International Journal of Otorhinolaryngology and Head and Neck Surgery
Shilpa C et al. Int J Otorhinolaryngol Head Neck Surg. 2025 Jun;11(3):285-287
http://www.ijorl.com
pISSN 2454-5929 | eISSN 2454-5937

DOI: https://dx.doi.org/10.18203/issn.2454-5929.ijohns20251514

Case Report

Unravelling Kartagener's syndrome in the face of recurrent polyposis: a case report

Shilpa C., Sandeep S., Sneha Unnikrishnan*

Department of ENT, JSSAHER, Mysuru, Karnataka, India

Received: 11 December 2024
Revised: 20 March 2025
Accepted: 21 March 2025

***Correspondence:**
Dr. Sneha Unnikrishnan,
E-mail: snehau345@gmail.com

ABSTRACT

Kartagener's syndrome (KS) is a rare autosomal recessive disorder characterized by a triad of situs inversus, chronic sinusitis, and bronchiectasis. It results from primary ciliary dyskinesia (PCD), caused by ultrastructural defects in the dynein arm of microtubules, leading to impaired mucociliary clearance. While chronic sinusitis is a well-recognized feature, nasal polyposis can sometimes be an initial presentation, making early diagnosis challenging. We report the case of a 40-year-old male who presented with persistent nasal polyposis and a history of recurrent sinus infections. Further evaluation, including imaging and clinical assessment, revealed that his nasal polyposis was part of the broader spectrum of KS. The presence of situs inversus and bronchiectasis confirmed the diagnosis. This case underscores the importance of considering KS in patients presenting with unexplained chronic nasal polyposis, particularly when associated with recurrent sinusitis. Early recognition and diagnosis are crucial in optimizing management and preventing long-term complications. A multidisciplinary approach is essential to improve clinical outcomes and quality of life for affected individuals.

Keywords: Kartagener's syndrome, Chronic nasal polyposis, Primary ciliary dyskinesia

INTRODUCTION

Kartagener's syndrome (KS) is a genetic condition with an autosomal recessive inheritance comprising a triad of situs inversus, bronchiectasis and sinusitis.[1,2] It was first described by Siewart in 1904, then it was Kartagener who recognized the etiological correlation between the elements of the triad and reported four cases in 1933.[4] Being the subset of PCD, the estimated prevalence of PCD is about 1 in 30,000.[2] While chronic sinusitis and bronchiectasis are well-recognized features of KS, nasal polyposis as an initial presentation is less common and may delay diagnosis.

CASE REPORT

A 40-year-old male patient_presented to our outpatient department with complaints of bilateral nasal obstruction for 27 years which aggravated since past one year.

There was history of being evaluated outside at different hospitals and he had undergone nasal surgeries thrice for similar complaints in the past. He was put on steroid nasal sprays following surgery which he subsequently stopped using. He mentioned undergoing nasal surgery initially at the age of 13 years then at 15 years followed by revision surgery at the of age 29 years. Symptoms recurred after a short period of time.

Diagnostic nasal endoscopy showed multiple pale greyish polypoidal mass in bilateral middle meatus.

Patient underwent detailed evaluation before being taken up for endoscopic sinus surgery for removal of sinonasal polyps. Routine investigations included blood, chest X-ray, ECG.

CT PNS revealed a post operative status of the nose and paranasal sinuses along with hyperdense and soft tissue

attenuation in all the sinuses suggestive of pansinusitis and sinonasal polyposis.

During systemic examination cardiovascular system revealed heart sounds heard on right 4th intercostal space. On evaluating the patient further, routine blood investigations were found to be normal, but chest X-ray PA view showed dextrocardia with right fundal shadow.

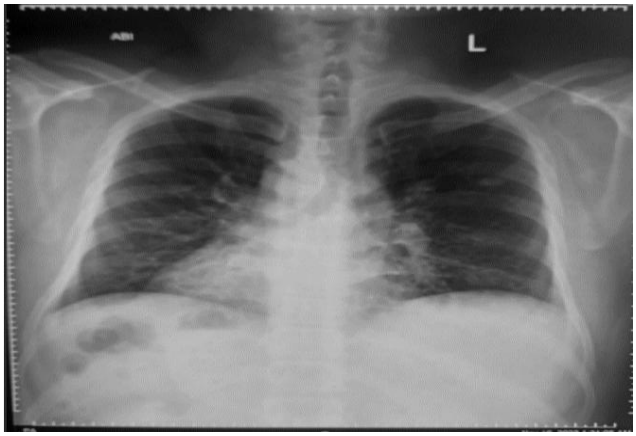

Figure 1: Chest X-ray of dextrocardia.

ECG showed changes pertaining to dextrocardia which included positive QRS complexes in aVR and absent R wave progression. The 2 D echo was obtained to confirm dextrocardia which again reported as situs solitus with mild mitral valve regurgitation and EF of 66%.

USG abdomen showed complete inversion of organs to opposite side.

HRCT thorax was ordered to rule out active pulmonary pathology. Which revealed situs inversus totalis and cylindrical bronchiectasis pattern in right upper lobe, left middle lobe and multiple ground glass density nodules with tree in bud appearance in bilateral lung fields.

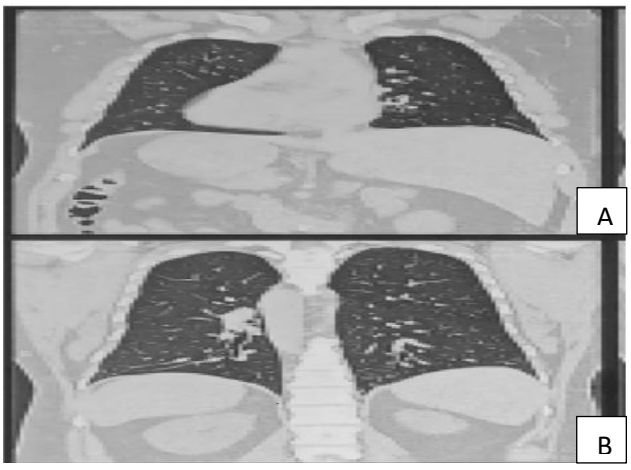

Figure 2 (A and B): HRCT thorax showing situs inversus totalis with bronchiectasis pattern in bilateral lungs.

Pulmonologist opinion was taken in view of HRCT thorax results, diagnosed as active lung infection and started on appropriate antibiotics, nebulisations and mucolytics. Sputum for AFB and culture showed no growth. Patient was discharged and surgery was deferred due to active lung infection. Later he was followed up on weekly basis and after 2 months patient underwent endoscopic sinus surgery for CRS with polyposis for clearance of polyps from nose and sinuses.

Postoperatively the patient received tablet Ambroxol (twice daily) for 15 days to improve mucus clearance, followed by budesonide nasal douching twice daily for 15 days to reduce sinonasal inflammation and prevent polyps. Long term care included hypertonic saline (3%) douching twice daily for 3 weeks.

During post operative follow up patient was started with long term intranasal corticosteroid nasal spray after 2 weeks, and was asked to continue for 6 months with regular follow up.

DISCUSSION

KS is a subset of a larger group of ciliary motility disorders called PCDs. It is a genetic condition with an autosomal recessive inheritance, comprising a triad of situs inversus, bronchiectasis and sinusitis.[1,4]

The main genes identified in the autosomal recessive PCD variants are (DNAI1) and (DNAI2) and heavy chain (DNAH5) and (DNAH11).[1] It was shown as the lack of dynein arm of cilia of central pair of microtubules responsible for the disturbance of mucociliary clearance due to congenital structural defect.[3,5]

A case series by Mishra et al indicated that both males and females with KS can have infertility due to diminished sperm motility and ovum transport, highlighting the need to address infertility in KS.[3,4,6] Infertility was not a feature seen in our patient.

VEGF levels are significantly elevated in nasal polyp mucosa.[6] *In vitro* culture experiments showed the use of Bevacizumab significantly reduced VEGF levels and nasal polyp cells.[6]

Maheshbabu et al found that budesonide nasal douching with saline significantly reduces postoperative polyps in chronic rhinosinusitis patients.[7,8]

A randomized trial by Pablo et al showed that Mometasone furoate sinus implants are effective in managing recurrent sinonasal polyps.[10]

The 2024 expert panel on adult pneumococcal vaccination advises that individuals aged 60 and above, and those with high-risk conditions such as COPD and asthma, receive the pneumococcal vaccine to lower infection risk.[9]

The primary treatment for nasal polyps in KS involves endoscopic removal followed by postoperative steroid therapy. Medical management includes antibiotics for sinus and lung infections and IV and oral steroids to reduce respiratory inflammation and edema. Biological therapies like bevacizumab and mepolizumab (anti-interleukin factors) are crucial for controlling nasal polyposis. However, further research is needed to better understand their role in recurrent nasal polyposis in KS.

CONCLUSION

Our patient had undergone nasal surgeries thrice in the past without being evaluated for KS. Once diagnosis of KS is made treatment in terms of aggressive steroid therapy, nasal surgery and adjuvant therapy can be given to control the disease and prevent recurrence. As clinicians we should keep KS as etiological factor while dealing with patients of recurrent nasal polyposis.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Handel MA, Kennedy JR. Situs inversus in homozygous mice without immotile cilia. J Hered. 1984;75(6):498.
2. Mittal V, Shah A. Situs inversus totalis: the association of Kartagener's syndrome with diffuse bronchiolitis and azoospermia. Arch Bronconeumol. 2012;48(5):179-82.
3. Afzelius BA, Mossberg B. Immotile cilia. Thorax. 1980;35:401-4.
4. Mishra M, Kumar N, Jaiswal A, Verma AK, Kant S. Kartagener's syndrome: A case series. Lung India. 2012;29:366-9.
5. Tangri N, Tangri P. 350 IJMSCR Kartagener Syndrome with Azoospermia: A Case Report. Int J Med Sci Curr Res. 2023;5(5):350-4.
6. Nemati S, Keihanian F, Saeidinia A, Bakhshaei M. Effect of Bevacizumab on Growth of Human Nasal Polyposis In Vitro; An Off-Label Use of Anti-Angiogenic Agent for Nasal Polyposis Treatment. Drug Des Devel Ther. 2019;13:3383-9.
7. Thanneru M, Lanke S, Kolavali S. The Effectiveness of Budesonide Nasal Irrigation After Endoscopic Sinus Surgery in Chronic Allergic Rhinosinusitis with Polyps. Indian J Otolaryngol Head Neck Surg. 2020;72(3):350-4.
8. Vakil AJ, Ojha T, Prasad S, Singh P. Comparison of Hypertonic Saline with Normal Saline in Nasal Irrigation Post Endoscopic Sinus Surgery. Indian J Otolaryngol Head Neck Surg. 2022;74(2):1518-22.
9. Koul PA, Vora AC, Jindal SK, Ramasubramanian V, Narayanan V, Tripathi SK, et al. Expert panel opinion on adult pneumococcal vaccination in the post-COVID era (NAP-EXPO Recommendations-2024). Lung India. 2024;41(4):307-17.
10. Stolovitzky JP, Kern RC, Han JK, Forwith KD, Ow RA, Wright SK, et al. In-office Placement of Mometasone Furoate Sinus Implants for Recurrent Nasal Polyps: A Pooled Analysis. Am J Rhinol Allergy. 2019;33(5):545-58.

Cite this article as: Shilpa C, Sandeep S, Unnikrishnan S. Unravelling Kartagener's syndrome in the face of recurrent polyposis: a case report. Int J Otorhinolaryngol Head Neck Surg 2025;11:285-7.