

Mounier-Kuhn syndrome — a case study

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Clinical information

A 21-year-old male presented to the casualty department complaining of swelling of both feet for 2 weeks and persistent headache. On further inquiry he also reported a 3-year history of coughing and a course of treatment for tuberculosis during the 6 months immediately preceding this presentation.

On examination the patient was found to be dyspnoeic and orthopnoeic with a productive cough and abdominal distension. Anaemia, peripheral oedema and digital clubbing were also noted as well as cyanosis which improved on oxygen. Urinalysis revealed no abnormality. He was afebrile (36°C), awake and fully oriented.

Respiratory system

A tachypnoea of 26/min was noted. Bilateral basal crepitations were found and an area of bronchial breathing, dull to percussion, in the left base. No respiratory distress was present.

Cardiovascular system

The patient's blood pressure was

113/73 mmHg and pulse 120 bpm. A hyperdynamic circulation was found with a loud S1, right ventricular S3 gallop and loud P2. Suspected bruits of mitral and tricuspid valve incompetence were noted. Jugular venous pressure was raised. The apex of the heart was displaced laterally into the 6th intercostal space.

Abdomen

Examination revealed ascites and liver enlargement of 2 cm. No rebound tenderness was present. A vasculitic skin rash could be seen over most of the torso and legs.

Radiological examination

Chest X-ray

A grossly dilated appearance of the trachea and main bronchi was observed. The right lung field showed an interstitial infiltrate including Kerley B lines, and also confluent shadowing in the perihilar areas and upper lobe. Bronchiectatic changes could be seen in the right lower zone. Right apical pleural thickening was present (Fig. 1).

On the left, lower lobe bronchiectasis and volume loss were noted with compensatory hyperinflation of the upper lobe. Blunting of the left costophrenic angle was present with some Kerley B lines. Enlargement of the cardiac shadow was also present

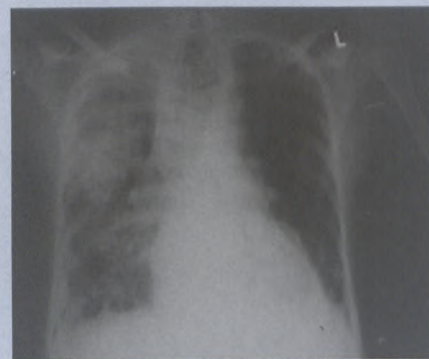


Fig. 1. The grossly dilated trachea and main bronchi are clearly seen with bronchiectatic changes in the right lower zone. The heart is also enlarged.

with prominence of upper-lobe vasculature.

CT chest — high resolution and mediastinal cuts

Tracheobronchial ectasia was again observed. The trachea measured 4.13 x 4.65 cm in diameter, while mainstem bronchial dilatation of 2.19 cm and 2.00 cm were noted on the right and left side respectively (Fig. 2). Thymic hyperplasia and right jugular prominence could also be seen. The rest of the mediastinum and both hilar regions were felt to be normal.

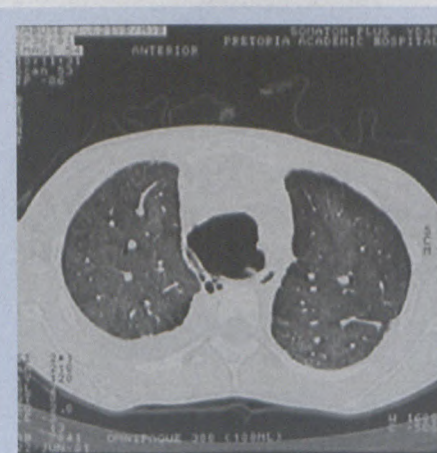


Fig. 2. Marked ectasia of the trachea is clearly seen. Ground-glass infiltrates suggest underlying infection.

A pleural effusion had formed on the right. Volume loss on the left resulted in ipsilateral cardiac shift. Both upper lobes and the middle lobe showed ground-glass infiltrates. Bronchiectasis was confirmed bilaterally in the peri-hilar regions and bases. Advanced fibrotic changes were seen in the left lower lobe, causing 'shagginess' of the pleuro-pericardial surfaces.

A confident diagnosis of tracheobronchomegaly (Mounier-Kuhn syndrome) was made. Complications of acute and chronic infection included visible pneumonitis, bronchiectasis and fibrosis. Thymic hyperplasia was likely reactive, due to repeated infections.

Cardiac ultrasound examination

A massive right ventricle was found with enlargement of the right atrium. Mild tricuspid incompetence was demonstrated with an increased pressure gradient. A pulmonary pressure of 70 mmHg was measured, indicating severe pulmonary hypertension.

Final diagnosis

1. Mounier-Kuhn syndrome (tracheobronchomegaly), complicated by: (i) pneumonia; (ii) bronchiectasis and fibrosis; and (iii) cor pulmonale with tricuspid incompetence.

2. Hyperplasia of the thymus due to chronic infection.

The patient was treated with Lasix, Berotec inhalations, Ciprobay and physiotherapy.

Discussion

Tracheobronchomegaly was originally recognised at autopsy by Czyhlaz in 1897, but the first clinical descrip-

tion by Mounier-Kuhn was given in 1932.^{1,2} Mounier-Kuhn syndrome primarily affects the supporting structures of the trachea and major bronchi, weakening them and leading to ectasia. Bronchi abruptly revert to normal by the 4th to 5th divisions.³

The syndrome is characterised by marked ectasia of the trachea and main bronchi, bronchiectasis and repeated lower respiratory tract infections.⁴ Presentation is typically during the 3rd or 4th decade, with a preponderance in males of African-American origin.^{2,4} A familial tendency is observed with an autosomal recessive inheritance pattern.²

Tracheobronchomegaly is the underlying cause in virtually all observed instances of tracheal dilatation. It is regarded as a primary dysplasia rather than a complication of recurrent infection; this is in keeping with the acute transition to normal calibre bronchi.^{2,5} Symptoms include recurrent respiratory infections, chronic productive cough and exertional dyspnoea.⁴ Weakness of proximal airway walls results in flaccidity and collapse, ineffective cough, retention of mucus and consequently,

repeated infections.¹

As the cartilagenous rings of the trachea and larger bronchi dilate, the intervening soft tissues bulge outward, forming diverticular, extraluminal protrusions of the air column. These lesions are best seen on the lateral chest view. In severe cases, associated tracheobronchomalacia leads to proximal airway collapse during expiration.

The increase in tracheal diameter can be pronounced as in this case or subtle enough to be easily overlooked.^{4,5} Widening of the trachea beyond 30 mm or mainstem bronchial diameters over 20 mm on the right or 18 mm on the left indicate tracheobronchomegaly.⁴ Peri-hilar emphysema and bullae can also be found (Fig. 3).⁴

Secondary tracheobronchomegaly has been described in patients with Ehlers-Danlos syndrome, Marfan syndrome, Kenny-Caffey syndrome, Brachmann-de Lange syndrome, connective tissue diseases, ankylosing spondylitis, long-chain deposition disease and following prolonged ventilation in premature infants. Other sources give a shorter differential for

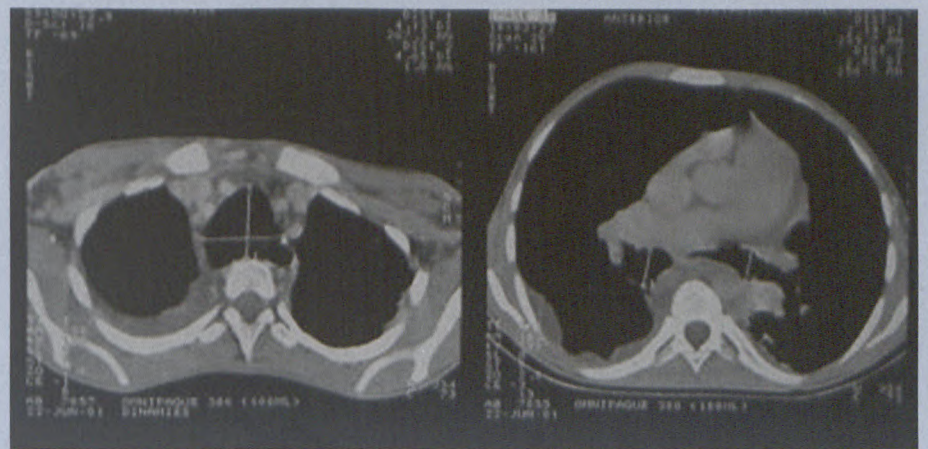


Fig. 3. The trachea measured 4.13 x 4.65 cm in diameter while mainstem bronchial dilatation of 2.19 cm and 2.00 cm were noted on the right and left side respectively.

tracheobronchomegaly, consisting of Mounier-Kuhn syndrome, Ehlers-Danlos syndrome, cutis laxa and relapsing polychondritis.^{4,5}

Tracheal stenting has proved of great benefit in patients with advanced stages of Mounier-Kuhn disease (Fig. 4).⁴

Summary

Measurement of the dilated trachea on a chest X-ray can make the diagnosis of tracheobronchomegaly. Normal tracheal diameter on a postero-anterior radiograph is 20 mm, with a standard deviation of 2 mm. In cases of tracheobronchomegaly the

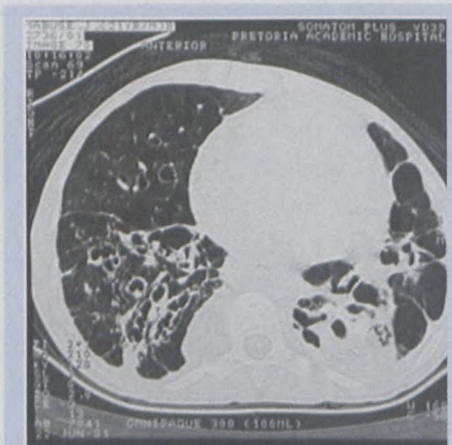


Fig. 4. Bilateral basal bronchiectasis with left lower lobe fibrosis causing 'shagginess' of the pleuro-pericardial surfaces.

measurement exceeds three standard deviations. A tracheal diameter of

more than 25 mm is consistent with the diagnosis of tracheobronchomegaly.¹

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