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Medical Imaging Diagnosis of a Rare Association of Kartagener Syndrome, Common Mesentery, Left Isomerism: A Case Report in Niamey (NIGER)

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ABSTRACT

Kartagener's syndrome, defined by the situs inversus-bronchiectasis-chronic sinusitis triad, is in 50% of cases associated with PCD, which can be responsible for numerous visceral positional defects ranging from complete situs inversus to ambiguous situs (isomerism) which is characterized by enormous organ lateralization defects during embryogenesis with one or more organs in symmetry or even duplicated. All these abnormalities can only be diagnosed with certainty with the help of medical imaging examinations, more specifically CT. We report here the case of a 67-year-old male patient admitted for radiological workup for cough, sinus syndrome to whom the diagnosis of kartagener syndrome (situs inversus-dilatation of the bronchi-chronic pan sinusitis) associated with a mesentery common, left isomerism (left lung isomerism and polysplenia) was done.

KEYWORDS: Kartagener syndrome, common mesentery, left lung isomerism, polysplenia, Niamey

ARTICLE DETAILS

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INTRODUCTION

Kartagener's syndrome is defined by the clinical triad: situs inversus-bronchiectasis-chronic sinusitis. It is a hereditary disease with autosomal recessive transmission and incomplete penetrance. This syndrome is part of the primary ciliary dyskinesias which constitute a heterogeneous group of genetic respiratory diseases linked to a constitutional anomaly of the cilia of the upper (sinuses) and lower (bronchi) respiratory mucous membranes [1]. More than 70% of cases present with a mutation of the gene coding for dynein, a protein essential for ciliary movements [2]. We report a rare case presenting not only Kartagener's syndrome on a left isomerism chart but also a complete inverted type common mesentery.

PATIENT AND OBSERVATION

This is a 67-year-old male patient admitted for an X-ray assessment for chronic cough evolving since childhood in a context of dyspnea and sinus syndrome. The imaging examinations carried out were: pulmonary X-ray, a thoraco-abdomino-pelvic (TAP) CT scan and a sinus CT scan.

Chest X-ray was performed with a Neusoft machine and revealed dextrocardia, widening of the upper mediastinum, gastric air pocket under the right diaphragmatic dome (figure 1).

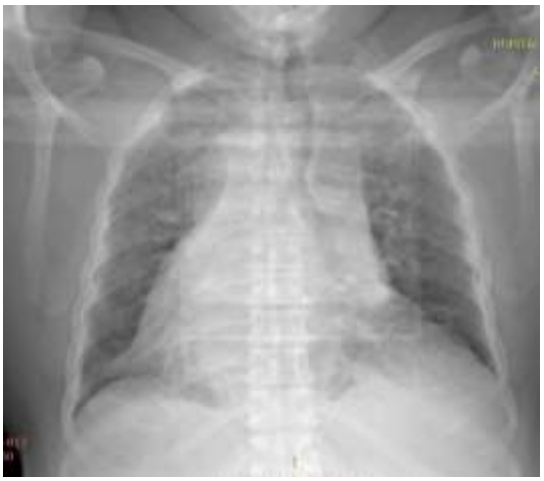

Figure 1: Pulmonary X-ray taken standing from the front in a 67-year-old patient showing dextrocardia, widening of the superior mediastinum, deviation of the trachea.

An x-ray of the sinuses of the face in Blondeau view, also indicated, showed bilateral fronto-maxillary sinusitis. Faced with this picture, a thoraco-abdomino-pelvic and sinus CT scan was requested.

For the thoraco-abdomino-pelvic scanner, the constants used were 120KV and 40mA and the cuts made were 1.5mm contiguous. The examination was performed in a fasting patient, in supine position, head first. Two series of acquisitions were performed, one without injection of contrast product and the other with injection of contrast product. The scan showed:

- At the thoracic level: the arch of the aorta is oriented to the right, the apex of the heart also oriented to the right, the descending aorta to the right (figures 2 and 3).

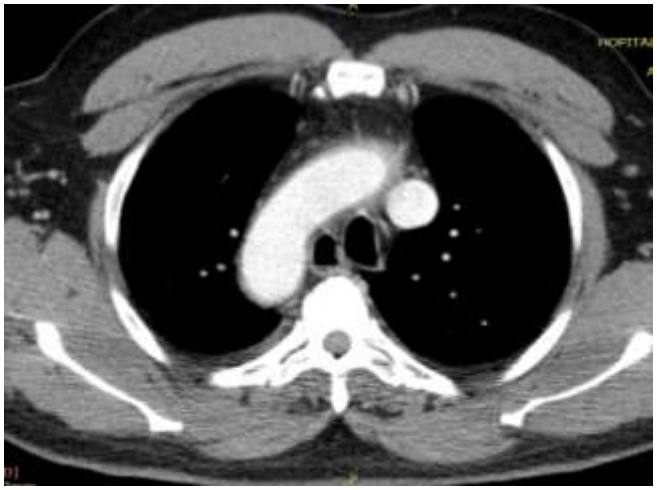

Figure 2: CT of the chest in axial section, mediastinal window, with injection of PDC, showing the arch of the aorta directed towards the right and the vena cava on the left.

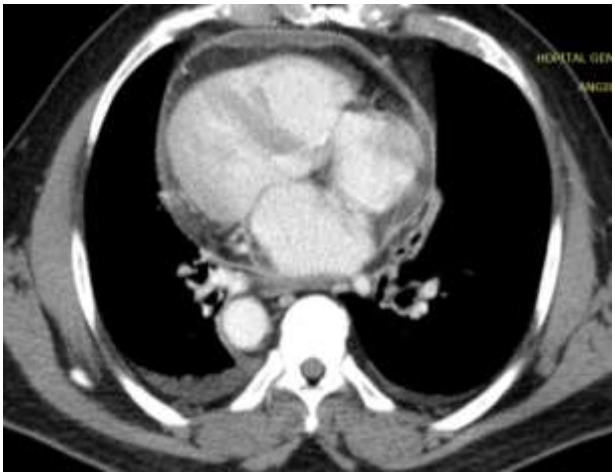

Figure 3: Chest CT in axial section, mediastinal window, with PDC injection, showing the apex of the heart on the right and the descending aorta on the right. NB: there is pericarditis.

It was also found two pulmonary lobes on the right, two pulmonary lobes on the left, a dilatation of the bronchi in the lower left lobe (figures 4, 5 and 6).

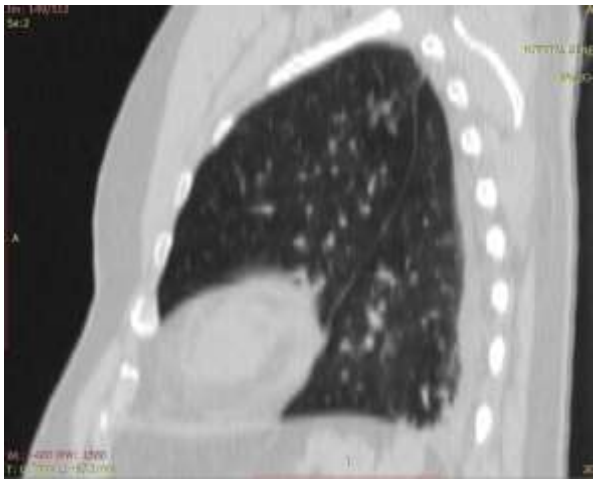

Figure 4: Chest CT in sagittal section, parenchymal window, showing 2 lung lobes on the right.

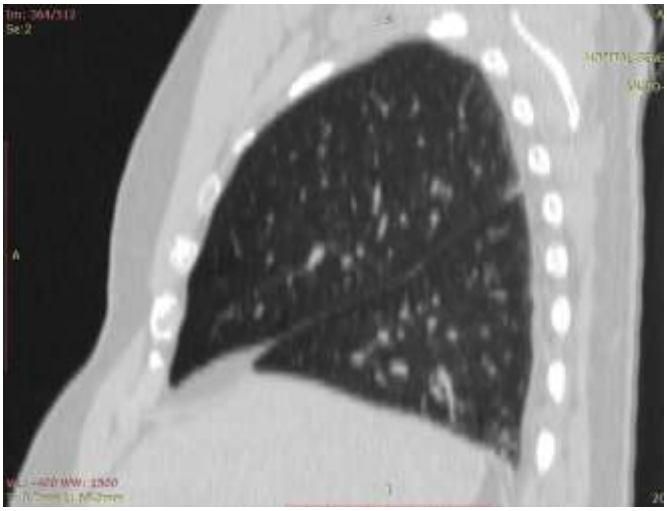

Figure 5: Chest CT in sagittal section, parenchymal window, showing 2 lung lobes on the left.

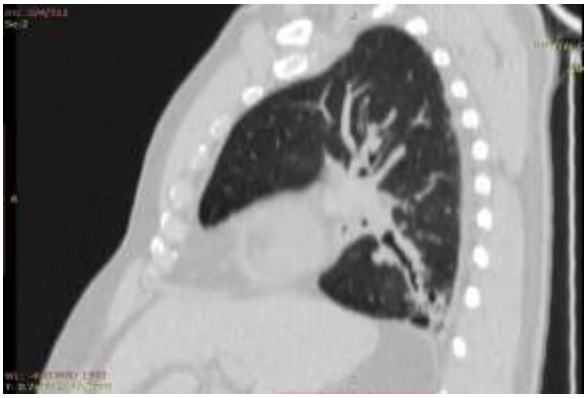

Figure 6: Chest CT in sagittal section, parenchymal window, showing bronchiectasis.

- At the level of the abdomen: the spleen and the stomach were in the right hypochondrium, the liver and the bile ducts

in the left hypochondrium, polysplenia, a complete common mesentery of the reversed type (colon on the right and hail on the left) (Figures 7-10).

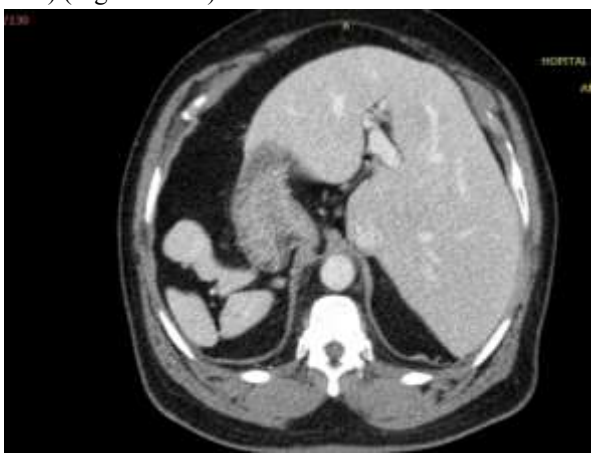

Figure 7: Abdominal CT in axial section at the height of the hypochondria, with injection of PDC, showing the liver in the left hypochondrium, the stomach and the spleens (three) in the right hypochondrium.

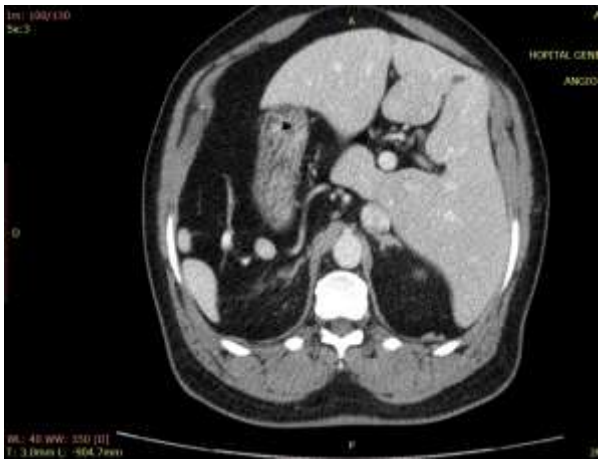

Figure 8: Abdominal CT in axial section at the height of the hypochondria, with injection of PDC, showing the liver in the left hypochondrium, the stomach and the spleens (three) in the right hypochondrium, IVC to the left of the aorta.

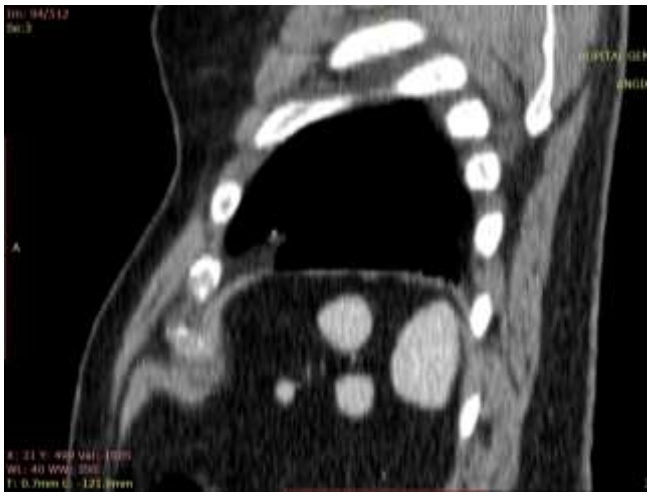

Figure 9: Thoraco-abdominal CT in sagittal section passing through the right hypochondrium showing polysplenia (four).

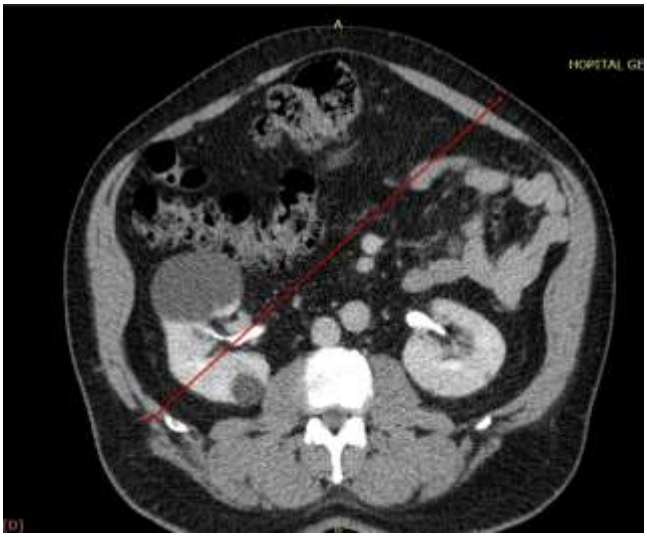

Figure 10: Abdominal CT in axial section, at the level of the kidneys, with injection of the contrast product showing the complete common mesentery of the inverted type with the colon on the right and the hail on the left.

For the CT scan of the sinuses, the constants used were 120KV and 40mA and the cuts made were 1mm contiguous. The examination was performed with the patient in the supine position, head first, arms along the body. A series of acquisitions without PDC injection were performed centered on the face. The scanner at this level showed chronic pan sinusitis (figures 11 and 12).

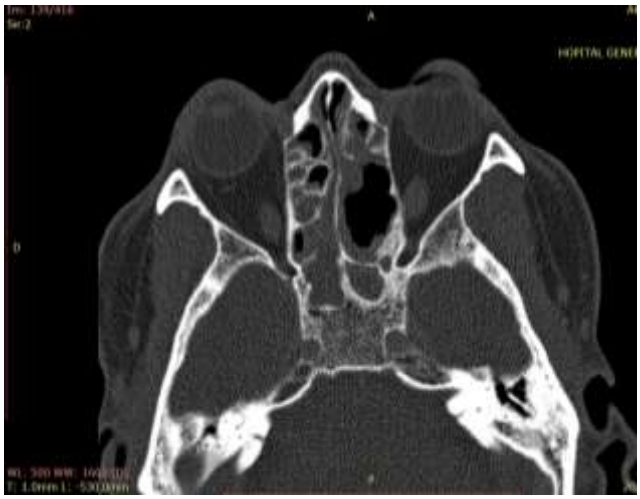

Figure 11: CT of the ethmoidal sinuses in axial section at the height of the orbits showing filling of the sphenoid sinuses, of certain ethmoidal cells with in-frame thickening of other cells.

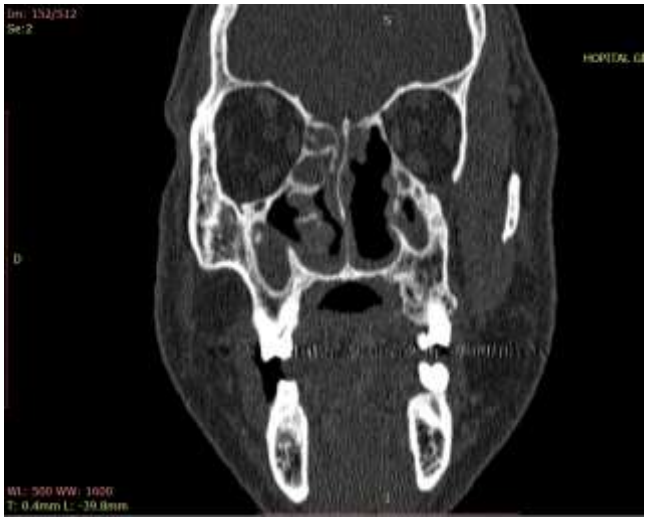

Figure 12: CT of the sinuses of the face in coronal section showing filling of the maxillary sinuses.

DISCUSSION

In 1904, Siewert [4] described a case of situs inversus coupled with bronchiectasis. However, the connection between these two signs could not be established. In 1935, Kartagener, the Swiss physician [5], was the first to identify a clinical syndrome associating chronic sinusitis and bronchiectasis as well as total situs inversus as well as familial cases. Thus, these signs have been grouped under the name "Kartagener's triad". Kartagener syndrome is a rare entity with an incidence of 1/32,000 births [4,5]. In half of the cases, Kartagener's syndrome is associated with primary ciliary dyskinesia [1,2] which is a heterogeneous genetic disease with autosomal recessive transmission, involving respiratory diseases linked to the ciliary constitutional anomaly [6]. Some authors [7] have nevertheless reported X-linked forms. Respiratory symptoms can begin early, with respiratory distress (50 to 70%), chronic bronchorrhea in childhood, and in adults, the picture is that of chronic obstructive pulmonary disease with diffuse bronchiectasis, or even a picture of chronic respiratory failure [4,7-8] as seen in our patient whose diagnosis was late following a chest X-ray. In Africa, several studies have been devoted to Kartagener syndrome but no case has been reported on an isomerism table. This is the first case described in Niger

Imaging (TAP CT and CT of the sinuses) confirms the classic triad of Kartagener's syndrome by highlighting the presence of bronchiectasis, sinusitis and a laterality defect. Radiologically, bronchiectasis appears early and is observed in 60 to 70% of cases at the time of diagnosis [9].

The Kartagener syndrome usually seen on situs inversus occurred in our case on an array of ambiguous situs, thus underlining the primary interest of our study. Situs ambiguous is a rare condition, if all lateralization defects are considered, the incidence is approximately 1/10,000 to 1/15,000 and women are more affected (F>M) [10] . The ambiguous situs is defined as being a defect of lateralization of the organs, several clinical pictures are possible and it can be associated with any type of malformations: cardiac, renal, digestive, etc... but we retain 2 main clinical pictures:

- Right isomerism: This would be a duplication of the right side; the thoraco-abdominal anatomical configuration of the right and left sides being identical to the image of the right side in relation to the axis of the body, we therefore have asplenia: duplication of the organs located on the right in the situs solitus and possible absence of the organs located on the left. Thus, we have: asplenia, central liver, inferior vena cava on the right and aorta on the left.
- Left isomerism: This would be a duplication of the anatomical configuration of the left side; the right and left sides being identical to the image on the left side, we therefore have polysplenia, VCI interruption with azygos/hemi-azygos continuation. [11]

Thus, in our observation we have 2 lungs of left configuration and polysplenia.

Situs ambiguus is in 50 to 100% of cases associated with a congenital heart defect [12]. If the patient does not show signs of heart disease incompatible with life, he may be asymptomatic, being diagnosed only incidentally in adulthood [13], often thanks to medical imaging examinations prescribed for other complaints. . In Africa, rare studies have focused on situs ambiguus [14].

The second interest of our observation is the association with a common mesentery. During embryological development, the digestive tract undergoes complex phenomena of reintegration, rotation and joining. When these phenomena are incomplete or flawed, they can lead to potentially pathological anatomical situations. In this case, migration anomalies of the mesentery: total absence of rotation, complete common mesentery, incomplete common mesentery and reverse rotation in case of situs inversus. Embryologically, the first rotation takes place before the 10th week of gestation when the primitive intestine is still located outside the abdomen. This rotation places the pre-yolk portion (hail) on the right and the post-yolk portion (colon) on the left; stopping at this stage is the origin of the complete common mesentery. The complete common mesentery then results from a stoppage of the intestinal rotation at 90°. Thus, we find the small intestine on the right and the colonic frame on the left; the cecum in an anterior and median position and the superior mesenteric artery to the right of the superior mesenteric vein [15]. In our observation, the prior ambiguous situs table explains why the hail is found on the left and the colon on the left.

CONCLUSION

Kartagener's syndrome remains a rare debilitating disease but which can be compatible with a normal life. It has most often been described in a situation of situs inversus but we retain that it is possible to find it in a situation of ambiguous situs. The association with certain rare malformations is also possible such as the common mesentery reported in our

observation. All this informs us about the importance of imaging examinations, especially scanners, in the diagnosis and follow-up of kartagener syndrome as well as associated malformations.

CONFLICT OF INTEREST

The authors declare no conflict of interest

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