Research Paper

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Case Report of Kartagener's Syndrome

Medical Science

KEYWORDS :

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ABSTRACT

KARTAGENER'S SYNDROME is a rare cilliopathic autosomal recessive genetic disorder that is characterize by impaired function of mucosal cilia.Patients generally present with recurrent respiratory tract infections most commonly caused by pseudomonas infection and bronchiectasis.Situs inversus may be associated in about half of the patients. Diagnosis can be confirmed by tests to prove ciliary dysfunction, mucosal biopsy and genetic studies.Treatment is supportive.In severe cases bilateral lung transplantation may be warranted. We present a case of a 13 year old male patient presented to us with history of recurrent episodes of URTI and pneumonia. Imaging studies revealed a triad of sinusitis,bronchiectasisand situs inversus. Ciliary dysfunction was demonstrated by positive saccharin test. Patient was managed conservatively and was advised regarding vaccination.Aim of this case report is to increase awareness about this rare disease so as to provide early diagnosis and improve the prognosis in affected individuals.

INTRODUCTION:

Kartagener's syndrome is an autosomal recessive conditionoccurring with a frequency of 1:30,000 – 1:40,000. It ischaracterized by the classic triad of situs inversus, bronchiectasisand sinusitis. The condition was first described by Siewert in 1904but details of the condition were given by Manes Kartagener in 1933 and it is known by his name ever since.

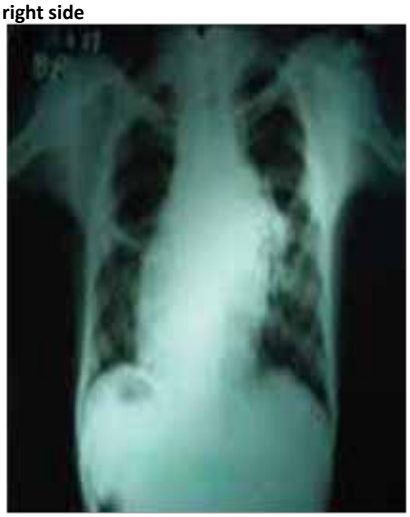
CASE REPORT

A 13 years old male patient presented with complaints of recurrent episodes of cough,cold and headache forone year and high grade fever with chills for 7 days with no significant past history and no bad habits.On physical examination patient had raised temperature of 101 degree F with pulse rate of 90/min/regular and 110/70mmhg of blood pressure in supine position.He was tachypneic with RR of 30/min. Nasal examination showed sinus tenderness and edematous mucosa.On respiratory examination coarse crepitations were present in bilateral lung fields. CVS examination was unremarkable except for a loud S2.No abnormality was found on Abdomen and CNS examination.

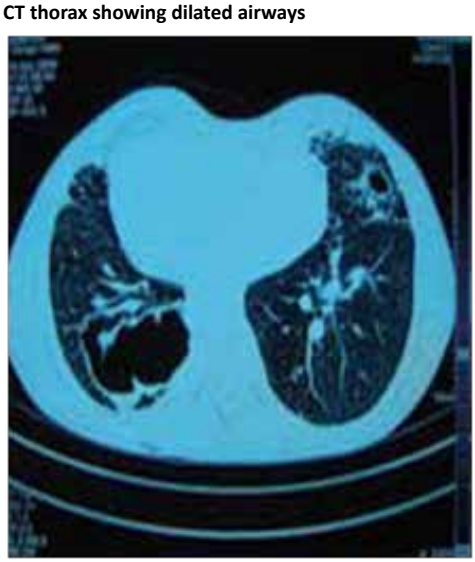
Patient's blood count showed polymorphoneutrophilic leucocytosis and XRAY CHEST-PA was suggestive of dextrocardia with bilateral lung field opacities suggestive of consolidation.XRAY PNS had changes of bilateral maxillary sinusitis.USG abdomen confirmed situs inversus by showing the liver on the left side and stomach and spleen on the right side.HRCT thorax confirmed dextrocardia as well as bilateral upper and lower lobes bronchiectasis.Changes of T inversion in lead 1,avL, V2 to V6 were evident in ECG s/o dextrocardia.A positive saccharin test confirmed ciliary dysfunction.

Patient was managed conservatively with higher antibiotics,nebulizations and chest physiotherapy. He was advised for pneumococcal and Hib immunization on follow up.

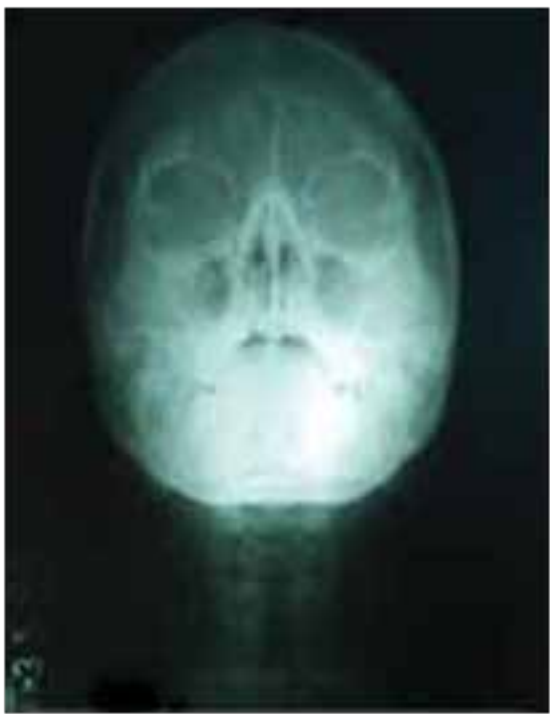
Chest X ray showing dextrocardia and gastric bubble on right side



CT thorax showing dilated airways



X ray PNS showing maxillary sinusitis



Discussion

Kartagener's syndrome in seen in 50% cases of primary ciliary dyskinesia. Primary ciliary dyskinesia is an inherited autosomal recessive condition characterized by bronchiectasis, sinusitis and otitis media. When situs inversus is associated with primary ciliary dyskinesia then it is referred to as Kartagener's syndrome. Numerous defects have been described in both the conditions including abnormalities in dynein arms, radial spokes and microtubules. As a result of these abnormalities cilia become dyskinetic and their propulsive action is diminished. This leads to impaired bacterial clearance resulting in recurrent lower and upper respiratory tract infections. Also because sperm motility depends on proper ciliary movement, males with this condition become invariably infertile. Additionally since visceral rotation during development depends upon ciliary motion, situs inversus is seen in Kartagener's syndrome. Demonstration of abnormal ciliary movement needs electron microscopic studies of biopsies obtained from nasal mucosa or trachea. However these procedures are invasive and available at specialized centres only. Diagnosis of this condition is usually clinical accompanied by imaging studies.

Medical care for a person with Kartagener syndrome focuses on prevention of repiratory infections,and prompt treatment of any that may occur.Antibiotics can relieve sinusitis,and inhaled medications and respiratory therapy can help if chronic lung disease develops.Small tubes may be placed through the eardrums to allow infections and fluid to drain out of the middle ear.Adults especially men may have difficulty with fertility,and may benefit from consulting a fertility specialist.

In many individuals the number of the respiratory infections begins to decrease by about age 20 and as a result many people with Kartagener syndrome have near normal adult lives

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