www.jmscr.igmpublication.org
Impact Factor (SJIF): 6.379
Index Copernicus Value: 79.54
ISSN (e)-2347-176x ISSN (p) 2455-0450
crossrefDOI: https://dx.doi.org/10.18535/jmscr/v6i10.41

IGM Publication
Journal Of Medical Science And Clinical Research
An Official Publication Of IGM Publication

Absence of Arm and a Broken Family

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Abstract
Here we present a case of 59 year old male patient brought to the casualty with dyspnea, swelling of both lower limbs of 7 days duration. Patient was treated many times as COPD acute exacerbation but on further detailed history and investigations was found to have kartagener's syndrome.

Introduction

The lower respiratory tract contains ciliated epithelium from the trachea to respiratory bronchioles. Each ciliated cell gives rise to approximately 200 cilia that vary in length from 5 to 6 micrometres and decrease in size as airway becomes smaller. Patients with primary ciliary dyskinesia exhibit a wide range of defects in ciliary ultra structure, which impair ciliary motility and mucociliary clearance. The most common defect, first described by Afzelius, is a reduction in number of dynein arms, which decreases the ciliary beat frequency. The clinical consequence of the immotile cilia syndrome includes chronic cough with expectoration, bronchiectasis, chronic rhinitis with nasal polyposis, chronic and recurrent sinusitis and often agenesis of the frontal sinuses. Obstructive lung disuses may develop. Most clinical manifestations develop from early childhood itself and neonatal asphyxia occurs often.

Siewart first described the combination of situs inversus, chronic sinusitis with bronchiectasis in 1904. However Manes Kartagener described this syndrome in detail. Afzelius was the first to recognize the relationship between kartagener syndrome and male infertility when he observed lack of dyne in arm in the sperms and cilia of some subjects

Case Report

59 year old male patient brought to the casualty with dyspnea, swelling of both lower limbs of 7 days duration. Patient gives history of recurrent episodes of dyspnea suggestive of a chronic respiratory illness from childhood. He is on regular medications initially oral bronchodilators, later inhalation therapy with steroid and long acting bronchodilators. He was hospitalized several times for exacerbations and also gives history of pulmonary tuberculosis at the age of 40 for which he took full course of ATT regimen. He got married at the age of 29. Marriage lasted only for 3 years

On examination patient is moderately built and nourished, dyspnoeic, pallor, cyanosis and clubbing were present. Bilateral pitting pedal edema was also present. No dermatological

manifestations of active TB or malignancy was there.

Respiratory system examination shows accessory muscles of respiration working with bilateral extensive coarse crepitations more on the right lower lobe and bilateral rhonchi more on right lower lobe present

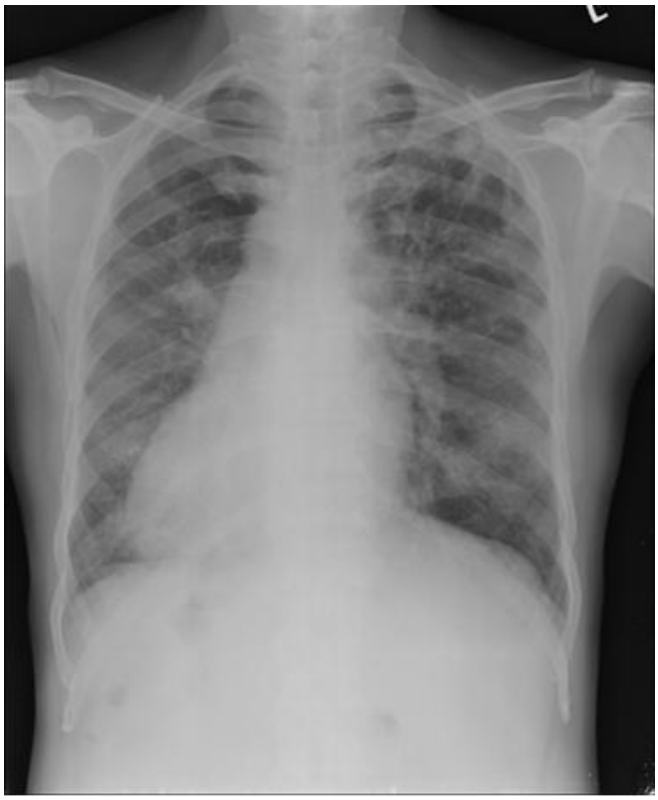
Cardiovascular system examination showed elevated JVP with prominent a wave with JVP of 7 cm from sternal angle. Cardiac impulse is felt on the 5th right intercostal space in the midclavicular line. A systolic murmur in the second left intercostal space heard.

Examination of abdomen revealed liver palpable on the left side, no splenomegaly but abdominal wall edema was present. Other systemic examination is non contributory.

So clinically we suspect the possibility of a COPD with asthmatic element presenting with corpulmonale. In view of clubbing, possibility of bilateral bronchiectasis more on the right side was also concerned. On re eliciting the history patient gives history of recurrent attack of bifrontal headache with nasal discharge suggestive of chronic sinusitis was also obtained. While obtaining family history we came to know that he has taken treatment for infertility and he is divorced. So clinically we suspect the possibility of a kartagener syndrome was concerned and investigated.

Investigations

- Hb-13.5
- TC-5,900
- Platelet count-2 L
- ESR-40
- PCV-44
- DC-P75, L25
- FBS-100, PPBS-120
- Urea/Creatinine-34/1.2
- Na/K-135/3.9
- S.bilirubin-1.2
- T Protein/Albumin-5.7/2.7
- SGOT/SGPT-46/11
- ALP-131
- Lipid profile-NAD
- Ana profile-Normal
- Serum electrophoresis-Normal
- CRP, VDRL,VIRAL Markers-Normal
- Alpha 1 antitrypsin- Normal
- Sputum culture and sensitivity – NAD
- Semen analysis-normal
- AFB staining and culture- NAD
- Blood culture -NORMAL
- ECG- Right ventricular hypertrophy, P Pulmonale +
- X-ray- dextrocardia, cardiomegaly +



Chest x-ray PA view showing dextrocardia and bilateral fluffy opacities

Echocardiography- RVH present

HRCT suggestive of bilateral bronchiectatic changes with dextrocardia

CT Paranasal Sinuses-Sinusitis

MRI Brain-Normal

Discussion

Kartagener syndrome isknown as primary ciliary dyskinesia. It is a rare genetic lung disorder. The defect is in the action of cilia lining the respiratory tract. It occurs in 1 out of 32,000 live births. The

triad consists of bronchiectasis, situs inversus and sinusitis. In situs inversus there is mirror image of organs. It is an autosomal recessive condition in which microscopic cells in the respiratory epithelium do not function normally. Dynein arms, radial spokes or central apparatus are missing leading to defective or impaired ciliary movement. Ciliary dysfunction impairs the clearing of mucus from sinus, respiratory tract, ear Abnormal monocilia in embryo cause defective rotation of organs in 50 percent of the patients. Impaired cilia of the oviduct cause infertility in females and non motile sperms cause infertility in males.

The importance is that early diagnosis helps in proper management of the cases and prevention of complications. In our patient, there is recurrent hospital admissions and he was treated many times as acute exacerbation of COPD and finally he presented with congestive cardiac failure. Early diagnosis could have prevented the development of cardiac failure. Fertility can also be attained by TESE-ICSI (spermatozoa extracted from testes). It improved the morphology of spermatozoa and fertilization rate. In our patient due to late diagnosis such techniques were not adapted and he ended up in having a broken family.

Conclusion

Before labelling every chronic case of breathlessness as COPD we should consider every other possibilities like ILD, bronchiectasis, rare conditions producing breathlessness because late diagnosis might result in irreversible damage and patient may end up in having serious complication like cardiac failure. These days even the infertility part of kartagener syndrome can be addressed and the patient can lead a normal life.