

Clinical image

Williams-Campbell Syndrome: A Rare Case of Bronchiectasis

Síndrome de Williams-Campbell, un caso raro de bronquiectasias

Alexander Ali Jiménez Romero^{a,*}, Olga Meca Birlanga^a, Begoña Inoriza^b, Francisca Lerenas Bernal^a,
Maria del Rosario Cabello Jabalquinto^a

^a Department of Pneumology, Hospital General Universitario Santa Lucía, C. Minarete, s/n, 30202 Cartagena, Spain

^b Department of Thoracic Radiology, Hospital General Universitario Santa Lucía, C. Minarete, s/n, 30202 Cartagena, Spain

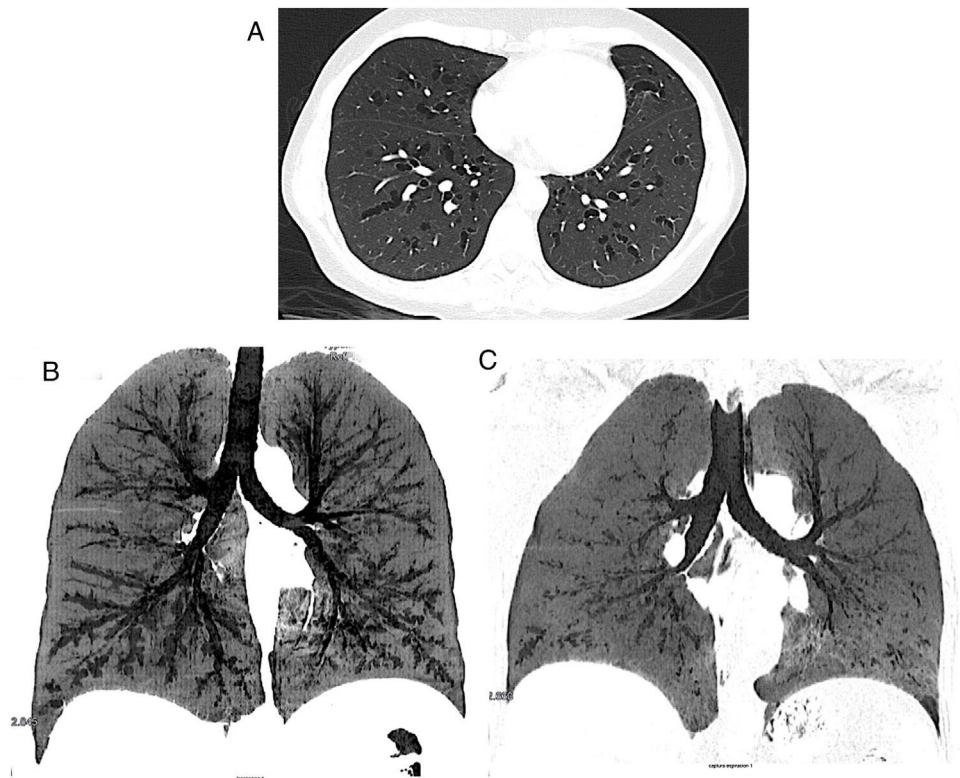


Fig. 1. (a) Shows the central bronchiectasis and how they change during the inspiration (b) and expiration (c) chest CT.

A 57-year-old female, with repetitive episodes of cough, upper airway symptoms and moderate exertion dyspnea, showed multiple central bilateral bronchiectasis in bronchus from 3rd to

8th generation sparing periphery on a chest computed tomography (CT) as part of her cough tests (Fig. 1a), respiratory functional tests were normal, but after analyzing the CT images, we decided to perform a low-dose CT during full expiration (Fig. 1b, c), this showed an approximately 80% collapse of the previously ecstatic bronchi, this was consistent with cartilage deficit and with Williams-Campbell syndrome.

* Corresponding author.
E-mail address: Alexandermed2012@gmail.com (A.A. Jiménez Romero).

Williams-Campbell syndrome is a rare syndrome consisting of a total or partial deficit of the subsegmental bronchi cartilage, usually affecting between the 4th and 6th generation, this results in bronchiectasis in the affected bronchi. This syndrome is generally diagnosed in childhood due to its congenital nature¹; however other authors speak about the possibility of secondary acquisition.²

The diagnosis of this syndrome usually requires bronchoscopy, in our case expiration/inspiration CT was enough for diagnosis. Taking into account CT its non-invasive methodology, easy of execution and good patient tolerance, and considering lung biopsy has several complications, the expiratory/inspiratory chest CT procedure should be included in patients with central diffuse cylindrical bronchiectasis.¹

Informed consent

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

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Authors' contribution

All authors have contributed significantly to the research and preparation, revision and final production of the manuscript and approve its submission.

Conflict of interest

The authors declare to have no conflict of interest directly or indirectly related to the manuscript contents.

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