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CASE REPORT

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KARTAGENER'S SYNDROME – A CASE REPORT

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A B S T R A C T

Kartagener's syndrome is a rare, autosomal recessive genetic ciliary disorder comprising a triad of situs inversus, chronic sinusitis, and bronchiectasis. It is also called as Primary ciliary dyskinesia or immotile ciliary syndrome, the pathology involves defective movement of cilia leading to recurrent chest infections, ear, nose, throat symptoms, and infertility. Male to female sex ratio is 2 : 1. Here we present a case of kartagener's syndrome in a 15 years old female patient and a brief review of its literature.

INTRODUCTION

Kartagener's syndrome is a genetic condition with an autosomal recessive inheritance, comprising a triad of situs inversus, bronchiectasis and sinusitis[1,2]. Although Siewart first described this condition in 1904, it was Kartagener who recognized the etiological correlation between the elements of the triad and reported four cases in 1933 [3,4] The estimated prevalence of Primary ciliary dyskinesia is about 1 in 30,000.

The ultrastructural genetic defect leads to impaired ciliary motility which causes recurrent chest, ear/nose/throat (ENT), sinus infections, and infertility. A high index of suspicion is needed to make an early diagnosis so that timely treatment options may be offered for infertility in these young patients. It seems likely that early diagnosis is important for the preservation of pulmonary function, quality of life, and life expectancy in this disease[5].

Case Report

A 15 year old female came to our outpatient department with complaints of nasal obstruction on both side for 3 months duration associated with bleeding from nose. No history of cough with expectoration, fever, nasal discharge, head ache. No history suggestive of otitis media, gastrointestinal disease, cardiac failure, or genital infection, There was no family history of recurrent sino-pulmonary infection.

On general examination, the apex beat was in the right fifth intercostal space in midclavicular line with area of cardiac dullness on the right side. The liver was situated on the left side, with tympanic stomach resonance detected on the right side. Sinus examination revealed tenderness over maxillary sinuses. Respiratory system clinically within normal limits.

On ENT Examination, B/L Nasal cavities showed multiple greyish white polypoidal mass seen, (Fig. 1) insensitive to touch, Probe test confirmed mass arising from Lateral wall of nose. Other clinical examination was found to be within normal limits.

DISCUSSION

Disorders of ciliary motility may be congenital or acquired. Congenital disorders are labeled as Primary ciliary dyskinesia. Nearly 50% of Primary ciliary dyskinesia patients have situs inversus. Such cases of Primary ciliary dyskinesia with situs inversus are known as Kartagener's syndrome [3,5].

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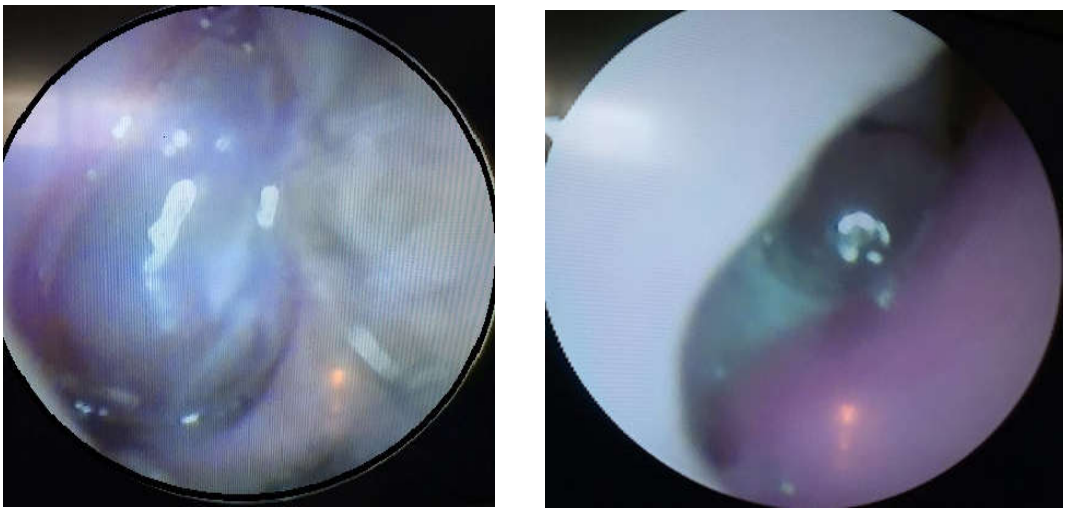

Figure 1 DNE Shows Bilateral nasal polyposis.

Echo cardiogram: Dextrocardia
USG Abdomen: Showed features suggestive of situs inversus totalis

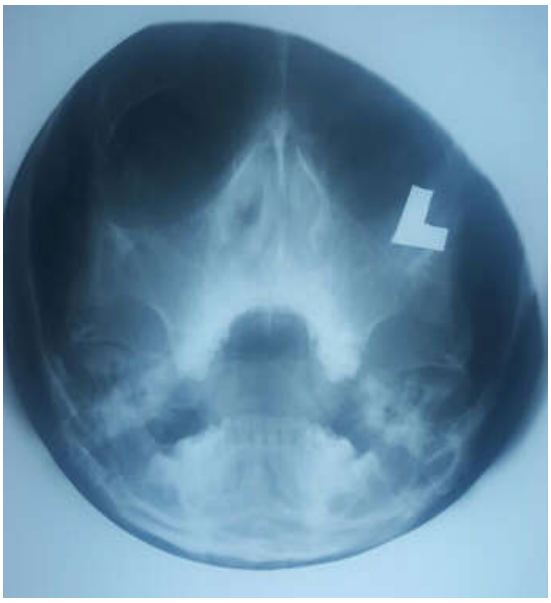

Figure – 2 X-ray paranasal sinus showing maxillary sinusitis and opacification seen in both nasal cavity

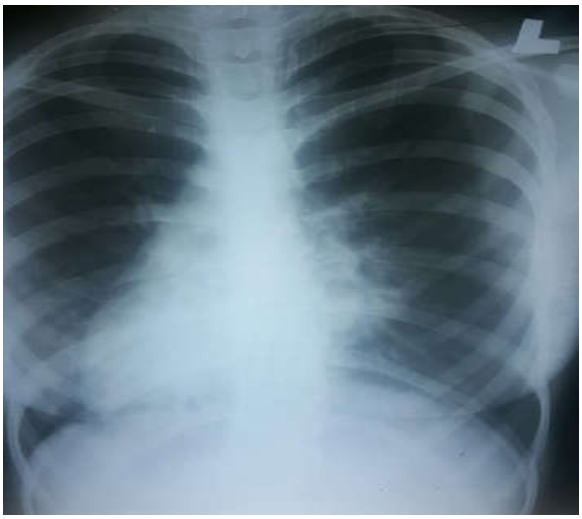

Figure – 3 X-ray chest, PA view, revealed right-sided heart and bilateral in homogeneous infiltrates in right middle zone and left lower zone.

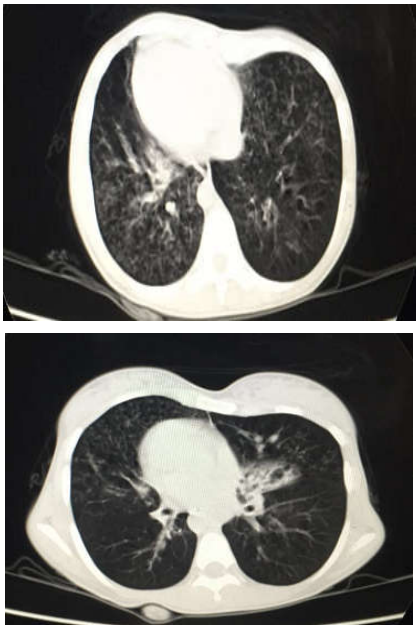

Figure – 4 On CT scan thorax, there was dextrocardia, B/L Central bronchiectasis involving left side lingual and right middle lobe. B/L Centrilobular nodular opacity.

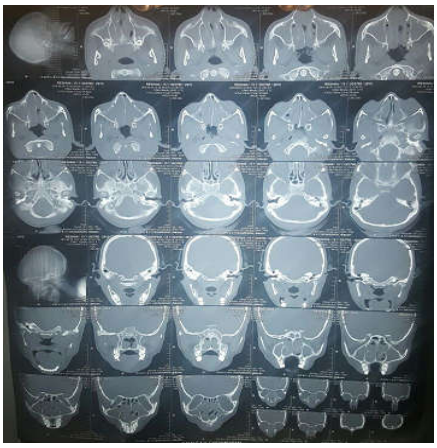

Figure – 5 CT PNS shows Soft tissue dense mass noted occupying entire Bilateral maxillary sinuses, Bilateral ethmoidal sinuses, Bilateral frontal sinuses extending in to the nasal cavity upto the chonae. Deviated nasal septum to the right side. Features suggestive of Sino nasal polyposis.

Primary ciliary dyskinesia is a phenotypically and genetically heterogeneous condition wherein the primary defect is in the ultrastructure or function of cilia. Patients with primary ciliary dyskinesia exhibit a wide range of defects in ciliary ultrastructure and motility, which ultimately impairs ciliary beating and mucociliary clearance [1,4].

Impair ciliary beating and mucociliary clearance are identified in approximately 90% of Primary ciliary dyskinesia patients and involve the outer dynein arms, inner dynein arms, or both. 38% of the Primary ciliary dyskinesia patients carry mutations of the dynein genes *DNAI* and *DNAH5*[4].

Patho physiologically, the underlying defect which leads to accumulation of secretions and consequent recurrent sinusitis, bronchiectasis, infertility, and situs inversus is the defective ciliary motility/immotility. The severity of symptoms and the age at which the condition is diagnosed is quite variable, even though the symptoms are present from birth [3,4,5].

Diagnostic criteria for this condition include clinical picture suggestive of recurrent chest infections, bronchitis, and rhinitis since childhood, along with one or more of the following: (1) situs inversus in the patient/sibling; (2) alive but immotile spermatozoa; (3) reduced or absent transbronchial muco ciliary clearance; and (4) cilia showing characteristic ultrastructural defect on electron microscopy[2,5,6].

Diagnosis of Primary ciliary dyskinesia

Screening tests: Exhaled nasal nitric oxide measurement which is usually low in Primary ciliary dyskinesia and saccharin test to assess mucociliary function of nasal epithelium.

Diagnostic tests: Ciliary beat pattern and frequency analysis using video recording, and electron microscopic confirmation of the ultrastructural ciliary defect. The samples for these tests for examining motility and ultrastructure of cilia may be obtained by biopsy of nasal mucosa and laparoscopic biopsies of tubal mucosa in females, as was done by Halbert *et al* [4,5].

In our cases, however, we did not perform these tests and the diagnosis was done clinico-radiologically (Fig. 2,3,4,5).

Infertility in male kartagener's syndrome patients is due to diminished sperm motility, while in females it is due to defective ovum transport because of dyskinetic motion of ovi ductal cilia, suggesting that the ciliated endosalpinx is essential for human reproduction. The development of assisted reproductive techniques has helped in these patients.

Treatment of this rare congenital disorder includes antibiotics to treat upper and lower airway infections. Hemophilus influenzae and *Staphylococcus aureus* are the most common organisms. Long-term low-dose prophylactic antibiotics may be necessary in children. Obstructive lung disease/bronchiectasis should be treated with inhaled bronchodilators, mucolytics, and chest physiotherapy. oral corticosteroids. Influenza and pneumococcal vaccination should be encouraged [5,7] .

CONCLUSION

Diagnosis of kartagener's syndrome is usually achieved by clinical features and radiological images. Kartagener's syndrome patients are frequently associated with repeated infection and infertility.

Early detection of kartagener's syndrome can reduce or even prevent the occurrence of severe respiratory infections, thus contributing to a better prognosis and an increase in the patient's survival.

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