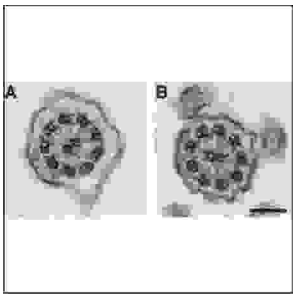
CASE REPORT PROF-983

KARTAGENER SYNDROME;
A CASE PRESENTATION



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ABSTRACT... We are reporting a case of Kartagener syndrome in a 19 year old young male, who presented to us with chronic sinusitis, bronchiectasis and dextrocardia.

INTRODUCTION

Kartagener syndrome is a rare subgroup of primary ciliary dyskinesia, which is characterized by chronic sinusitis, bronchiectasis and situs-inversus. Kartagener syndrome makes half[6] of the cases of primary ciliary dyskinesia. It has a prevalence[3] of 1 in 32000 live births. It was first described by Siewert in 1904. Manes kartagener recognized the clinical triad (Chronic Sinusitis, Bronchiectasis and Situs-inversus) as a congenital syndrome in 1933. Pathologically[4] there are ultra-structural defect of the cilia, there may be reduction of the dynein arms, absence of radial spokes and the defects of central microtubule. Due to these ultra-structural defects there are unco-ordinated movements of the cilia resulting in ineffective mucociliary clearance. Patient presents with repeated upper and lower respiratory tract infections leading to chronic sinusitis, otitis media, chronic bronchitis, recurrent pneumonia and bronchiectasis. Young male patients may presents with infertility due to immotile sperms. On examination dextrocardia may be found. X-ray of paranasal sinuses may show thick mucosa, hypo-plastic frontal sinuses and sinus cavity. Chest X ray[7] may include crowded bronchial markings related to peribronchial fibrosis and small cystic spaces at the base of lung. HRCT is the diagnostic study of choice for bronchiectasis. Ultrasonography abdomen may show situs-inverses.

In the past bronchography was the gold standard to demonstrate bronchiectatic changes but due to the nature of the test, it has been replaced by HRCT. Semen analysis shows immotile sperms. Mucosal biopsies show ultra-structural defect under electron microscope. Saccharine test[5] may be delayed. Pulmonary function test

demonstrate obstructive pattern.

CASE REPORT

This nineteen years old male presented to us with an upper respiratory tract infection and shortness of breath. He gives the history of repeated chest infections since childhood. During these episodes there was history of excessive production of yellow coloured, sticky sputum associated with cough and high grade fever, which was relieved by antibiotics and antipyretics. He also gives history of episodic frontal headache, post nasal drip and rhinorrhea. There is history of episodes of hemoptysis and progressive shortness of breath. General physical examination at the time of admission showed, pulse 88/min, blood pressure was 110/70, temperature 100F, respiratory rate is 20/min. Clubbing and cyanosis was positive. Examination of respiratory system revealed coarse crepitations with scattered rhonchi all over the chest. Apex beat was palpable on the right side of the chest in right lateral position. First and second heart sounds were normal.

Gastrointestinal tract and central nervous system were normal. Nose examination showed thick nasal mucosa with mucopurulent discharge. Ear examination was normal. Lab. Investigation showed Hb = 11.5 gm/dl, TLC = 12300 / , ESR = 40 mm/1st hour, Polymorph = 71%, lymphocytes = 26%, RBS = 113 mg/dl, urea = 35 mg/dl, urine complete examination was normal, semen analysis showed 12 millions sperm/ml. 100% sperm were immotile. Chest X ray showed bilateral lower zone infiltrate with dextrocardia. X ray PNS showed haziness of maxillary and frontal sinuses along with mucosal thickening. CT chest showed fibrocystic changes with areas of infiltration in both lower lobes involving basal segments. Heart apex and his appendages were lying on right side. Liver was lying on left side and spleen on right side suggestive of situs-inversus and bronchiectasis. Ultrasonography abdomen revealed similar findings showing transposition of liver and inferior vena cava on the left side and spleen and aorta on right side. Audiometry test was normal. Saccharine test was positive. On the basis of bronchiectasis, chronic sinusitis, situs-inversus and immotile sperms the diagnosis of Kartagener Syndrome was made.

DISCUSSION

Kartagener syndrome is a rare form of primary ciliary dyskinesia. It occurs in 1 in 32000 live births. It is characterised[1] by chronic sinusitis, bronchiectasis and situs-inversus. All these feature were present in our patient. Nasal polyps are recognized in 30% of affected individual which were not found in our patient. Recurrent sinusitis and otitis media are common manifestation a of primary ciliary dyskinesia. The later may lead to conductive deafness. The ear examination in our patient was normal. Lower[2] respiratory tract infections leading to chronic bronchitis recurrent pneumonia and bronchiectasis are other common manifestation which were present in our patient. Dextrocardia is present in half of patients with primary ciliary dyskinesia in European series. In Polynesians with ciliary dyskinesia organ position is normal. Young syndrome is an other variant of primary ciliary dyskinesia, who present with chronic sinusitis and bronchiectasis but they have azoospermia due to obstruction at the level of vasa deferentia. Other causes of early onset bronchiectasis are cystic fibrosis, allergic broncho-pulmoponary aspergillosis, immunoglobulin deficience and post infectious bronchiectasis. Treatment of this condition comprises mainly of broncho-dilators, postural drainage and antibiotics.

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