

ACUTE RESPIRATORY SYMPTOMS IN MOUNIER-KUHN SYNDROME

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Key-word: Trachea, abnormalities

Background: A 73-year-old female patient presented at the emergency room with cough and dyspnea. There was green sputum production and she had fever. She had given up smoking 15 years before. Her medical history was positive for recurrent bronchitis. Physical examination revealed rhonchi, a body temperature of 38.2°C and tachypnea of 35 breaths per minute. Oxygen saturation was 80% without oxygen supply. Chest radiography did not reveal a consolidation.

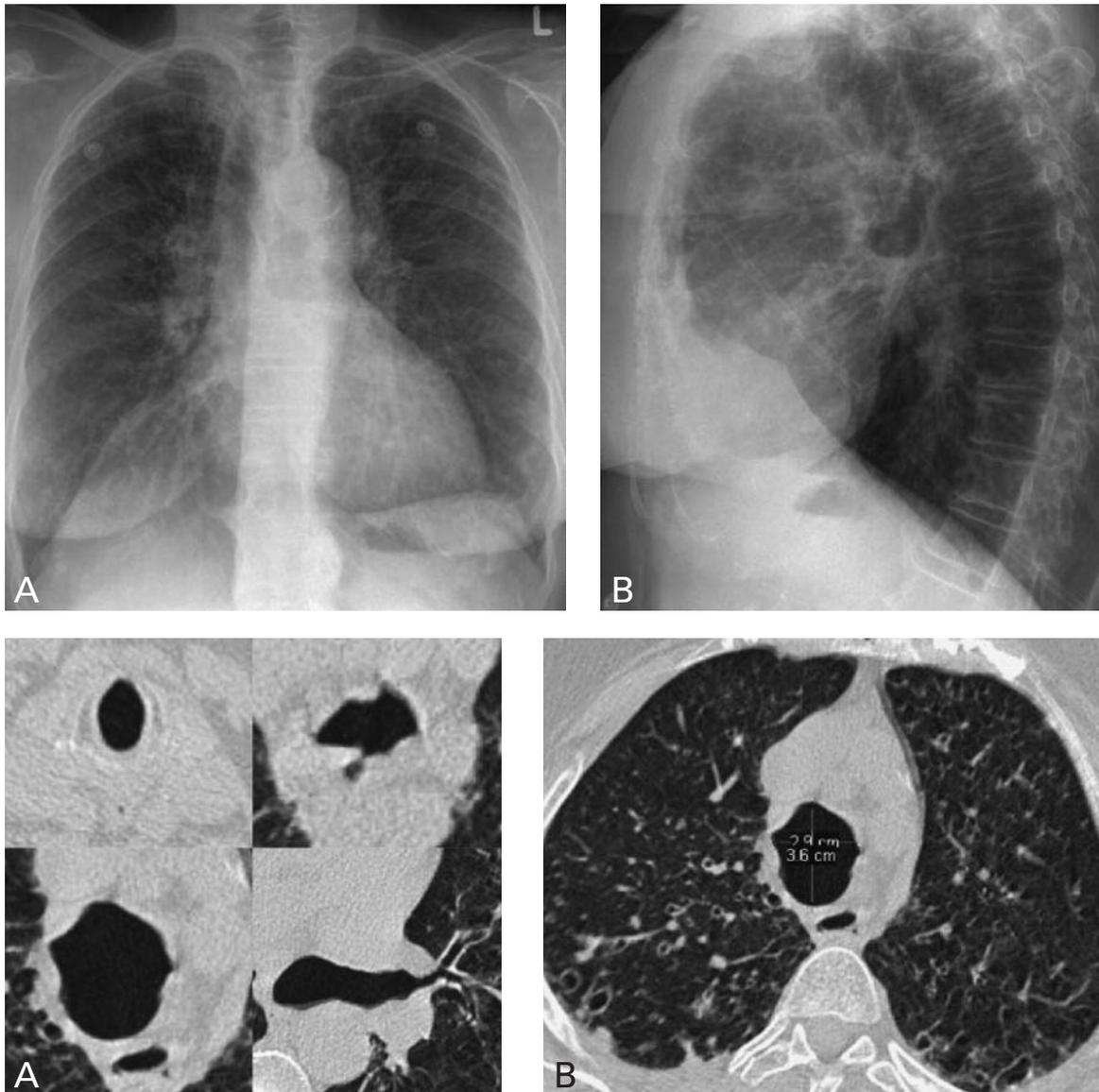


Fig.

1A	1B
2A	2B

Work-up

Chest radiograph (Fig. 1) demonstrates on the posterior-anterior (PA) view (A) a large transverse dimension of the trachea (3.2 cm) and left main bronchus (2.4 cm), large lung volumes, irregular diaphragmatic contours. Lateral chest radiograph (B) demonstrates the large trachea (3.7 cm) to a better extent.

Thin-section CT scan of the lungs (Fig. 2) includes four detail views of axial CT images (A) showing the wide trachea and left main bronchus (PA: 2.5 cm) with an abrupt change to more normal dimensions. In addition, there are tracheal diverticulae. Axial thin CT-section (B) demonstrates the large diameters of the trachea (transverse: 3.1 cm, PA: 3.6 cm) and the presence of bronchiectasis.

Radiological diagnosis

The radiological diagnosis was made of *syndrome of Mounier-Kuhn*, with tracheobronchomegaly, tracheal diverticulae and bronchiectasis.

Discussion

Tracheobronchomegaly is a rare disorder. It appears to impair mucous clearing and therefore leads to recurrent infections. The best diagnostic clue is a tracheal dilatation. The lateral chest radiograph is usually the most conspicuous. Normal values of the trachea have been published for chest radiography and computed tomography.

On a chest radiograph, normal antero-posterior tracheal diameters (profile views) (mean \pm standard deviation) are approximately 17 ± 2 mm and 21 ± 3 mm for females and males respectively. Diameters of the trachea in the coronal plane (mean \pm SD) are 17 ± 2 mm and 20 ± 2 mm for females and males respectively.

The other imaging findings of tracheobronchomegaly or Mounier-Kuhn disease include

dilatation of the main bronchi, usually with an abrupt change to normal caliber, diverticulae of the trachea and bronchiectasis.

On expiration, a collapse of the trachea can be seen.

Mounier-Kuhn syndrome appears to be autosomal recessive in some families and in some cases the disease is associated with the disease of Ehlers-Danlos or Marfan. There is also a congenital variant of the disease. Overall, most cases are sporadic. The disease is usually diagnosed at a younger age than in our patient, between 30-50 years. A strong male predominance has been observed.

In the presented case, the clinical symptoms and the imaging findings were very characteristic for the Mounier-Kuhn Syndrome, with exception for age at presentation and sex.

The diagnosis was made based on the CT scan, although, in retrospect, the chest radiograph also demonstrated the dilatation of the trachea and main bronchi as well. At presentation, the patient was treated as if having a pneumonia based on the clinical findings in the emergency room. The case illustrates the importance of looking carefully at the trachea and main bronchi on both the PA and lateral chest radiograph.

Bibliography

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