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# EMPYEMA THORACIS IN KARTAGENER SYNDROME: AN UNUSUAL PRESENTATION WITH ABDOMINAL HETEROTAXY, LEVOCARDIA, AND POLYSPLENIA

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**Abstract:**

*Kartagener's syndrome (KS) is a rare autosomal recessive disorder classified under primary ciliary dyskinesia (PCD), characterized by the classic triad of bronchiectasis, chronic sinusitis, and situs inversus. However, atypical presentations can occur, posing diagnostic challenges. We present a unique case of a 35-year-old female with KS, exhibiting abdominal heterotaxy with isolated levocardia, empyema thoracis, and polysplenia. The patient, who had a lifelong history of recurrent respiratory tract infections, presented with a chronic cough, pleuritic chest pain, and an abdominal mass on the right side. Imaging including a high-resolution computed tomography (HRCT) of the chest revealing bilateral bronchiectasis, granuloma and pleural effusion on the left side, and a computed tomography (CT) with contrast of the abdomen & pelvis revealed a left-sided liver, and multiple spleens on the right depicting as a mass, consistent with situs inversus, yet the heart remained in its normal left-sided position (levocardia). The absence of prior flu vaccinations, and history of recurrent respiratory infections, underscores the importance of early recognition and intervention in KS to prevent complications such as empyema thoracis. This case highlights the variability in KS presentations and emphasizes the need for awareness of its potential impact on fertility, as evidenced by the patient's decade-long history of primary infertility. Treatment remains symptomatic, focusing on managing respiratory infections and preventing future complications.*

*Keywords: Kartagener's syndrome, Abdominal Heterotaxy, Bronchiectasis, Empyema thoracis, Situs inversus, Infertility*

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## INTRODUCTION:

Kartagener's syndrome (KS) is classified as a subgroup in various ciliary motility illnesses recognized as primary ciliary dyskinesia (PCD)[1]. Kartagener's syndrome is an autosomal recessive disease characterized by diminished ciliary motility leading to a unique spectrum of presentations. The classic triad of Kartagener's syndrome includes bronchiectasis, situs inversus, and chronic sinusitis[2]. The prevalence of Kartagener's syndrome is roughly estimated to be 1 in 30,000 live births. Normal ciliary motility is an essential component of respiratory defense mechanisms, sperm motility, and congruous orientation of the viscera during development. The gene mutations (dnai1 and dnah5) in KS result in reduced ciliary movement leading to infertility, left-right misalignment of body organs, and repeated respiratory tract infections [3] . The criteria based on which we can diagnose Kartagener's syndrome comprises a past medical history of recurrent Sino-pulmonary infections with one or more of the given features: (a) situs inversus, heterotaxy or dextrocardia, (b) infertility due to impaired ciliary motility, (c ) impaired clearance of tracheobronchial secretions and irritants, (d) electron microscopic evidence of structural defects in cilia [4-5] . Investigations for the diagnosis of KS include evidence of bronchiectasis and situs inversus on a CT scan of the chest and abdomen respectively. Furthermore, screening tests include exhaled nasal nitric oxide level determination and saccharin test for assessing ciliary function. Tem for structural defects in cilia and genetic testing for *dnai1* and *dnah5* mutations confirm Kartagener's syndrome. [4-6]

## CASE PRESENTATION:

A 35-year-old married female presented with complaints of chronic cough, breathing difficulty, right flank pain, and left-sided pleuritic chest pain radiating to the left scapula and tip of the left shoulder. The cough was dry, minimally productive of greenish to grey sputum. Chest pain exacerbated with coughing.

On arrival, her bp was 120/70 pulse 106, temp of 102 Fahrenheit and spo2 87% on room air.

Her past medical & surgical history along with medication history was insignificant. She reported repeated upper respiratory tract infections and common flu that would occur once every three months or so accompanied by greenish to grey discharge that was foul smelling. She reported having this cycle of infections from childhood around 4 years of age. She did not receive any flu vaccines previously.

On examination, she was oriented in time, place and person. She was febrile, having a fever of 101 Fahrenheit, and pale conjunctiva. The chest examination revealed decreased air entry in the left lower lung, fine crepitations in the middle and lower zones of her lungs bilaterally, and dull percussion on the left lower side of the chest. She did not have any clubbing or cyanosis. Abdomen was soft and non-tender. The cardiovascular examination was normal.

The chest x-ray done was suggestive of bronchiectatic changes bilaterally and blunting of the left cardiopulmonary angle pointing towards left sided pleural effusion as shown in in Figure 1.

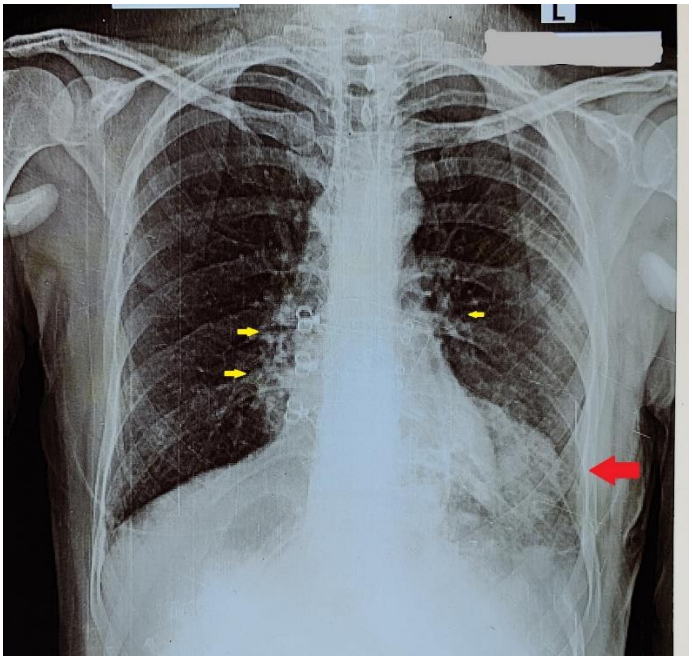

**Figure 1: Chest Xray showing cardiopulmonary angle blunting on the right side and bronchiectatic changes bilaterally (Red arrow - Cardiopulmonary angle blunting on the left side. Yellow arrows - bronchiectatic changes bilaterally)**

Computed tomography (CT) with contrast of the abdomen & pelvis revealed a left-sided abdominal mass reporting the presence of liver on the left side with polysplenia in the right hypochondrium, collectively as a part of abdominal situs inversus (Figure 2). Also, a small non-obstructing 5 mm calculi was noted in the right renal pelvis which was the cause of her right flank pain.

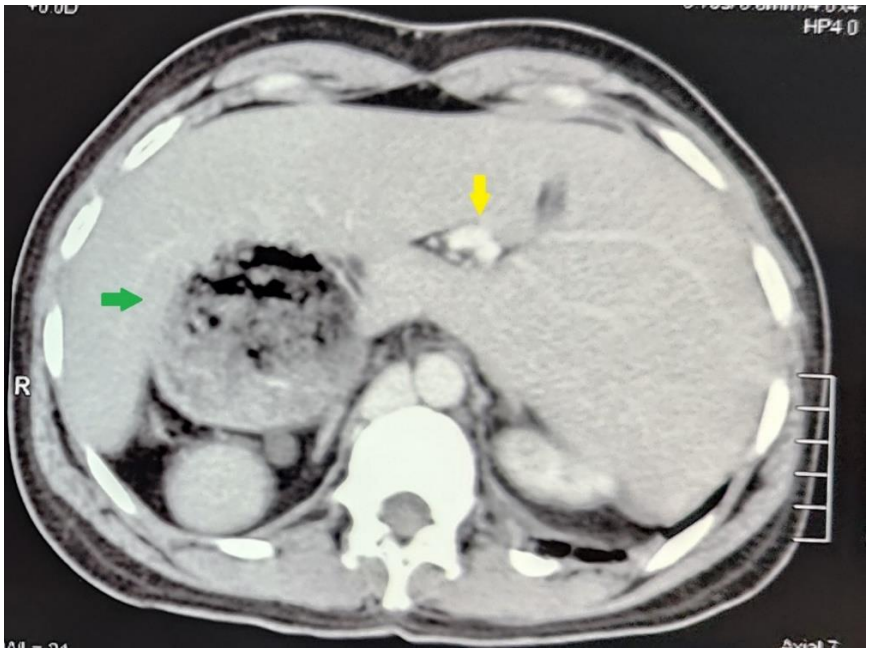


**Figure 2: CT abdomen with contrast showing liver on the left side and stomach on the right (Yellow arrow - liver hilum on left side. Green arrow - stomach on right side)**

High-resolution computed tomography (HRCT) of the chest revealed bronchiectatic changes in bilateral middle and lower zones, along with granulomas and pleural effusion in the basal left lung as shown in in Figure 3.

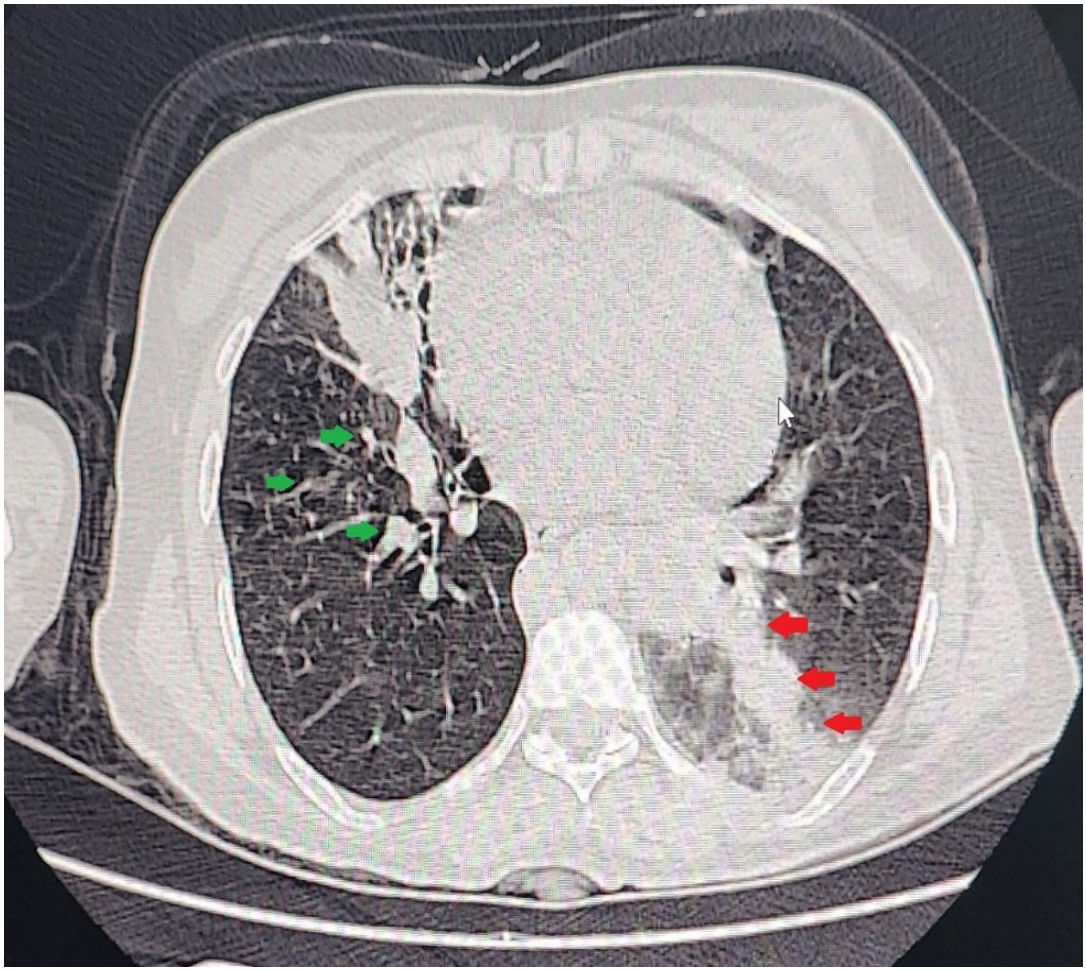

**Figure 3: High resolution computed tomography of the chest (HRCT) showing bronchiectatic changes bilaterally and granuloma in the left (Green arrows - bronchiectatic changes on the right. Red arrows - granulomas in the basal segments of left lung)**

Base line laboratory investigations including complete blood picture and inflammatory markers were done.

**Table 1: Complete blood picture timeline**

| Complete blood picture | | | | |
|---|---|---|---|---|
| | On arrival | 3rd day of admission | 7th day | 10th day (discharge) |
| White blood cells | 22.1 x 10 | 25.7 x 10 | 18.5 x 10 | 10.4 |
| Hemoglobin | 11 | 11 | 11.2 | 11.5 |
| Platelets | 714 | 693 | 840 x 10 | 810 x 10 |
| Neutrophils % | 85 % | 90 % | 84 % | 83 % |
| Lymphocytes % | 10 % | 6 % | 14 % | 11 % |

Ultrasonography of the chest revealed thick fluid collection of 332 ml in the lower segment of the left lung and no collection in the right lung. Diagnostic pleural tap was done and sent for pleural fluid R/E, culture and sensitivity testing, and fluid cytology.

Blood cultures were taken and no growth after 48 hours; no growth after 96 hours.

Gene Xpert (sputum): no acid-fast bacilli detected

Pleural fluid culture: no growth after 48 hours

Pleural fluid cytology: no malignant cells seen. Note is made of smears showing neutrophils, lymphocytes and plasma cells scattered in a thick proteinaceous background.

Pleural fluid r/e: volume - 8 ml, color - purulent, cell count - 1550, neutrophils - 90% & lymphocytes 10%, protein - 4.4 gm/dl indicative of empyema.

Gene Xpert, sputum culture, and sputum acid-fast bacilli (AFB) were done as part of a workup to rule out pulmonary tuberculosis. All the investigations were negative for tuberculosis.

Sputum culture: no growth after 48 hours

Sputum Acid-fast bacilli: no AFB seen
Sputum gene Xpert: no mycobacterium tuberculosis detected

She was diagnosed with Kartagener syndrome. Although Kartagener syndrome mostly presents with dextrocardia, isolated levocardia in Kartagener syndrome is a rare presentation with the presence of multiple spleenules.

She was started on broad spectrum antibiotic i.e. Inj. Piperacillin-tazobactam 4.5 gram three times a day intravenously, 2 liters of oxygen via nasal cannula improving her oxygen saturation to 95%, paracetamol tablets for pain relief and pyrexia, and mucolytics in the form of n-acetylcysteine sachet were added to help thin her secretions. She reported improvement in her breathing difficulty and pleuritic chest pain.

Additionally, ultrasound guided pig-tail catheter was placed in her left hemithorax to facilitate drainage of the empyema (Figure 4).

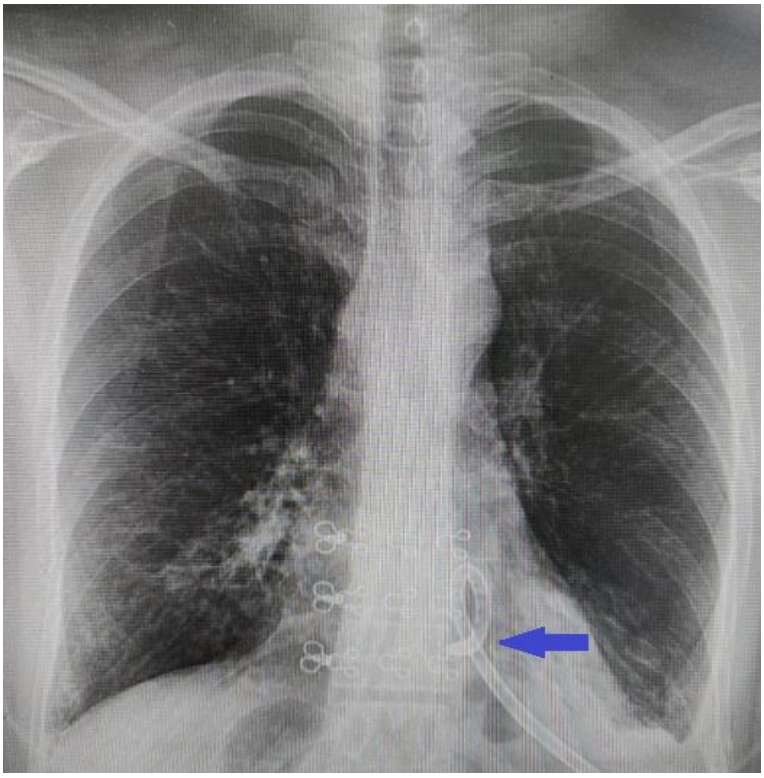

**Figure 4: Chest x-ray showing pigtail insertion (Blue arrow: pig tail tube in the pleural cavity on the left side)**

The patient responded well to treatment and was discharged after 10 days of inpatient stay. She was subsequently sent home with the pig-tail catheter in place and was extubated 2 weeks later on follow up.

On follow-up visit, she reported good health with no signs and symptoms. A repeat chest x-ray showed complete resolution of empyema and optimal lung expansion (Figure 5).

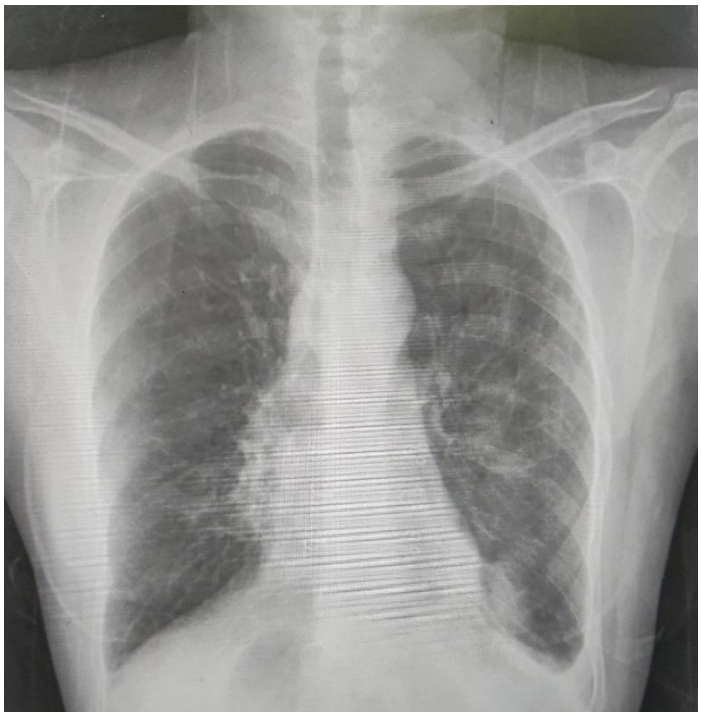

**Figure 5: Follow-up chest x-ray showing complete lung expansion and empyema resolution**

She was previously not aware of her condition. She had been married for the last 10 years, has no history of contraceptive use, and has not conceived to date. She has 4 brothers and 4 sisters; all married and have children. She was counselled about her diagnosis in detail and referred to a genetic specialist and fertility specialist to help with her family building.

## DISCUSSION:

Kartagener syndrome's hallmark feature is situs inversus, dextrocardia. However, this case is unique due to levocardia, where the heart remains in its usual left-sided position despite the mirror-image arrangement of other organs (abdominal heterotaxy). The presence of multiple spleens adds to the rarity of this presentation.

The disease symptoms and manifestations can range from recurrent upper respiratory infection (sinusitis, rhino sinusitis) to recurrent ear infection to chronic productive cough due to bronchiectasis, ectopic pregnancy, and subfertility in females [7]. Clinical and radiological evidence of bronchiectasis develop as the disease progresses,[8] as in our patient.

Our patient had a history of recurrent sinopulmonary infections and infertility. Imaging findings showed bronchiectasis, and situs inversus with levocardia. Laboratory screening and confirmatory testing which required a better clinical setup were not done

Early recognition of Kartagener syndrome is critical to prevent severe complications such as empyema thoracis. Chronic respiratory infections, a common feature of the syndrome, often lead to repeated hospitalizations and significant morbidity if not managed promptly. Additionally, primary infertility is a well-documented issue in both males and females with Kartagener syndrome due to dysfunctional cilia in the reproductive tract.

Unfortunately, there is no definitive treatment for Kartagener syndrome. Treatment is symptomatic. Chest infection and bronchiectasis are treated with bronchodilators, antibiotics, mucolytics, and corticosteroids. Influenza and pneumococcal vaccines are also necessary to prevent recurrent infections.[9]

Early diagnosis and follow-up are essential in these patients to prevent complications. These patients also need counseling regarding conception.

## CONCLUSION:

This case underscores the importance of early diagnosis of Kartagener syndrome. Awareness and recognition of this condition can lead to timely

interventions, reducing the risk of complications such as empyema thoracis and recurrent upper respiratory tract infections (URTIs). Furthermore, acknowledging the potential for primary infertility in young couples with Kartagener syndrome is crucial for providing appropriate counseling and management, thereby alleviating the mental distress associated with infertility.

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