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Case Report

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A rare case of kartagener's syndrome: a clinical encounter

Abstract

Primary ciliary dyskinesia (PCD) is a rare congenital autosomal recessive disorder affecting the structure of cilia. It is seen in 1 in 10,000 -20,000 live births.[1] It is characterised by a triad of situs inversus, bronchiectasis and infertility. Although congenital in origin, the condition is frequently diagnosed later in life due to the absence of clear clinical markers, nonspecific presentation, and the need for complex diagnostic testing. Most commonly, a diagnosis is made later in life after multiple hospital admissions and a high degree of suspicion. Early diagnosis is very crucial to prevent permanent lung damage and to preserve fertility, wherever applicable. This case report aims to describe the clinical aspect of Kartagener's syndrome in a 24-year-old female.The peculiarity of the case gives insight into different types of presentation of PCD.

Keywords: primary ciliary dyskinesia, kartagener's syndrome, nasal polyposis

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Abbreviation: PCD, primary ciliary dyskinesia; OME, otitis media with effusion; EM, electron microscopy

Case report

A 24-year-old married female presented to our outpatient department with complaints of on-and-off left ear discharge since the age of 10 years, along with progressively decreased hearing, and a feeling of heaviness in the right ear and progressively increasing bilateral nasal blockage with facial pain for more than 5 years. Her history revealed bronchial asthma and episodes of shortness of breath for which the patient had multiple hospital admissions. She had been on short-acting beta 2 adrenergic agonists and inhalational corticosteroids for the past 10 years. She had also received anti-tubercular medication (ATT) for similar complaints 5 years back, but no improvement was noted.

Her family history revealed parental consanguineous marriage. There was no history of similar complaints or any other chronic conditions in the family. On enquiring regarding fertility history, the patient was nulligravid and reported inability to conceive despite regular unprotected intercourse for the last 2 years. However, she was not seeking any fertility treatment at the time of presentation.

Physical examination of the head and neck revealed that there was a central perforation in the left ear with active mucopurulent discharge, and the middle ear mucosa was congested. Examination of the right ear revealed a collection of fluid behind the tympanic membrane with multiple air bubbles. Audiometry showed bilateral mild conductive hearing loss (Figure 1).

On endoscopic examination of nose bilateral nasal polyposis with mucoid discharge was seen (Figure 2).

On auscultation, the heart sounds were audible on the right side, with minimal bilateral wheeze. An electrocardiogram (ECG) was done which was suggestive of dextrocardia(right axis deviation), showing inversion of all complexes in the lead I (inverted P wave, negative QRS, inverted T wave), upright P wave in the aVL lead and an absent R wave progression and prominence of S wave in the anterior leads and low voltage in leads V4-V6.

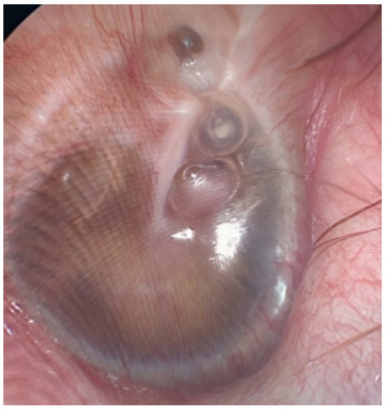

Figure 1 Otitis media with effusion in right middle ear.

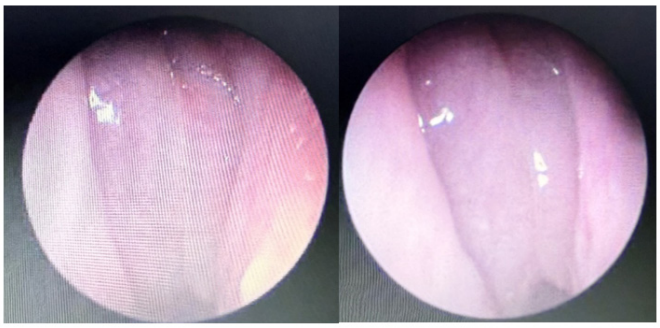

Figure 2 Showing nasal polyps in bilateral middle meatus on nasal endoscopy.

Chest X-ray (PA view) revealedbilateral prominent bronchopulmonary markings, dextrocardia, aortic arch on the right side and fundal gas shadow on the right side (Figure 3).

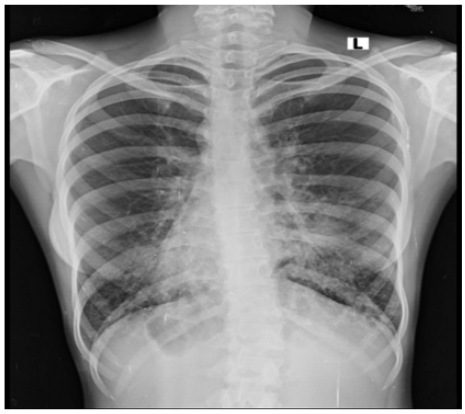

Figure 3 Chest X-ray (PA view) showing dextrocardia, with aortic arch and fundal gas shadow on the right side with bilateral prominent bronchopulmonary markings.

Haematological and biochemical parameters were within normal limits. Serum IgE was also done to rule out an allergic origin. X ray mastoid (Schuller's view) was suggestive of diploic mastoid on both sides (Figure 4).

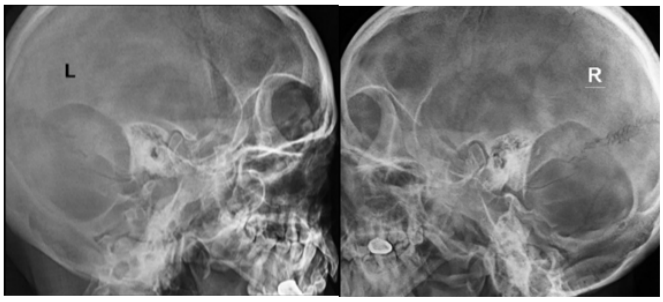

Figure 4 Showing diploicpneumatisation of mastoid bone.

Ultrasound (USG) of the abdomen confirmed situs inversus with the liver and gall bladder on the left side and spleen on the right side. A provisional diagnosis of primary ciliary dyskinesia (Kartagener syndrome) with left chronic otitis media, right OME with bilateral nasal polyposis was made and the patient was started on conservative management which consisted of Oral antibioticsalong with bronchodilators andintranasal steroids. She responded well to management and was kept on regular follow-up.

Discussion

Kartagener's syndrome is a rare autosomal recessive genetic disorder affecting ciliary function. It is characterised by sinusitis, situs inversus, and bronchiectasis. It was first described by *A Siewart* in 1904. Later, *Manes Kartagener* described the classical triad and reported it in four patients, in 1933.[2] Camner et al. in 1975 describe the causal relationship between sinopulmonary infections alongside immotile cilia syndrome.[3] It is a type of primary ciliary motility disorder. It affects more than 50 genes, most importantly DNAI1 & DNAH5, which are responsible for coding for the dynein protein.[4] These defects impair the function of the cilia responsible for respiratory system defence and embryonic visceral orientation during embryogenesis.[5,6] The prevalence ranges from 1 in 10,000 to 20,000 births. Diagnosis is often made late in adulthood after multiple hospital admissions requiring correlation of symptoms with a high degree of suspicion. Early diagnosis is important to preserve quality of life and life expectancy by preventing known complications.

Due to diminished ciliary function in almost every patient of Kartagener's syndrome, pansinusitis is seen. In later stages, it may even lead to hypoplasia of sinuses / silent sinus syndrome. Impaired eustachian tube function will cause otological manifestations like serous otitis media, chronic otitis media and recurrent otitis media with effusion alongside with audiological symptoms.[7] Bronchiectasis is localised and irreversible dilatation of the part of the bronchial tree, resulting in airflow obstruction and diminished clearance of secretions.[2] Stasis of secretions leads to recurrent respiratory infections. The classic symptoms include a chronic productive cough and difficulty in breathing. Later in life, it can lead to further diminution of pulmonary function due to pulmonary fibrosis. The mainstay of management for pulmonary complications includes culture-directed aggressive antibiotic therapy with bronchodilators. In non-responding patients, surgical interventions such as lobectomy or segmental resection,[2,8] may be beneficial.

In male patients, infertility is encountered more frequently than in female patients. In male patients, the infertility can be explained by immotile spermatozoa, whereas in females, it is due to impaired ovum transport from the ovaries to the fallopian tube. In females, defective ciliary movement can also lead to ectopic pregnancies. Infertility can often be managed with assisted reproductive techniques such as in vitro fertilisation.[9]

Diagnosis currently relies on a multidisciplinary team approach and electron microscopy examination of defective ciliary ultrastructure (>11Hz/s).[5] The sample for EM is obtained by either brush biopsy or nasal scraping of the respiratory cilia. In 3-30% cases of PCD the cilia can display normal structure. High-speed videomicroscopy can be performed to asses ciliary beat.[5] Different patterns of ciliary beat pattern (CBP) could be associated with specific ultrastructural defects. Another easily available test for screening is saccharin test which assess mucociliary function of nasal epithelium.[2] For screening purpose of the patients with PCD nasal nitric oxide (NO) level is measured which directly co relates with mucociliary function. It is found to very low in patients with PCD when compared with normal control subjects. This test can be performed in 5 years or older patients. It is a rapid, non-invasive and outpatient procedure. However, it still requires further research, as the diagnostic cutoff value has not yet clearly defined.

The objective of management includes control of the progression of the disease and symptomatic relief. Early diagnosis facilitates vaccination against Influenza and Pneumococcal disease, helping to prevent recurrent upper and lower respiratory tract infections, thereby reducing infection rates and hospital admissions. Treatment options include conservative management, surgical interventions for non-responders and adjuvant therapies.

Culture-based antibiotic treatment is found to be beneficial alongside inhalational/intranasal corticosteroids, mucolytics and bronchodilators. The role of chest physiotherapy is equally crucial in the management of Kartagener's syndrome. Surgical options may be offered to patients of KS with appropriate counselling regarding disease progression and recurrence chances. Functional endoscopic sinus surgery (FESS), lobectomy, or pneumonectomy can be offered on a case-by-case basis. Genetic counselling should be offered to patients or couples with Kartagener syndrome to reduce the risk of disease transmission to the offspring.

Conclusion

The author describes a rare case of Kartagener syndrome, that initially presented as a case of right ear chronic otitis media, along

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with its workup, management and a review of literature. The author highlights that there is no specific treatment protocol/ guidelines for this condition. Early diagnosis and management can preserve pulmonary function and thus prolonging the life expectancy of the patient.

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None.

Conflicts of interest

The authors declare that there are no conflicts of interest.

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