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A Rare Case Report Kartagener Syndrome in a Seventeen Year Old Female

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Abstract: Kartagener's syndrome is an autosomal recessive disorder characterised by classical triad of situs inversus, bronchiectasis, and sinusitis. This syndrome is seen in 1 in 20000 to 1 in 60000. A 17 year old female student presented with recurrent cold, headache from the past 7 years. Last year she developed cough with expectoration .Her family history was not significant. On ENT examination she had tenderness over on frontal sinus. Ear and Throat examination were normal. Clinical examination revealed apical impulse on right side at 5th intercostals space at 1inch lateral to mid clavicular line. Heart sounds were prominent on right side. Respiratory examination was normal. Chest X-ray revealed Dextrocardia while Electrocardiography and 2D- Echo supported it. X-ray paranasal sinuses showed features of sinusitis. High Resolution computed Tomography showed bronchiectatic changes in mid zones and situs inversus. Spirometry shows borderline obstructive pattern. Sputum for Acid-fast bacilli negative and culture for pathogenic organisms remained sterile. Saccharin test was positive after 44 min. Invasive procedures were not done and diagnosis was made clinicoradiologically. As this syndrome is rare in females, fertility issues and hearing defects can be anticipated during course of time. Hence, thorough counselling and diagnosis (Otorhinological &Chest examination) should be made at the earliest. Early diagnosis and counselling will help the patients to take precautionary measures to prevent complications (i.e. fallopian tube blockage, conductive hearing defects, etc) in future..
Keywords: Kartagener's syndrome, autosomal disorder, bronchiectasis

INTRODUCTION

Kartagener syndrome is an autosomal recessive disorder characterised by classical triad of situs inversus, bronchiectasis and sinusitis [1]. This condition was first described by Siewert in 1904. Details were given by Manes Kartagener in 1933 and it is known by his name ever since. Kartagener syndrome is part of the larger group of disorders referred as Primary Ciliary Dyskinesia. Defective ciliary motility causes symptoms.

CASE REPORT

A seventeen year old female student presented with several episodes of rhinosinusitis for past seven years (three to five episodes per year). She complained of cough with expectoration since one year. Her parents' marriage was not consanguineous. On ENT examination she elicited tenderness over frontal sinus. Ear and Throat examination were normal. Clinical examination revealed apical impulse on right side at fifth intercostal space at one inch medial to mid clavicular line. Heart sounds were prominent on right side. Respiratory examination revealed crepitations in infra axillary regions bilaterally. Liver was palpated on left hypochondrium.

Chest X-ray Postero-anterior view showed cardiac silhouette and arch of aorta to the right side and the left dome of diaphragm is elevated, which was evident of situs inversus thoracis. HRCT-Chest and Abdomen: Arch of aorta on the right side, dextrocardia (situs inversus thoracis), liver on left side and fundus of stomach on right side (situs inversus abdominalis) confirming situs inversus totalis. Bronchiectatic changes seen on right middle lobe, lingular segment on left.

Sputum examination neither showed acid fast bacilli nor was positive for Grams stain.

2D-echo showed Dextrocardia and normal valves with ejection Fraction 65%.

ECG positive waves in aVR, and if the precordial leads placed on the right side.

Saccharin test was positive after 44 minutes.

Spirometry showed mild obstructive pattern.

The patient was treated for exacerbations with suitable antibiotics and physiotherapy. She was explained of her condition.

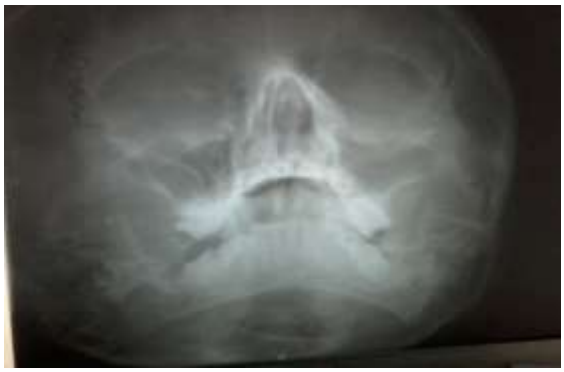

Fig. 1: Showing haziness in the frontal sinus

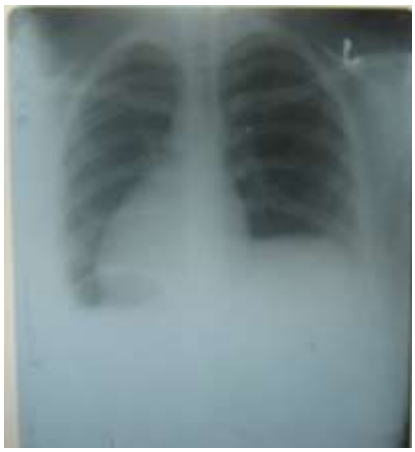

Fig. 2: Shows dextrocardia

Fig. 3: Showing dextrocardia (situs inversus thoracis)

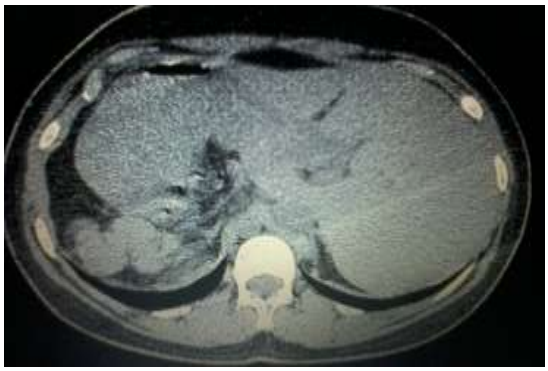

Fig. 4: Showing Situs inversus abdominalis

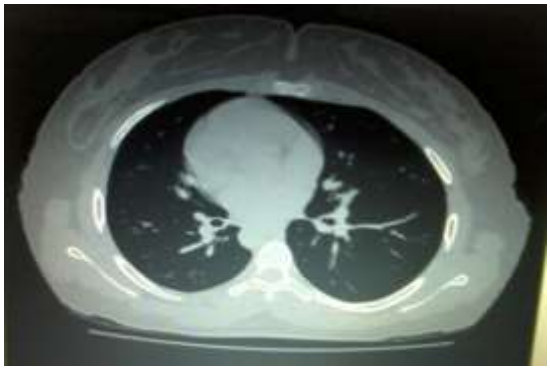

Fig. 5: Showing mild bronchiectatic changes

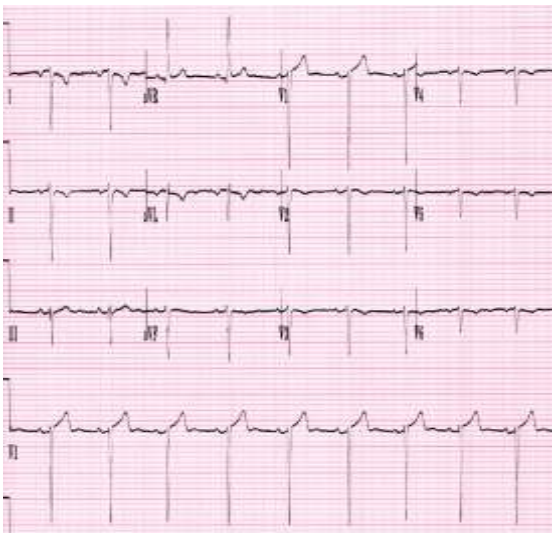

Fig. 6: Showing ECG changes

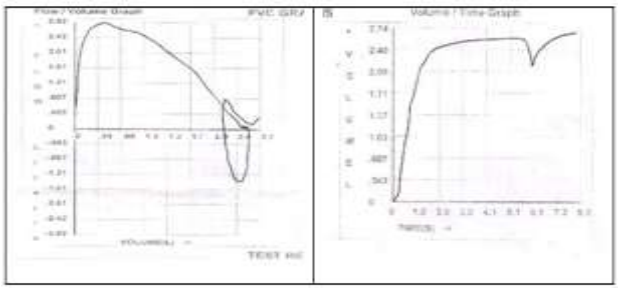

Fig. 7: Spirometry showing borderline obstructive changes

Table 1: Borderline Obstruction noted

Pred.	Actual	% Pred.
2.334	2.71	116.1
2.142	1.824	85.15
84.47	67.29	79.67

DISCUSSION

Majority of patients present to Physician 20-40 times for rhinosinusitis, before the actual diagnosis of Kartagener Syndrome is made. The diagnosis is made difficult because patient present with usual complaints of bronchitis and sinusitis [2]. 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]. This syndrome is rare in females, can occur 1 in 20000 to 30000.As this is a ciliary disease, it affects nasal mucosa, paranasal sinuses, middle ear, eustachian tube, pharynx ,fallopian tubes in females, epidydimis in males. Women with Kartagener syndrome are infertile sometimes due to lack of cilia motility in the lining of fallopian tube. Chronic respiratory and sinus infection are controlled by antibiotics and chest physiotherapy(to clear the mucus) [4]. To confirm this diagnosis, we have to perform exhale NO, biopsy of nasal mucosa and laproscopic studies of tubal mucosa [5]. But these tests were not performed and the diagnosis is made on clinic-radiological basis. The suspicion is made when patient was found to be having dextrocardia which is usually present in more than fifty percent of individuals in Primary Ciliary dyskinesia. Kartagener is most common in Primary Ciliary Dyskinesia. Thorough otorhinological examination along with chest examination would help in early diagnosis .Most patients have normal life span .Chronic rhinosinusitis with dextrocardia should be suspected for Kartagener syndrome and bronchiectasis should be evaluated. The delayed diagnosis can lead to adverse side effects, and inappropriate treatment [6]. The treatment of bronchiectasis include respiratory supervision at regular intervals, combined with physiotherapy, and treatment of upper and lower respiratory tract infections [7]. Physiotherapy aims to clear impacted secretions caused due to impacted sputum due to bronchiectasis and is proved to be better than bronchodilator therapy.

Fertility issues and conducting hearing deafness can be anticipated in due course of time. Infertility in male KS patients is due to diminished sperm motility, while in females it is due to defective ovum transport because of dyskinetic motion of oviductal cilia, suggesting that the ciliated endosalpinx is essential for human reproduction [8].

CONCLUSION

Fertility depends on degree and efficiency of cilia in fallopian tubes which is variable in women with this syndrome [9]. 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, wherever feasible. Also, although unproven, it seems likely that early diagnosis is important for the preservation of pulmonary function, quality of life, and life expectancy in this disease [10].

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