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Kartagener's syndrome in adults: A case report

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Abstract

Kartagener's syndrome is a special case of primary ciliary dyskinesia (PCD).
It is characterized by a clinical triad of sinusitis, bronchiectasis, and situs inversus, which may be complete or incomplete. It is a rare congenital disorder with autosomal recessive inheritance, occurring in approximately one person in 32,000. We report the case of a 40-year-old female patient, admitted for preoperative work-up of nasosinusal polyposis.

Keywords: Kartagener syndrome, headache, imaging.

Introduction

Kartagener syndrome is a peculiar entity among primary ciliary dyskinesias (PCD) characterized by a clinical triad of sinusitis, bronchiectasis, and complete or incomplete situs inversus [1]. It is a rare congenital disorder with autosomal recessive inheritance and impaired mucociliary clearance, occurring in approximately one in 32,000 individuals [2]. Airway infections during the course of the disease classically manifest themselves in childhood [2]. A review of the African literature revealed studies published on this condition in North Africa [3]. We report the case of a 40-year-old female patient admitted for preoperative assessment of nasosinusal polyposis and recurrent bronchorrhea.

Observation

Patient G, aged 40, mother of 03 children, from a rural background, with a low socioeconomic level, followed for naso-sinusal polyposis with no particular family history. She reported chronic bronchorrhea and dyspnea for over 7 years. She presented for surgical management of her sinus pathology. Clinical examination on admission revealed a patient in good general condition, with bilateral snoring rales on pleuropulmonary auscultation. Cardiovascular examination was unremarkable. Kartagener's syndrome was suspected. A thoraco-abdomino-pelvic CT scan of the mediastinal windows confirmed dextrocardia with the tip of the heart on the right and bilateral cylindrical foci of bronchiectasis without mucoid impaction on the parenchymal windows. Abdominally, the liver is located in the left hypochondrium, while the stomach and spleen are in the right hypochondrium, confirming complete situs inversus.

Discussion

Kartagener's syndrome was first described by Manes Kartagener in 1935, the syndrome that bears his name and combines chronic sinusitis, situs inversus (complete or incomplete), and bronchiectasis [6]. Kartagener syndrome accounts for around 50% of primary ciliary dyskinesias (PCD). It is a rare genetic disorder with autosomal recessive inheritance; X-linked or dominant modes of inheritance have also been described [6]. It involves mutation of genes coding for dynein on chromosomes 5, 9, and 7, responsible for morphological and/or functional abnormalities of the cilia [6]. The age of diagnosis of the condition varies according to authors, with revelation of the disease as early as childhood [3, 4,]. In our case, the patient was 40 years old, similar to Amadou Doumbia et al [1] who reported a case in a 35-year-old adult.

We noted no gender predominance in the literature, and our patient had no family history of consanguinity or respiratory disease. Imaging plays a vital role in the diagnosis of the pathology. In our case, the thoracoabdominal CT scan (Figure 1) showed a complete situs inversus, i.e. dextrocardia, left location of the liver and gallbladder, and right location of the spleen and stomach.

In the lung parenchymal window, CT scans showed discrete cylindrical bronchiectasis of the middle lobe, lingula, and lower lobes (Figures 2 and 3). Bronchial involvement is most common in the middle and lower lobes, with bronchiectasis increasing with age [1]. The symptomatology of Kartagener syndrome is dominated by respiratory signs beginning in childhood. These signs are present in all patients and are

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specific only for their chronicity and recurrence [1]. They include chronic bronchial congestion, a daily hacking cough with mucopurulent secretions, and phases of exacerbations and superinfections. In our case, the patient presented with obstructive bronchopneumonia with diffuse bronchiectasis, as described in the literature [6], and a chronic phlegmy cough with no evidence of dyspnoea. The evolution of the respiratory pathology is variable, and depends in particular on how early the diagnosis is made and how rigorously the patient is managed [2]. Involvement of the upper airways is marked by sinusitis and otitis associated with abnormal mucociliary clearance. In our case, the patient presented with repeated episodes of nasal obstruction and chronic recurrent rhinorrhea evolving since childhood. A CT scan of the facial sinuses revealed near-total filling of the left compartment of the sphenoidal sinus, with framelike thickening of its right compartment. Filling of the anterior and posterior ethmoidal cells was more marked on the left. Polypoid filling of the nasal cavity associated with turbinate hypertrophy more marked in the inferior turbinates, all suggestive of nasosinus polyposis (Figure 4 and 5). Nasosinus polyposis is present in around 30% of patients.

Treatment is purely symptomatic, relying mainly on respiratory physiotherapy. Antibiotic therapy is indicated for pulmonary or ENT respiratory exacerbations, and in certain cases of chronic colonization. The patient benefited from antibiotic treatment (amoxicillin-clavulanic acid combined with moxifloxacin). In terms of ENT, patients should be educated to blow their nose effectively, while insisting on nasal hygiene with saline lavage.

Conclusion

Kartagener's syndrome remains a rare and disabling disease but can be compatible with a normal life if treated early. However, in forms with significant pulmonary lesions, the patient's prognosis is at risk in the short term, due to severe multi-visceral damage.

Figure :

Figure 1: Thoracic and abdominal CT scan, coronal section in mediastinal window, visualizing a heart oriented to the right, highlighting the stomach on the right, and the liver located in the left hypochondrium.

Figure 2 and 3: Chest CT scan, axial and coronal section in parenchymal window, showing bilateral bronchiectasis without mucoid impactions.

Figure 4 and 5: CT scan of the sinuses showing near-total filling of the left compartment of the sphenoidal sinus, with framed thickening of its right compartment.

Filling of the anterior and posterior ethmoidal cells, more marked on the left.

Polypoid filling of the nasal cavity, with turbinate hypertrophy more marked in the inferior turbinates.

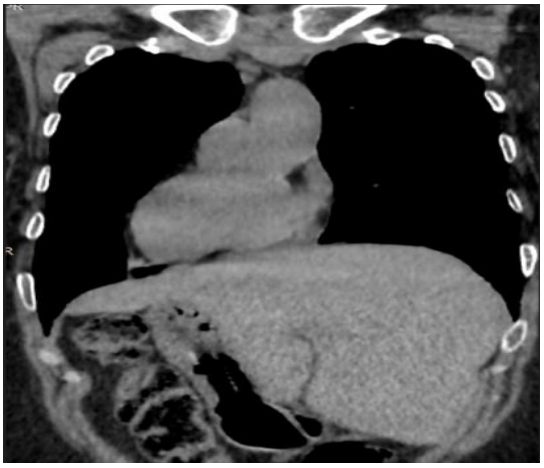
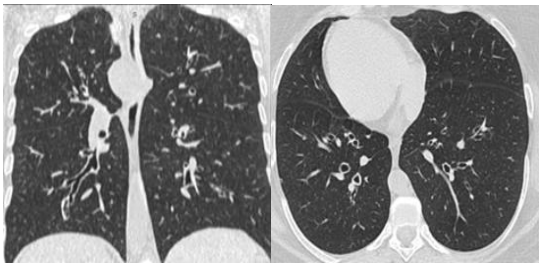
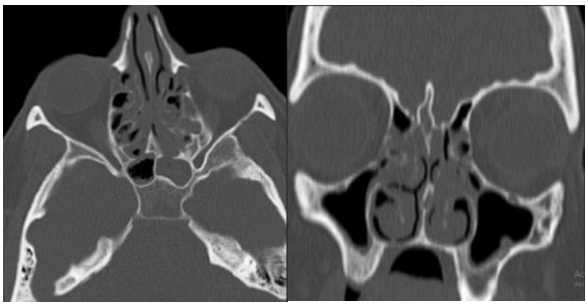


Figure 1: Thoracic and abdominal CT scan: Dextrocardia + Liver located in the left hypochondrium and stomach to the right.



Figures 2 et 3: thoracic CT scan with parenchymal window in coronal and axial section: cylindrical bronchiectasis in the middle, lingular and lower lobes on both sides.



Figures 4 et 5: Axial and coronal CT scan of the sinuses in the bone window, noting: near-total filling of the left compartment of the sphenoidal sinus, with framelike thickening of its right compartment.

Filling of the anterior and posterior ethmoidal cells, more marked on the left.

Polypoid filling of the nasal cavity, with turbinate hypertrophy more marked in the inferior turbinates.

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