Bronchiectasis in primary ciliary dyskinesia: Radiological keys for its diagnosis

Published on 19.05.2018

DOI: 10.1594/EURORAD/CASE.15717
ISSN: 1563-4086
Section: Chest imaging
Area of Interest: Lung
Procedure: Diagnostic procedure
Imaging Technique: CT
Imaging Technique: Conventional radiography
Special Focus: Genetic defects Case Type: Clinical Cases
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Patient: 36 years, female

Clinical History:

A 36-year-old woman consulted for greenish bronchorrhea and moderate effort dyspnea. The patient suffered from pneumonia when she was a child and she had suffered from repetitive respiratory infections since then. She had family members diagnosed with bronchiectasis, so a chest X-ray and a chest Multidetector Computed Tomography (CT) were performed.

Imaging Findings:

The posteroanterior and lateral chest X-ray revealed parallel and ringlike opacities consistent with multiple bronchiectasis with bilateral and diffuse distribution (black arrows in Fig. 1). Chest CT showed bronchiectasis with universal distribution, they were predominantly cylindrical (blue arrows in Fig. 2a) and varicose (white arrowheads in Fig. 2b) in the upper lobes, cystic in the middle lobe (anterior red arrow in fig. 2c), and varicose and cystic in lower lobes where they were more numerous (red arrows in Fig. 2c, 2d and 3). Some of the bronchiectasis were occupied. In addition, there was a bilateral mosaic attenuation with patchy areas of lower density suggestive of air trapping areas (yellow asterisks in Fig. 2a, 2b and 3), due to the coexisting constrictive bronchiolitis in patients with bronchiectasis.

Discussion:

Primary ciliary dyskinesia is a genetic disease with autosomal recessive inheritance, in which there is an abnormality of the dynein arms of epithelial cilia that leads to an alteration in mucociliary clearance predisposing to pulmonary infections, airway damage, bronchiectasis, sinusitis and otitis media [1, 2]. It is a disease that is diagnosed relatively late because of its nonspecific symptoms in children [2].

Although in the chest X-ray we can see signs of bronchiectasis such as "tram-track" appearance of parallel and
ringlike opacities, the thoracic CT is the reference standard because it shows the bronchiectasis distribution and additional findings that help us make an aetiological diagnosis [1, 3]. The typical imaging findings of primary ciliary dyskinesia in thoracic CT are bilateral and diffuse bronchiectasis with variable severity, which predominate above all in the lower lobes, as well as in the middle lobe and lingula. Other frequent findings are tree-in-bud nodules related to acute infection, mucous plugging due to alteration in mucous clearance, and the mosaic attenuation secondary to areas of air trapping because of the coexisting constrictive bronchiolitis [1, 2].

In the differential diagnosis, we must take into account those entities that present with bronchiectasis predominantly in the lower lung fields: chronic aspiration of gastric contents in the context of patients with gastroesophageal reflux disease or alterations in oesophageal motility; immunodeficiencies (especially hypogammaglobulinaemia) that are usually diagnosed in childhood and usually present other associated symptoms such as intestinal infections; and the alpha-1-Antitrypsin deficiency that typically causes panlobular emphysema and sometimes has bronchiectasis but rarely they appear before emphysema [1, 2]. Other diseases to consider in this case are: cystic fibrosis, in which upper lobes predominance of bronchiectasis is seen in many but not in all cases, being also a diffuse distribution a common finding; and allergic bronchopulmonary aspergillosis, which usually manifests with cystic or varicoid bronchiectasis with central or proximal upper lobe predominance [1].

The definitive diagnosis of primary ciliary dyskinesia can be made by measuring mucociliary clearance through different studies such as the saccharin test or a test with radioisotopic tracers, exhaled nitric oxide can be measured, or a biopsy can be done to study ciliary structure under an electron microscope. In our case, the patient underwent a dynamic scintigraphic study, with instillation of the radioisotopic tracer in the nasal meatus, demonstrating that there was no movement of the tracer, which was compatible with mucociliary akinesia [4, 5].

**Differential Diagnosis List:** Bronchiectasis due to primary ciliary dyskinesia, Primary ciliary dyskinesia, Chronic aspiration of gastric contents, Immunodeficiencies (hypogammaglobulinaemia), Alpha-1-Antitrypsin deficiency, Cystic fibrosis, Allergic bronchopulmonary aspergillosis

**Final Diagnosis:** Bronchiectasis due to primary ciliary dyskinesia

**References:**


**Description:** Cylindrical (blue arrows in Fig. 2a) bronchiectasis in the upper lobes. Bilateral mosaic attenuation with patchy areas of air trapping areas (yellow asterisks in 2a and 2b). **Origin:** Department of Radiology, H.G.U. Morales Meseguer, Murcia, Spain

**Description:** Varicose bronchiectasis (white arrowheads in Fig. 2b) in the upper lobes. Bilateral mosaic attenuation with patchy areas of air trapping areas (yellow asterisks in 2a and 2b). **Origin:** Department of Radiology, H.G.U. Morales Meseguer, Murcia, Spain
Description: Cystic bronchiectasis in the middle lobe (anterior red arrow in Fig. 2c), and varicose and cystic in lower lobes where they were more numerous (red arrows in Fig. 2c and 2d). 

Origin: Department of Radiology, H.G.U. Morales Meseguer, Murcia, Spain

Description: Varicose and cystic bronchiectasis in lower lobes where they were more numerous (red arrows in Fig. 2c and 2d). 

Origin: Department of Radiology, H.G.U. Morales Meseguer, Murcia, Spain
Description: Bronchiectasis predominates in the lower lung fields (red arrows) and there is a bilateral mosaic pattern secondary to areas of air trapping (yellow asterisks). Origin: Department of Radiology, H.G.U. Morales Meseguer, Murcia, Spain
Description: Parallel and ringlike opacities (black arrows) consistent with multiple bronchiectasis with bilateral and diffuse distribution. Origin: Department of Radiology, H.G.U. Morales Meseguer, Murcia, Spain
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