

Recurrent pneumonia as a result of Mounier–Kuhn syndrome

Introduction

In patients with recurrent pneumonia, anatomical causes should be considered. This includes obstructive, possibly malignant, lesions in elderly patients and other rarer causes in younger patients. Primary immunodeficiency should also be considered when two or more severe bacterial infections occur in an otherwise healthy adult.

Discussion

Mounier–Kuhn syndrome is generally considered to be an acquired, sporadic dilatation of the large airways, although a congenital basis has been proposed. Pathologically described by Czylharz in 1897, tracheobronchomegaly was first clinically described in 1932 by Mounier and Kuhn (Meyer et al, 1990). Although the pathogenesis is not fully understood, it is likely to be the result of atrophy of smooth muscle and elastic tissue in the upper airways (Celik et al, 2011).

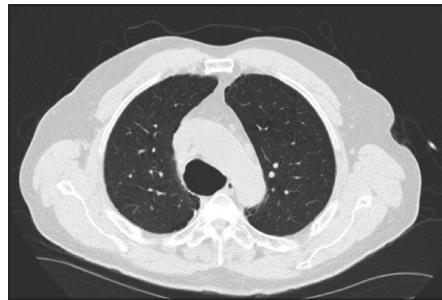
Mounier–Kuhn syndrome is associated with connective tissue diseases such as Ehlers–Danlos syndrome, and other secondary causes of the tracheobronchial dilatation include Marfan's syndrome and ankylosing spondylitis (Celik et al, 2011). The incidence is low – less than 100 cases have been reported – although the number of diagnosed cases has increased recently as computed tomography has become more commonplace. This syndrome demonstrates a male preponderance, and the usual age of onset is in the third or fourth decade of life (Bourne et al, 1995).

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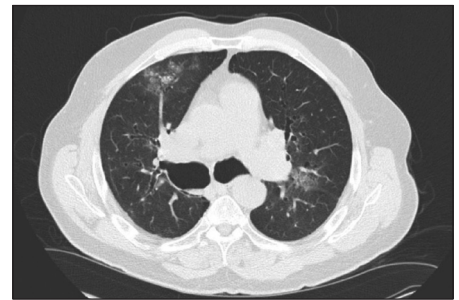
Mounier–Kuhn syndrome has a wide variety of clinical presentations ranging from completely asymptomatic to acute respiratory failure. Owing to the low incidence, there are no data suggesting usual causative organisms. The diagnostic criteria for Mounier–Kuhn syndrome in adults

Figure 1. Computed tomography scan of the trachea. The maximal transverse diameter of the trachea was 5.7 cm.



are based on diameters of the large airways; the transverse diameter of the trachea must exceed 3.0 cm in diameter, the right main bronchus must exceed 2.4 cm and left main bronchus must exceed 2.3 cm (Noori et al, 2010). This anatomia-

Figure 2. Computed tomography scan of the bronchi. The right and left main bronchi were measured at 3.8 cm and 3.2 cm respectively, which suggests Mounier–Kuhn syndrome.



Case Report

A 69-year-old man presented to the authors' tertiary level care emergency department with a 1-week history of dyspnoea, fevers and productive cough with greenish sputum. Upon presentation, the patient appeared well, was in no acute distress and was afebrile. Clinical examination revealed crackles on his left lung base. Past medical history was significant only for hypertension, hyperlipidaemia and a pneumothorax over 10 years earlier. On further questioning, the patient had experienced two prior episodes of pneumonia in the preceding 4 months. The patient denied any sick contacts or recent travel. The patient had a 20 pack/year history of tobacco use and stopped smoking 25 years ago.

Initial work up revealed an elevated white blood cell count of 16.8×10^9 /litre and no other haematological abnormalities. Chest radiography revealed right middle and lower lobe infiltrates with left basilar atelectasis. Blood cultures remained negative throughout admission. Sputum cultures revealed only normal respiratory flora. The patient was started on empiric antibiotics for community-acquired pneumonia. The patient had previously been treated with fluoroquinolones for two episodes of pneumonia with complete resolution of symptoms. The patient was admitted for work up of recurrent pneumonia in an otherwise healthy and active man.

The patient underwent computed tomography of the chest which revealed multifocal pneumonia and diffuse tracheobronchomegaly with diffuse bronchiectasis suggestive of Mounier–Kuhn syndrome. His serum immunoglobulin levels were normal apart from low IgD, which is of unclear significance.

In this patient, the maximal transverse diameter of the trachea was 5.7 cm (Figure 1). The right and left main bronchi were measured at 3.8 cm and 3.2 cm respectively (Figure 2), which suggests Mounier–Kuhn syndrome (Meyer et al, 1990; Bourne et al, 1995). The aetiology of recurrent pneumonia was identified as anatomical in nature. Following the diagnosis of Mounier–Kuhn syndrome, antibiotics with anaerobic coverage were added. The pulmonary team advised spirometry, which showed no signs of airway collapse or tracheomalacia. Within 2 days, the patient's leukocytosis and dyspnoea had resolved. The patient was discharged on oral antibiotics to complete 10 days of therapy, and appropriate follow up was scheduled with pulmonary specialists.

cal abnormality predisposes patients to pneumonia as a result of ineffectual clearance of secretions (Bourne et al, 1995).

Conclusions

Despite the high prevalence of community-acquired pneumonia, recurrent pneumonia should always be further investigated in order to elucidate the aetiology. In this case, a previously unknown structural defect in the large airways predisposed the patient to recurrent, possibly aspiration pneumonias. Imaging modality of choice is computed tomography scan. A high index of suspicion is also necessary. This diagno-

sis helped the authors choose appropriate antibiotics and also arrange appropriate follow up for secondary prevention of pneumonia. **BJHM**

Bourne TM, Raphael JH, Tordoff SG (1995)

Anaesthesia for a patient with tracheobronchomegaly (Mounier-Kuhn syndrome). *Anaesthesia* **50**: 545–6

Celik B, Bilgin S, Yuksel C (2011) Mounier-Kuhn syndrome: a rare cause of bronchial dilation. *Tex Heart Inst J* **38**: 194–6

Meyer E, Dinkel E, Nilles A (1990)

Tracheobronchomegaly: clinical aspects and radiological features. *Eur J Radiol* **10**: 126–9

Noori F, Abduljawad S, Suffin DM et al (2010) Mounier-Kuhn syndrome: a case report. *Lung* **188**: 353–4

LEARNING POINTS

- Recurrent pneumonia or infections in an otherwise healthy patient should provoke further testing.
- Evaluation for both structural and immunological causes should be pursued in this clinical setting.
- Mounier–Kuhn syndrome is a rare, but important anatomical cause of recurrent pneumonia in young patients.
- The imaging modality of choice is computed tomography and a high index of suspicion is necessary.

IMAGES IN MEDICINE

Idiopathic calcinosis cutis

An 83-year-old woman presented with hard lumps over both shins. The past medical history was significant for congestive cardiac failure, type 2 diabetes, hypertension and diverticulosis. She was bedbound as a result of spinal cord compression.

Medication included baclofen, folic acid, aspirin, omeprazole, furosemide, bisoprolol, gliclazide, pregabalin, alendronate, Adcal and clonazepam. Examination revealed multiple firm nodules over both shins (*Figure 1*) and calves. There was no tenderness, ulceration or erythema to suggest infection.

Radiographic examination demonstrated an extensive latticework of calcification in a subdermal plane, with additional vascular calcification (*Figure 2*).

The serum calcium, phosphate, parathyroid hormone and creatinine kinase levels, and auto-antibody screen were normal.

Histology from a punch biopsy showed irregular epidermal acanthosis with parakeratosis and full thickness atypia, in keeping with Bowen's disease. The underlying dermis showed chronic inflammation, vascular proliferation and perivascular aggregates of eosinophilic material. No evidence of invasive carcinoma was seen. Congo red staining was weakly positive, but immunohistochemistry for amyloid A was negative.

Idiopathic calcinosis cutis was diagnosed, a relatively rare condition whereby insoluble calcium phosphate crystals form in the skin. Most cases are not idiopathic, occurring in response to tissue injury, systemic metabolic defect, inheritable disease or an autoimmune condition (usually systemic sclerosis or dermatomyositis) (Gutierrez and Wetter, 2012).

There is no accepted first-line therapy, with many proposed, including surgical

excision, laser therapy, extracorporeal lithotripsy, diltiazem, minocyclin, colchicine, probenecid, warfarin, bisphosphonates, aluminium hydroxide and intralesional steroids (Reiter et al, 2011). Management hinges on the extent of the condition, tissues involved and symptomatology, which in this case did not require treatment. **BJHM**

Gutierrez A Jr, Wetter DA (2012) Calcinosis cutis in autoimmune connective tissue diseases. *Dermatol Ther* **25**(2): 195–206

Reiter N, El-Shabrawi L, Leinweber B, Berghold A, Aberer E (2011) Calcinosis cutis: part II. Treatment options. *J Am Acad Dermatol* **65**(1): 15–22

Figure 2. Radiographic image of the right lower leg.



Figure 1. Photograph of shin of right leg.



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