Kartagener's syndrome, Bronchiectasis, Situs inversus, Dextrocardia, Sinusitis.

CASE REPORT: A 20 year old male patient presented to the ENT OPD with complaints of nasal obstruction and nasal discharge, on and off since childhood. Headache since 2 months. Recurrent episodes of lower respiratory tract infection, often requiring hospitalization. His nasal obstruction was insidious in onset, gradually progressive, aggravated during winter months and subsided on taking medications. Nasal discharge was copious, thick and persistent, no h/o bleeding from the nose and patient's sensation of smell was reduced. Headache was bilateral, dull aching, associated with postnasal drip. No other significant past history. Patient has 2 younger male siblings, who are asymptomatic.

On examining the nose, there was septal deviation to the right with bilateral hypertrophy of inferior turbinate. Tenderness was noted over the frontal and maxillary sinuses on both the sides. Ear, throat and neck examination was normal. Nasal endoscopy confirmed anterior rhinoscopy findings. Thick mucoid discharge was noted in the middle meatus in both nasal cavities. Systemic examination revealed grade III clubbing (Fig. 1) and coarse crepitation. Further examination by general physician revealed dextro-cardia, situs inversus totalis with spleen on the right side and liver on the left side.

X-ray of nose and PNS, showed opacification of bilateral maxillary, frontal and ethmoid sinuses. Nasal septum was deviated to the right (Fig. 2). The X-ray findings were confirmed by CT scan of nose and PNS. However sphenoid sinus was normal, Osteomeatal complexes were normal bilaterally and no bony erosions or irregularities were seen. Chest X- ray highlighted trachea to be normal in position, dextrocardia and bronchiectatic changes in both main bronchi. Cardio-phrenic angles were free of fluid (Fig. 3). Ultrasound abdomen revealed Situs inversus totalis with no significant sonological abnormality in any individual organs. HRCT Thorax showed Dextrocardia with abdominal situs inversus involving left sided liver and right sided stomach and spleen. Descending aorta was on the right side and IVC on the left side. Bilateral bronchiectasis with complete collapse of left lower lobe and honey comb pattern. Right upper and lower lobe showed bronchiectasis with left middle lobe involvement. Scarring was noted in the right hilum

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(Fig. 4). Routine investigations were normal and insignificant. Nasal ciliary motility test was abnormal with significantly prolonged saccharine clearance time.

A clinical diagnosis of Kartagener's syndrome was established. A differential diagnosis of the following was kept in mind: Primary ciliary dyskinesia, Young's syndrome, Cystic fibrosis and Mucoviscidosis.

Patient underwent functional endoscopic sinus surgery and correction of septal deviation under general anesthesia. Thick sticky mucopurulent secretion was seen in the middle meatus, middle meatal antrostomy was done and thickened and unhealthy mucosal lining was noted in the maxillary sinus. Ethmoidal clearance was achieved. The frontal recess area was widened and thick secretions were cleared out. Post-operative period was uneventful. Bronchiectasis was treated with broad spectrum antibiotics and chest physiotherapy. Patient remained symptom free on follow up after 2 years.

DISCUSSION: The triad of bronchiectasis, sinusitis, and situs inversus was first described by Siewert in 1903, although its usual eponym is Kartagener's syndrome – after the Swiss paediatrician who described four cases with similar features in 1933 (Ref 1).

Primary ciliary dyskinesia is a rare, ciliopathic, autosomal recessive genetic disorder that cause defect in the cilia lining the upper and lower respiratory tract, eustachian tube, middle ear and fallopian tube (Ref 2). Incidence is 1 in 30,000. 50% of the individuals fall into the subgroup of Kartagener's syndrome characterized by Situs inversus, bronchiectasis, sinusitis and recurrent infection resulting in nasal polyposis in 40% of the patients (Ref 6). Young's syndrome, in addition to features of Kartagener's syndrome, has azoospermia. Cystic fibrosis is a disorder affecting fluid secretion in exocrine glands and epithelial lining of GIT, respiratory system and reproductive system. It is characterized by chronic respiratory tract infection which leads to bronchiectasis, affecting 1 in 3200 caucasians and 1 in 3100 Asians (Ref 5). Patients with Kartagener's syndrome have lack of ciliary activity which facilitates bacterial activity and predisposes sinus and bronchus to infection. Absence or shortening of the dynein arm, that is responsible for the coordinate bending of the cilia is seen in 50% of cases (Ref 3). Impaired cell motility seen during embryogenesis results in situs inversus.

Nasal mucociliary clearance can be measured by the 'nasal saccharin transit time' test in which a saccharin pellet is placed on the anterior end of the inferior turbinate and the time taken for the subject to notice the taste is recorded. This test requires the patient's co-operation and is not reliable in children under 10 years of age. Nasal cilia are easily accessible, and can be obtained from the inferior turbinate without anesthesia by a non-invasive brush technique. Ciliary beat frequency can then be assessed by light microscopy and photometric techniques, and cilia fixed on electron microscopy (Ref 4).

Syndromes with ciliary dysfunction affects mucociliary clearance of nasal and paranasal sinus secretions. This in-turn leads to stagnation of secretions and obstruction of paranasal sinus opening. Retained secretions facilitate secondary bacterial infection, which can turn chronic and leads to recurrent sinus infection. Treatment can be medical management of infection and liquefaction of nasal secretions to facilitate its clearance. Surgically, the narrow osteomeatal

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complex can be widened to prevent stagnation of secretions and better ventilation of the sinuses. However these patients require lifetime follow up at regular interval.

CONCLUSION: One should always bear in mind the diagnosis of Kartagener's syndrome or ciliary dyskinesia when there is thick mucoid discharge in the nasal cavity with under lying recurrent LRTI. On the other hand, whenever there is a case of situs inversus look for sinusitis and bronchiectasis which can yield a diagnosis of Kartagener's syndrome. Also one needs to have a proper methodology of investigations in such cases to arrive at the diagnosis of Kartagener's syndrome. Our presentation highlights the rarity of this condition and it's wide ranging ENT and multidisciplinary systemic findings.

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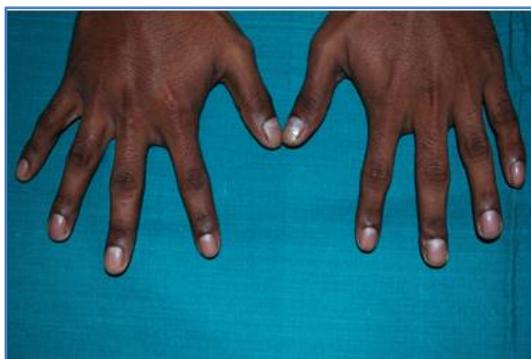


Fig. 1: Clubbing of fingers



Fig. 2: X-ray nose and PNS showing sinusitis with deviated nasal septum

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Fig. 3: X-ray chest showing dextrocardia

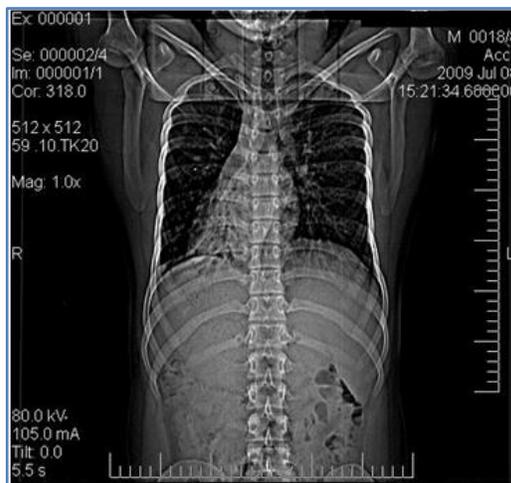


Fig. 4: CT thorax demonstrating bilateral bronchiectasis, dextrocardia and situs inversus

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