

MOUNIER-KUHN SYNDROME

E. Schiettecatte¹, E. Van Hedent¹, R. Van Herreweghe², K. Verstraete³

Key-word: Bronchi, abnormalities

Background: A 38-year-old male with no medical history complains about hemoptysis, chest pain, dyspnea on exertion and a cough. He has a history of occasional smoking, but had quit smoking 1 year previously. There is a normal saturation and normal spirometry. Clinical examination and auscultation are also normal.

Because of these complaints CT scan of the thorax was performed. For further diagnostic work-up bronchoscopy was performed afterwards.

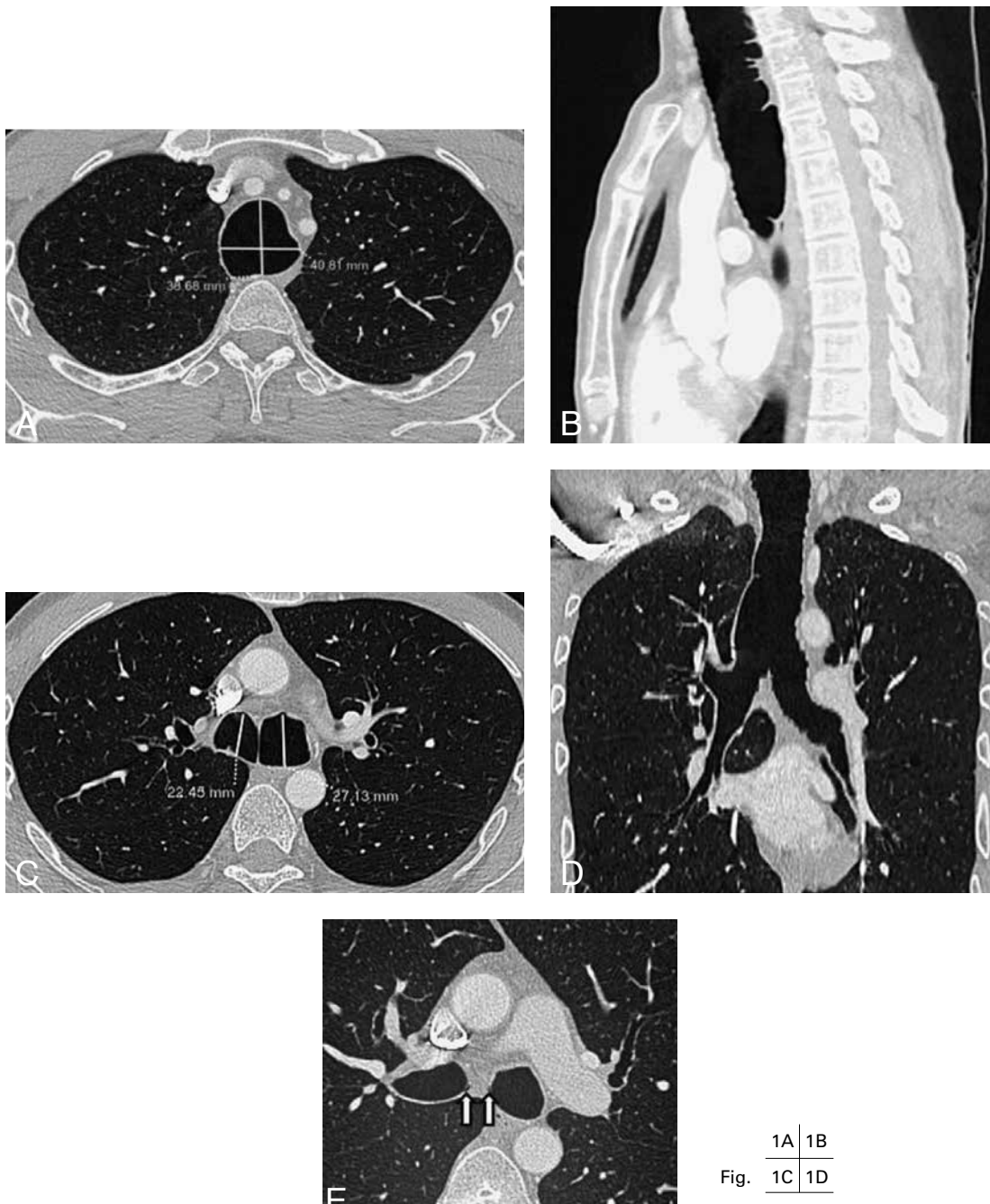


Fig.

1A	1B
1C	1D
1E	

Work-up

CT scan of the thorax (Fig. 1) shows on axial image of the chest at the level of the trachea, lung window setting (A) dilated aspect of the trachea with antero-posterior diameter of 38,68 mm and latero-lateral diameter of 40,81 mm. Reformatted image of the trachea in the sagittal plane, lung window setting (B) reveals dilated aspect of the trachea with irregular, undulated appearance of the tracheal wall due to protrusion of the musculomembranous tissue between the cartilaginous rings.

Axial image of the chest at the level of the main bronchi, lung window setting (C) visualizes dilated aspect of the main bronchi with diameter of 22,45 mm (right) and 27,13 mm (left). On reformatted image of the chest at the level of the trachea and bronchi, lung window setting (D), dilated aspect of the trachea and main bronchi is seen. Axial image of the chest at the level of the main bronchi, lung window setting (E) visualizes dilated aspect of the main bronchi with small diverticula (arrows).

Radiological diagnosis

Based on the CT findings and the clinical appearance the diagnosis of *Mounier-Kuhn syndrome* (tracheobronchomegaly) was established.

Discussion

Mounier-Kuhn syndrome (MKS) is characterized by tracheobronchial dilatation and was first recognized by Czyhlarz in 1897. In 1932 Mounier-Kuhn described the endoscopic and radiographic appearance of the enlarged airways. MKS is more common in men than in women and is typically diagnosed in the 3th or 4th decades of life. The cause is not fully known. It is linked to familial susceptibility and presumably inherited by autosomal recessive mechanism, but is most often sporadic and can also occur secondary as a complication of diffuse pulmonary fibrosis. There is an association with cutis laxa and Ehlers-Danlos syndrome. The etiology of MKS is uncertain, but supports the concept of a congenital defect or severe atrophy of the longitudinal elastic fibers with thinning of the muscularis mucosa. This results in dilatation of the cartilaginous and membranous portions of the trachea and main bronchi. Diverticula occur by protrusion of the

musculomembranous tissue between the cartilaginous rings.

The symptoms of MKS are non-specific and resemble those of bronchitis and bronchiectasis and many patients have few or no symptoms. Possible symptoms are: a harsh cough, copious or purulent sputum production, progressive dyspnea, recurrent respiratory tract infections, spontaneous pneumothorax, hemoptysis and finger clubbing.

On plain chest radiology the dilatation of the trachea and bronchi may be visible and may adequately display the tracheobronchomegaly, however CT scan is usually required for accurate airway measurement and assessment of complications (diverticulosis, scarring and bronchiectasis). CT characteristics are dilatation of the central airways, diverticulosis and an irregular, corrugated appearance of the trachea due to protrusion of the musculomembranous tissue between the cartilaginous rings.

Diagnostic criteria for tracheobronchomegaly for males are: tracheal diameters surpassing 25 mm in laterolateral direction, 27 mm in anteroposterior direction, diameter of the left main bronchus > 18 mm and of the right main bronchus > 21 mm.

Diagnostic criteria for tracheobronchomegaly for females are: tracheal laterolateral diameter > 21 mm, anteroposterior diameter > 23 mm, left main bronchial diameter > 17,4 mm and right main bronchial diameter > 19,8 mm.

On bronchoscopy, the increased caliber of the trachea and bronchi is evident and semicircular folds of mucous membrane with saccular pouches and pooled secretions may be seen.

Treatment is mainly supportive with no specific therapy for asymptomatic patients.

Bibliography

1. Burcin C., Salhih B., Canan Y.: Mounier-Kuhn Syndrome: A rare cause of bronchial dilatation. *Tex Heart Inst J*, 2011, 38: 194-196.
2. Falconer M., Collins D.R., Feeney J., et al.: Mounier-Kuhn syndrome in an older patient. *Age and Ageing*, 2008, 37: 115-116.
3. Schwarts M., Rossoff L.: Tracheobronchomegaly. *Chest*, 1994, 106: 1589-1590.
4. Van Schoor J., Joos G., Pauwels R.: Tracheobronchomegaly – the Mounier-Kuhn syndrome: report of two cases and review of the literature. *Eur Respir J*, 1991, 4: 1303-1306.