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Kartagener Syndrome Found Incidentally During Assessment of Respiratory Infection: A Case Report

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Abstract **Case Report**

Kartagener syndrome, a very rare genetic disease with autosomal recessive transmission, is described by the triad of bronchiectasis, chronic sinusitis and complete or incomplete situs inversus. This syndrome is part of primary ciliary dyskinesia, it's characterized by clinical symptoms of respiratory infections or/and infertility. Situs inversus is often discovered incidentally. The diagnosis is confirmed by specialized centers through analysis of ciliary ultrastructure and genetic testing. Treatment is supportive and in accordance with recommendations.

Keywords: Kartagener's syndrome, Imaging, CT Scan, sinusitis, situs inversus, bronchiectasis.

INTRODUCTION

Kartagener syndrome is a distinct entity among primary ciliary dyskinesia, characterized by a clinical triad of sinusitis, bronchiectasis, and complete or incomplete situs inversus [1]. It is a rare congenital disease with autosomal recessive transmission [2, 3]. Respiratory tract infections in the course of the disease typically manifest from childhood [2]. Kartagener syndrome, in addition to upper and lower respiratory tract infections, may cause also male infertility [4].

We report a case of this rare pathology revealed by a respiratory infection, discussing the contribution of imaging in the diagnosis and reviewing the literature.

OBSERVATION

A 39 year-old man, followed for recurrent respiratory infections since his childhood, some episodes of sinusitis, with no history of another medical disease or surgery, who presented to the emergency room with breathing difficulty, Sadoul stage II dyspnea associated with cough and mucous expectorations for a year and a half, with recent worsening characterized by stage III dyspnea and mucopurulent expectorations.

Clinically he had a heart rate of 108 beats per minute; blood pressure was 10/5.5 cm Hg, oxygen saturation of 91% in ambient air. On physical examination, he was polypneic, with crackling rales on auscultation in both lung bases and a right sided apex beat, all evolving in the context of unquantified fever without weight loss. a scout view (Equivalent to a standard frontal X-ray in the chest CT scan) showed bilateral areolar and cylindrical bronchiectasis, left scissuritis, left pleural effusion and dextrocardia (Figure 1). The Chest CT scan showed complete situs inversus (Figure 2, 3, 4 and 6), bilateral bronchiectasis, associated with infectious bronchopneumopathy (Figure 5 (a and b)), probably related to Kartagener syndrome. The infectious assessment was positive. He received antibiotic and symptomatic treatment, bronchial drainage physiotherapy with anti-flu and anti-pneumococcal vaccination with a good clinical improvement of these respiratory symptoms.

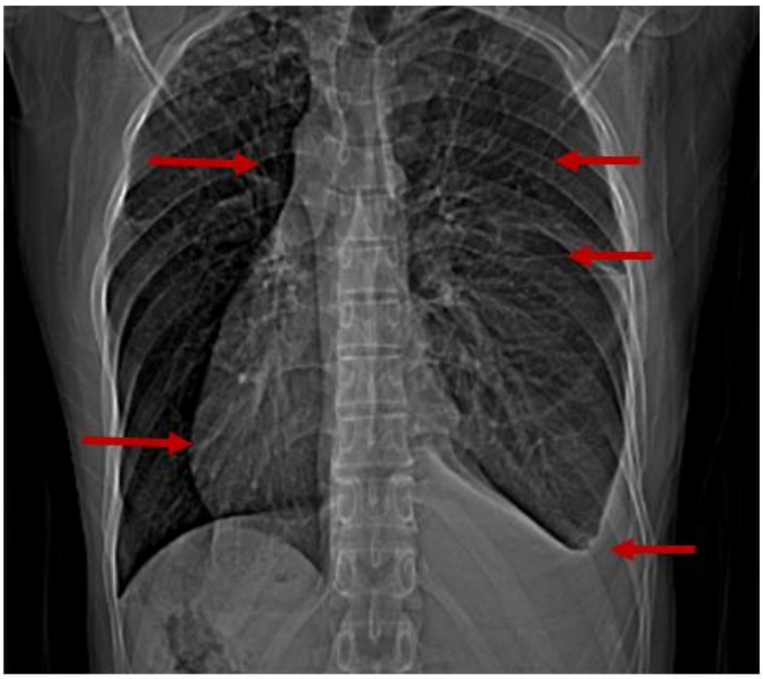

Figure 1: Scout view (Equivalent to a standard frontal X-ray in the chest CT scan) showing: bilateral areolar and cylindrical bronchiectasis, left scissuritis, reticular infiltrates, left pleural effusion and dextrocardia

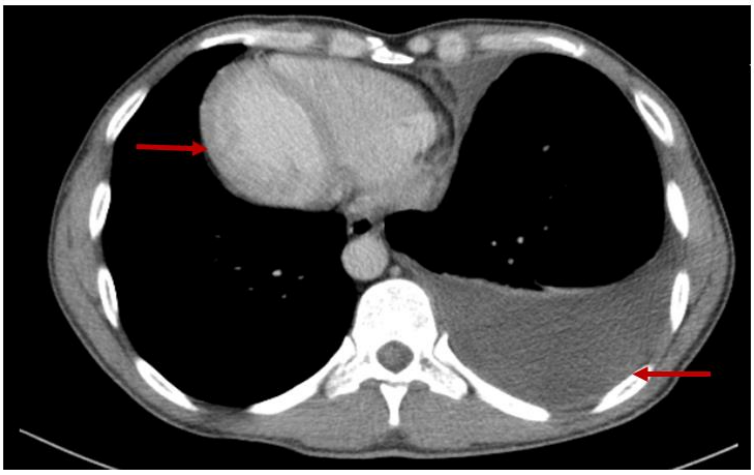

Figure 2: Chest CT scan, axial section in mediastinal window, showing dextrocardia with left pleural effusion

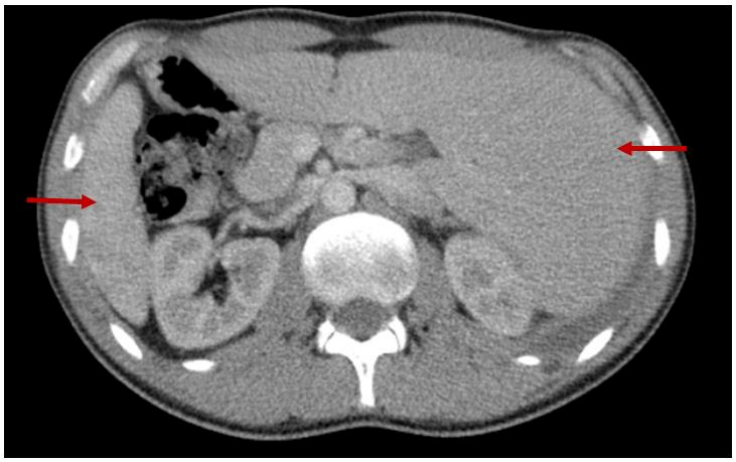

Figure 3: Chest CT scan, axial section in mediastinal window, showing the spleen and stomach on the right; the liver is located in the left hypochondrium

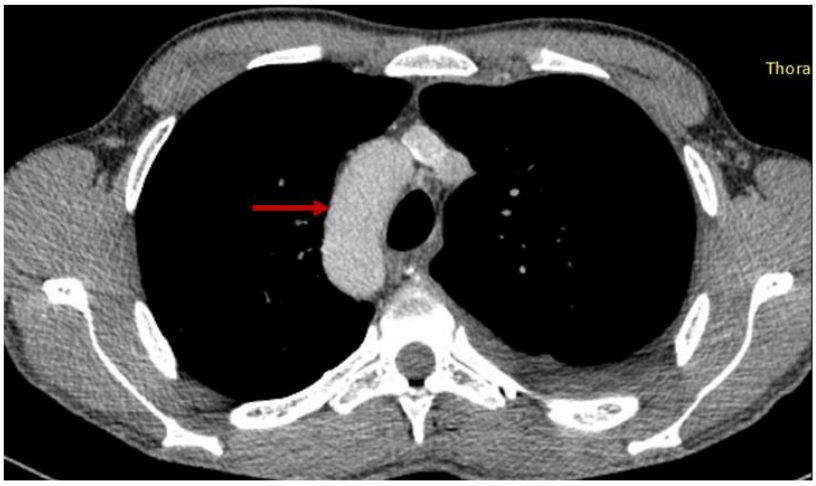

Figure 4: Chest CT scan, axial section in mediastinal window, showing aortic arch oriented to the right

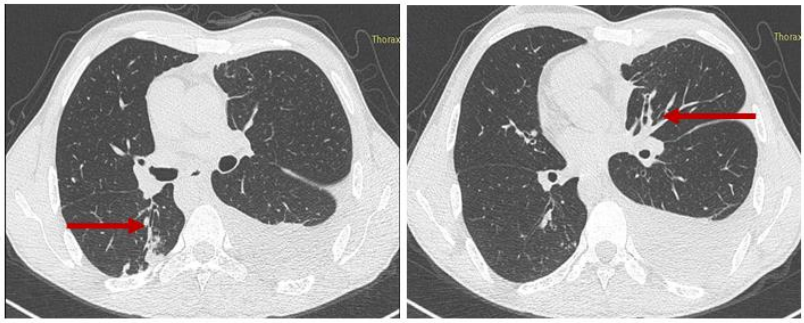

Figure 5 (a and b): Chest CT scan, axial section in parenchymal window, showing bilateral bronchiectasis with some mucoid impactions and thickening of the bronchial walls

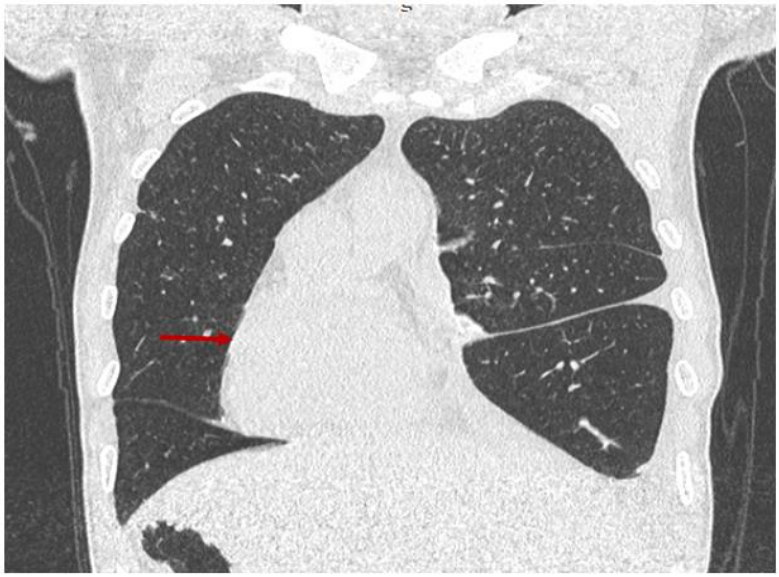

Figure 6: Chest CT scan, coronal section in parenchymal window, showing dextrocardia.

DISCUSSION

In 1935, Manes Kartagener first identified the syndrome that now bears his name, which is characterized by a combination of chronic sinusitis, situs inversus and bronchiectasis [5]. Kartagener syndrome is responsible of 50 % of cases of primary ciliary dyskinesia (PCD). Although it is a rare genetic disorder that is inherited in an autosomal recessive manner, other modes of inheritance, such as X-linked or dominant, have also been reported [6].

The syndrome results from mutations in genes that code for dynein, located on chromosomes 5, 9 and 7, causing structural and/or functional abnormalities in the cilia [6]. The age of diagnosis varies among researchers,

with some reporting it as early as childhood [3, 5–8]. In our case, the patient was a 39 year-old man, similar to the case reported by Prisca Gabrielle *et al.*, [1], who also observed the syndrome in an adult. Moreau [6] observed that males were more commonly affected, while Melki [8] reported a series of six female patients. Some researchers have also documented cases in neonates [3, 5]. In our case, there was no evidence of a family history of chronic respiratory disease or parental consanguinity.

Kartagener's syndrome is characterized by respiratory signs that manifest during childhood and dominate the symptomatology. These signs, which are present in all patients, are marked by their chronicity and annual recurrence [1]. They include chronic bronchial congestion, a persistent cough that produces mucopurulent secretions on a daily basis, and episodes of broncho pneumopathy, bronchiectasis and mucopurulent sputum, as described in the literature with cough and dyspnea [6]. ENT involvement, particularly in the upper airways, is characterized by sinusitis and otitis, which are associated with abnormal mucociliary clearance. In this case, the patient had a history of recurrent episodes of sinusitis since childhood. Nasosinus polyposis is observed in 30 % of patients and is a common feature in various inflammatory nasosinus diseases [3, 5, 6, 8].

Imaging is crucial in evaluating this condition; it allows the detection of radiological signs of this syndrome that require further genetic study to confirm the diagnosis. On the Chest CT scan (Figure 2, 3, 4, 5 and 6), we noted a complete situs inversus, which included dextrocardia and spleen and stomach located in the right side, while the liver and gallbladder were located on the left side. Furthermore, there was a rotational anomaly affecting the lungs and abdominal vessels, with the inferior vena cava situated on the left side of the aorta. A moderately abundant left pleural effusion was noted which may be the cause of the worsening of his symptoms. The CT scans also revealed cylindrical and cystic apical traction bronchiectasis of the right upper lobe and right Fowler's lobe with mucoid impaction and wall thickening, viewed in the lung parenchymal window. Additionally, there were branching micronodules and foci of sub pleural condensation. Bronchial involvement is typically seen in the middle and lower lobes, with an increase in bronchiectasis prevalence as age advances [1]. The progression of respiratory pathology varies, with early diagnosis and strict management being crucial factors [2]. Respiratory physiotherapy for bronchial drainage, antibiotic therapy for surinfection, and sufficient vaccination coverage against influenza and pneumococcal are the mainstays of managing Kartagener's syndrome [3, 5, 6]. Engaging in physical activity is strongly advised, as it facilitates bronchial drainage and the ability to exertion [9].

CONCLUSION

Kartagener syndrome remains a rare disease that may be compatible with a normal life if the diagnosis and support are early. However, in the forms involving significant lung lesions, the patient's prognosis is involved in the short term, due to respiratory complications but also cardiovascular.

Conflict of interest: The authors report no conflicts.

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