What is the cause of this patient's chronic productive cough?

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A 70-year-old male HIV-seronegative smoker presented with a productive cough and dyspnoea. Clinical findings included digital clubbing, central cyanosis and features of cor pulmonale. Bilateral crackles were audible on auscultation of his chest.

A high-resolution computerised tomography scan of his chest revealed features in keeping with Mounier-Kuhn syndrome (MKS) (congenital tracheobronchomegaly) and bilateral cystic bronchiectasis (Fig. 1). The transverse diameter of his trachea measured 32 mm (normal for males 13 - 25 mm).[1] MKS is characterised histologically by atrophy of smooth muscle and elastic tissue in the tracheal and bronchial walls. It is more common in males than females and is usually diagnosed in the 3rd or 4th decade of life,[2] although there is a report of a male diagnosed at 86 years of age.[3] Abnormal dilatation of the tracheobronchial tree, often with diverticula, causes impaired function of the mucociliary escalator, recurrent respiratory tract infections and bronchiectasis.[2] The vast majority of cases are sporadic.

1. Boiselle PM. Imaging of the large airways. Clin Chest Med 2008;29:181-193.
2. Simon M, Vremaroiu P, Andrei F. Mounier-Kuhn syndrome. J Bronchol Intervent Pulmonol 2014;21:145-149.
3. Geppert EF. Recurrent pneumonia. Chest 1990;98:739-745.

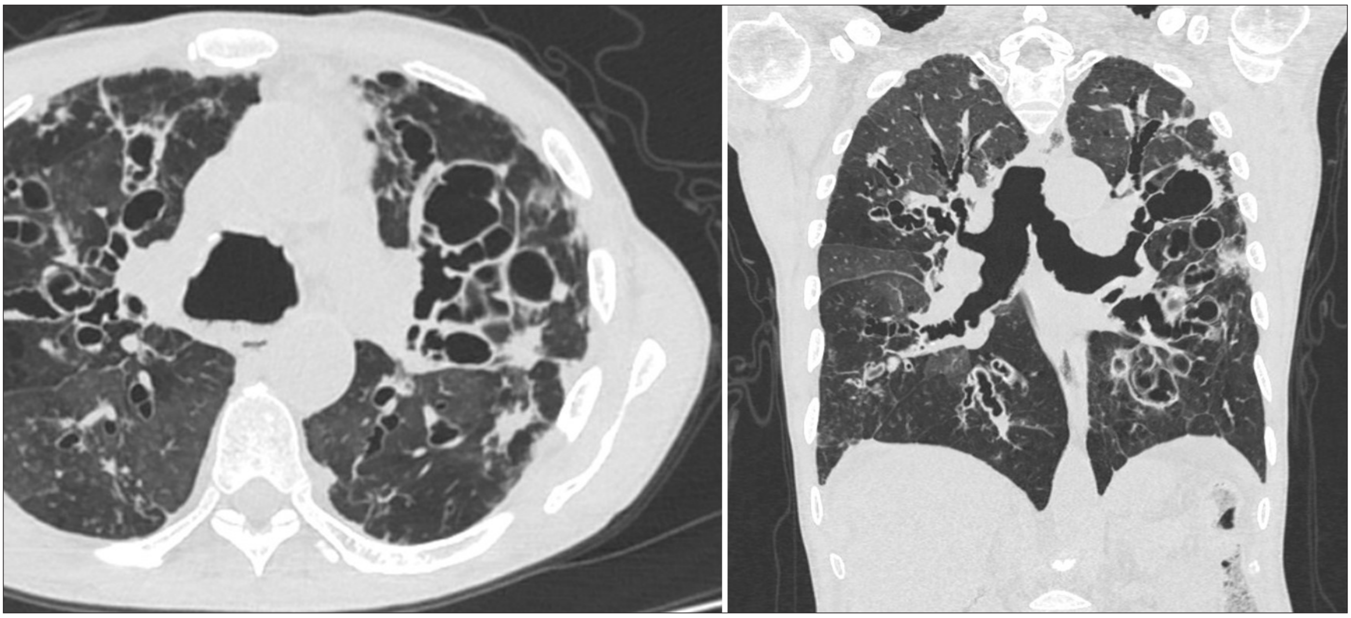

Fig. 1. Axial and coronal views of high-resolution computerised tomography scan demonstrating tracheobronchomegaly and bilateral cystic bronchiectasis.