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Original Research Article

OUR APPROACH TOWARD A RARE CASE OF KARTAGENER'S SYNDROME

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ABSTRACT:
Kartagener's Syndrome is a hereditary autosomal recessive disease of deficiencies in the activity of ciliary movement and consists of the triad of bronchiectasis, situs inversus, and sinusitis. Medical Management includes mucolytic or mucoactive agents and antibiotics with good pseudomonal coverage. Surgical intervention includes Functional Endoscopic Sinus Surgery for chronic sinusitis, Grommet insertion for serous otitis media, lobectomy or even lung transplantation. We present the case of a 17 year old boy who presented with complaints of recurrent cold, headache, bilateral nasal obstruction and discharge for 5 years. Heart sounds were present on the right side of chest and liver dullness was observed on the left side on percussion. Diagnostic Nasal Endoscopy showed polypoidal masses in bilateral nasal cavities. He was diagnosed with Kartagener's Syndrome based on clinical and radiological findings. Functional Endoscopic Sinus Surgery was done and the polypoidal masses were removed from the nasal cavities and bilateral maxillary sinuses and wide maxillary antrostomy done on both sides. The patient was followed up until Postoperative Day7. The patient was then lost to follow up.

1. INTRODUCTION:

Kartagener's Syndrome is a hereditary autosomal recessive disease of deficiencies in the activity of ciliary movement and consists of the triad of bronchiectasis, situs inversus, and sinusitis.(1) In KS, the gene mutation at *DNAI* and *DNAH5* leads to impaired ciliary motility, which predisposes to recurrent sinopulmonary infections, infertility, and errors with left–right body orientation.(2) The prevalence of PCD varies from 1 in 20 000 to 1 in 60 000 live births, of which, 50% may develop situsinversus. In men, PCD may lead to infertility due to impaired spermatozoa motility secondary to defective sperm flagella, although it is not a definite or consistent finding.(3) Medical Management includes mucolytic or mucoactive agents such as hypertonic saline nebulization and acetylcysteine, antibiotics with good pseudomonal coverage, daily chest physiotherapy and supportive pulmonary care. Mucoactive agents help in making it easier to clear secretions. Surgical intervention in such

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cases includes Functional Endoscopic Sinus Surgery for chronic sinusitis, Grommet insertion for serous otitis media and in cases of severe pulmonary involvement, lobectomy or even lung transplantation may be required. An early and accurate diagnosis of this illness is crucial to avoid complications and improve the quality of life of patients.(4)

CASE REPORT:

A 17 year old boy presented to the ENT OPD of our institute with complaints of recurrent cold, headache, bilateral nasal obstruction and discharge since 5 years. He had 2 episodes of fever a week back. On general examination, his vitals were stable and there was cervical lymphadenopathy on bilateral submandibular region. Anterior rhinoscopy showed bilateral thick nasal discharge and otoscopy was suggestive of serous otitis media. On systemic examination, heart sounds were present on right side of chest; with apex beat over right 5^{th} intercostal space.Liver dullness was observed on the left side on percussion. Diagnostic Nasal Endoscopy showed polypoidal masses in bilateral nasal cavities (Figure 1). Computed Tomography of the Paranasal Sinuses was done which showed soft tissue densities completely filling bilateral maxillary sinuses with polypoidal masses in bilateral nasal cavities, mild mucosal thickening in bilateral sphenoid sinusesand gross Deviated Nasal Septum towards left side(Figure 2). An ultrasonogram whole abdomen was done to further evaluate the unusual clinical findings on cardiac and abdominal examination, which showed findings suggestive of situs inversus i.e, Liver on the left side and spleen on the right side. A High Resolution Computed Tomography of the chest and abdomen was done to further evaluate pulmonary and abdominal findings which showed findings of Situs inversus with right sided aortic arch, left sided liver and right sided spleen. There were cylindrical bronchiectatic changes in bilateral lungs with secondary bronchitis (Figure 3). Echocardiography was normal with mild Tricuspid Regurgitation. Blood investigations were unremarkable except for mild leukocytosis and raised Erythrocyte Sedimentation Rate. Previous Fine Needle Aspiration Cytology report of cervical lymphadenopathy was reported to be reactive lymphadenitis.

The patient was posted for Septoplasty and Functional Endoscopic Sinus Surgery and the polypoidal masses were removed from the nasal cavities and bilateral maxillary sinuses (Figure 4), wide maxillary antrostomy done on both sides and septal deviation corrected. Concurrently, intravenous antibiotics, oral steroids, bronchodilators and a mucoactive agent were added to the treatment regimen. Nasal Pack was removed on Postoperative Day 2 and saline with steroid irrigation was started. He was discharged on Postoperative Day 5. The patient was followed up until Postoperative Day 7 in which we removed the crusts from the nasal cavity and sinuses (Figure 5). The patient was then lost to followup since he was from a remote area and did not visit again for routine followups.

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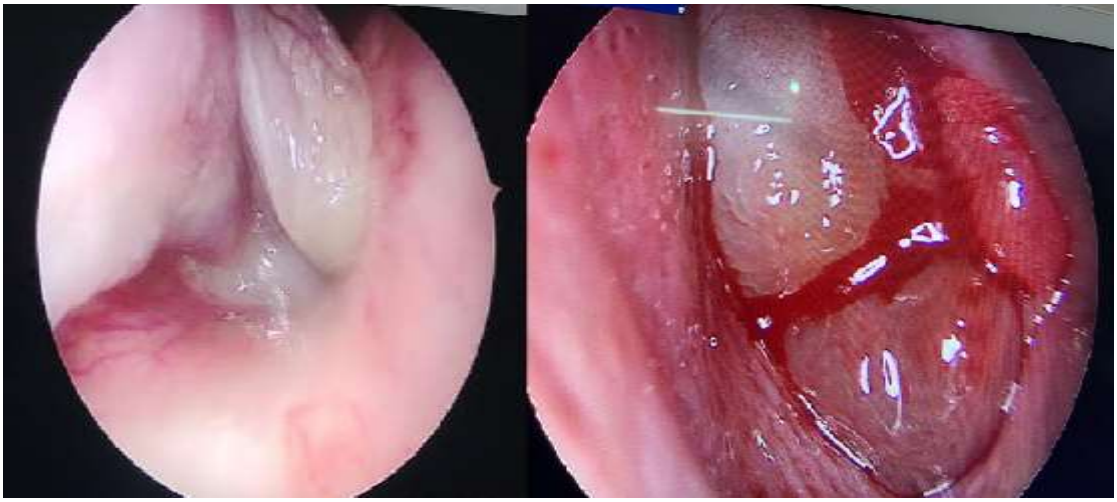

Figure 1: Bilateral nasal polyposis on diagnostic nasal endoscopy

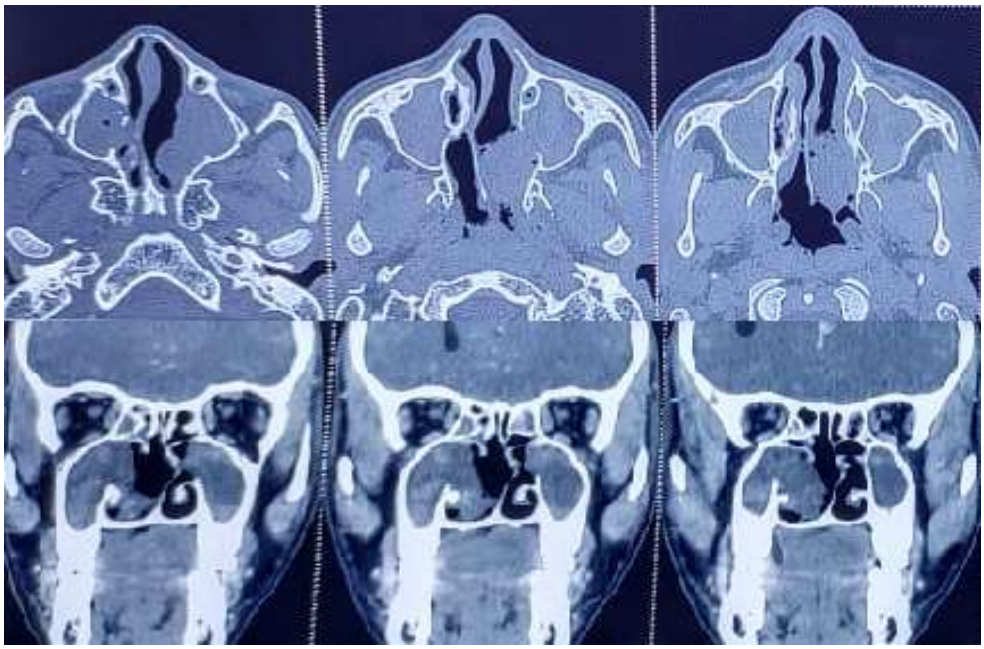

Figure 2: Computed Tomography of Paranasal Sinuses showing bilateral maxillary sinuses filled with polyp extending into the nasal cavities

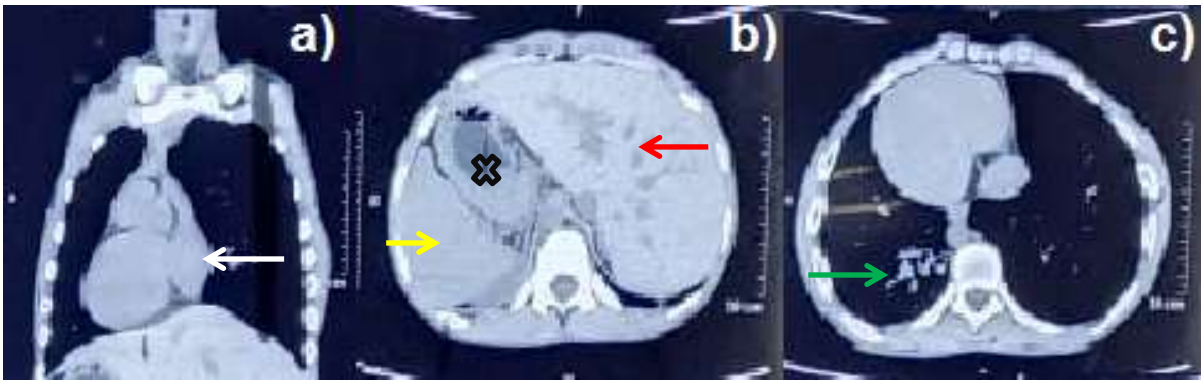

Figure 3: Findings in High Resolution Computed Tomography of the chest

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(a) Coronal section showing heart placed on right side (white arrow)
(b) Axial section showing liver on left side (red arrow), spleen (yellow arrow) and stomach (marked x) on right side
(c) Axial section showing bronchiectatic changes (green arrow)

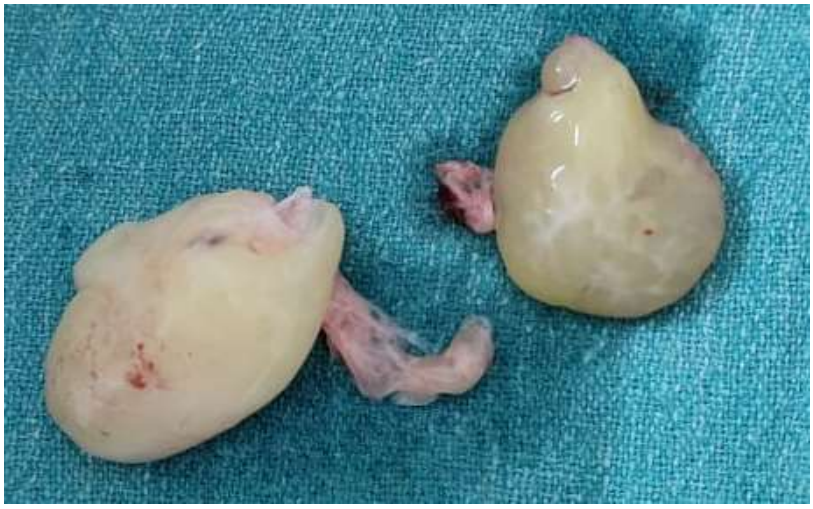

Figure 4: Polypoidal masses removed from bilateral maxillary sinuses and nasal cavities

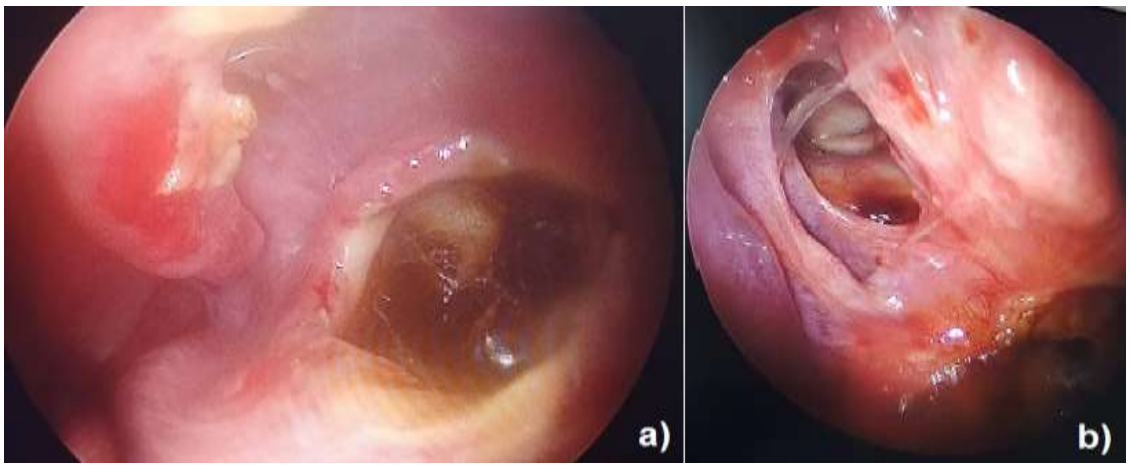

Figure 5: Postoperative Day 7 nasal endoscopy showing - a)wide antrostomy on the left side with slight crusting and b) wide antrostomy on right side

2. DISCUSSION:

Primary Ciliary Dyskinesia (PCD) is a ciliary motility disorder that can be congenital or acquired, caused due to ineffective or absence of beating of motile cilia in the different body systems. Patients with PCD who present with a clinical triad of chronic sinusitis, bronchiectasis, and situs inversus are clinically diagnosed to be suffering from Kartagener's syndrome.(3) The diagnostic criteria recommended for Kartagener's syndrome is a history of chronic bronchial infection and rhinitis from early childhood, with one or more of the following: (a) situs inversus or dextrocardia in the patient or a sibling, (b) living but immotile spermatozoa, (c) tracheobronchial clearance, which is absent or nearly so.(5) Immotile spermatozoa could not be demonstrated in our case since the patient refused the test. The "gold-standard" diagnostic test for PCD has been electron microscopic ultrastructural

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analysis of respiratory cilia obtained by nasal scrape or bronchial brush biopsy, but in most cases, diagnosis is made on the basis of clinical and radiological evaluation.(6) The current therapeutic strategy for KS includes medication, surgery, and some adjuvant therapies. Medication focuses on modifying mucus viscosity and controlling inflammation and infection in the airway, and includes mucoactive agents, steroids, bronchodilators and antibiotics. Adjuvant therapy includes nasal irrigation, pneumococcal and influenza vaccinations. However, the efficacy of both medical and adjuvant therapy seems to be unsatisfactory, most probably because of retained secretions in the airway. Functional Endoscopic Sinus Surgery is done to reduce disease burden and to establish the drainage of sinuses and improve access for regular cleaning.(7)

3. CONCLUSION:

Early diagnosis is essential for the effective management of Kartagener's syndrome (KS) and to prevent associated complications. While cystic fibrosis and KS are commonly identified in early infancy, KS can occasionally remain asymptomatic for extended periods.(8) Timely detection of KS is essential for a better prognosis and symptom control, as respiratory problems in KS patients stem from cilia abnormalities. Therefore, when a patient presents with recurrent respiratory tract infections and shows organ malposition on radiology, first consideration should be given to Kartagener’s syndrome as the underlying cause.

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