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Case Report

Familial Kartageners Syndrome: A Case Report

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Abstract

In recurrent lower respiratory tract infections the cause may be either general impairment of immune mechanism, abnormalities of mucus or abnormalities of cilia.

We report a case where the patient was having recurrent lower and upper respiratory tract infections and had situs Inversus totalis. Kartagener's syndrome is an inherited disorder transmitted in autosomal recessive manner with variable penetrance. There is no specific treatment for this condition but failure to recognize the condition early in life may subject the patient to unnecessary repeated admissions and investigations and inappropriate treatment.

Keywords: Kartagener's Syndrome; Dextrocardia; Situs Inversus; Bronchiectasis; Sinusitis

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Case Reports

A 24 year old male patient hailing from a rural area presented with productive cough, rhinorrhoea and headaches for last 16 years, with episodic fever and worsening of symptoms. His previous record showed a lot of investigations including investigations for tuberculosis such as repeated chest X-Rays and examination of the sputum. Record also revealed that he received several courses of antibiotics, antihistamines, bronchodilators, inhaled and oral corticosteriods but the response was only partial and temporary. Recived AntiTubercular Therapy twice. Because of his ill health, duration of symptoms and uncertainty about the cause of the disease, patient was

admitted in our hospital. On examination he was febrile, pulse rate 110/minute, blood pressure 110/84 mmHg, he had nasal discharge, chest auscultation showed bilateral diffuse wheeze and coarse crackles. His heart sounds were heard best on the right side of the chest. There was no digital clubbing. Initial suspicion was that of bronchial asthma with recurrent chest infections and the possibility of cystic fibrosis was also kept in mind. However chest X ray showed not only bronchiectasis especially in the lower zones but also dextro cardia (Fig 1). Sinus radiographs showed mucosal thickening opacified sinus cavities and other features of chronic sinusitis (Fig 2). Semen analysis showed azoospermia. Contrast Enhanced Computed Tomography (CECT) showed sinusitis, bronchiectasis and situs Inversus (fig 3). Considering the clinical picture of the patient, sinusitis, bronchiectasis and situs Inversus, the clinical diagnosis of Kartagener's Syndrome was made. The condition was explained to the patient and he was treated with antibiotics, antipyretics, mucolytics and inhaled bronchodilators.

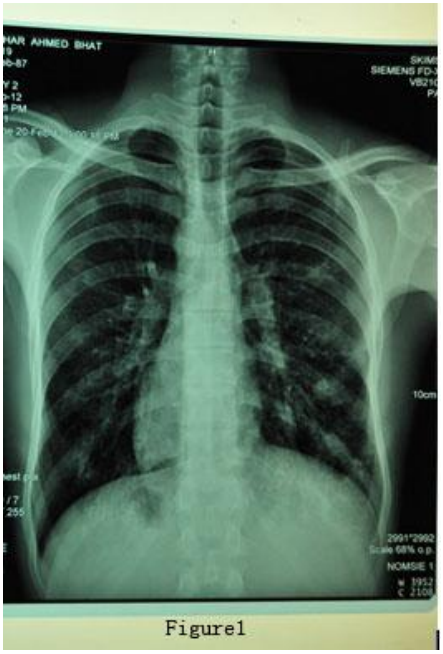

Figure1

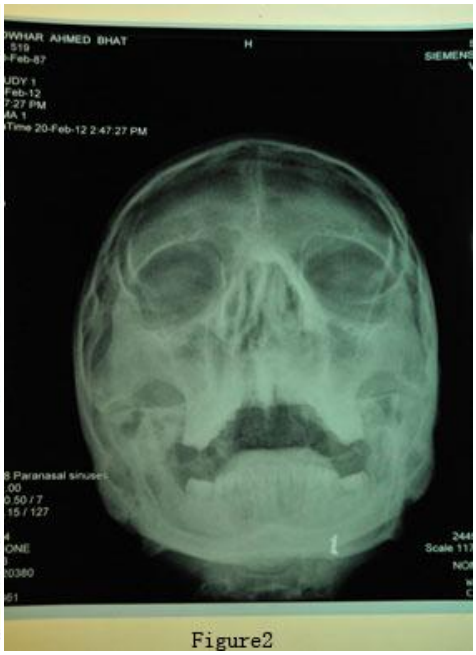

Figure2

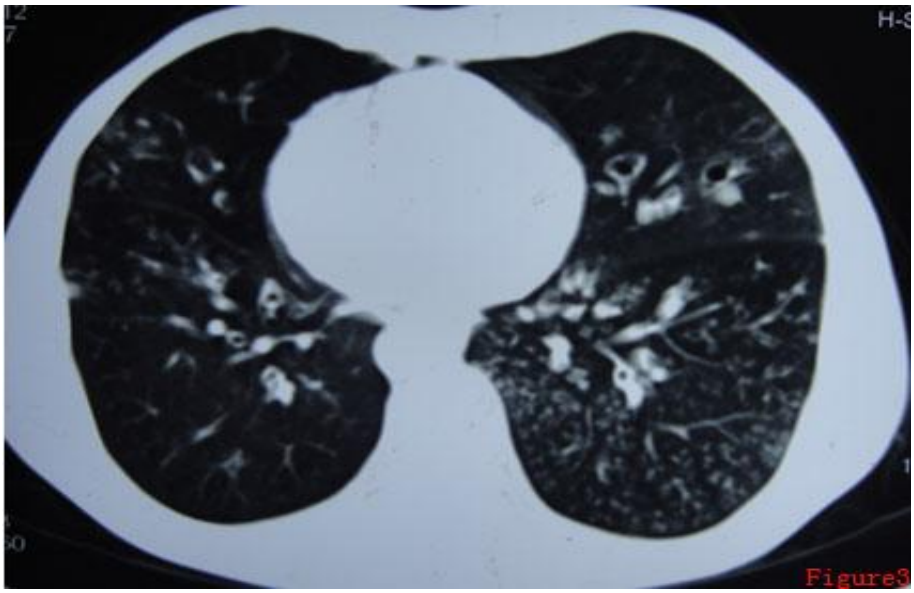

Figure3

He had a significant family history as his brother also had similar complaints and was married for 8 years without any children. He was operated once for a lung abscess.

Discussion

Kartagener's Syndrome is a rare, an autosomal recessive disorder [1] characterised by a triad of dextrocardia, bronchiectasis and sinusitis [2]. The condition was described for the first time by Siewert in 1904, therefore some people call it Siewert Syndrome but the details of the condition were given by Manes Kartagener in 1933 and it is commonly known as Kartagener's Syndrome. Sinusitis, situs inversus, bronchiectasis and male infertility occurring in this condition are attributed to abnormal ciliary motility. Cilia may be immotile or may show uncoordinated and inefficient movement patterns. Ciliary movement disorders may be congenital or acquired. Congenital ciliary disorders are labelled as primary ciliary dyskinesias (PCDS). Approximately one half of the patients with PCD have situs inversus. Those patients having PCD with situs inversus are known as Kartagener's Syndrome [3].

It has been proposed that abnormal ciliary beating is necessary for visceral rotation during embryonic development. In patients with PCD half of the patients will have situs inversus i.e. will be cases of Kartagener's Syndrome and the other half normal situs because of random rotation [4]. Abnormal ciliary motility results in general impairment of respiratory defence mechanisms due to problem with bacterial clearance leading to recurrent upper and lower respiratory tract infections. In abnormalities of cilia, structural abnormalities of dynein arms are the most common [5], although an abnormality of the radial spokes and microtubules can also account for the condition. In rare cases no structural ciliary abnormality is detected even though ciliary function is abnormal and the clinical syndrome is typical [6-7]. Patients with Kartagener's Syndrome may have either situs solitus i.e. dextrocardia only or situs inversus totalis where all the viscera are on the opposite side [4], including left sided appendix [8]. The patient in this case was having situs inversus totalis.

Lung CT- Scan demonstrates peribronchial thickening and bronchiectasis, which is mostly marked in the lower zones [9]. High resolution CT shows that pulmonary disease related to PCD predominantly involves the middle and lower lobes of the lungs [10]. Although pneumonia is frequently seen in Kartagener's Syndrome, lung abscess infrequently accompanies it [11].

Cutaneous lesions in Kartagener's Syndrome are nummular eczema, recurrent deep folliculitis and pyoderma gangeraenosum [12]. No cutaneous lesions were seen in our patient.

The diagnosis of Kartagener's Syndrome is made clinically and confirmed through transmission electron microscopy [13]. Screening test for PCD includes nasal nitric oxide and in vivo tests of ciliary motility such as the Saccharin test. Specific diagnosis requires examination of cilia by light and electron microscopies with epithelial culture in doubtful cases14. Since there is no specific therapy for PCD, it is recommended that upon diagnosis, secondary infections be treated with potent antibiotics and prophylactic interventions [15].

In the 3 families, 17 siblings were studies: 4 had Kartagener's syndrome and another 7 showed recurrent respiratory infections [16].

Some interesting associations of this syndrome such as reversible airways obstructions (5 out of 11 patients), pulmonary

tuberculosis (3 out of 11 patients). Pleuritis (3 out of 11 patients) reported by Goyal et al. [17]

In a conclusion, prompt and appropriate treatment of respiratory infections can minimize irreversible lung damage in such cases.

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