Malaysian Family Physician 2009; Volume 4, Number 2&3
ISSN: 1985-207X (print), 1985-2274 (electronic)

Online version: http://www.e-mfp.org

Test Your Knowledge

Respiratory Clinics
DRY COUGH WITH PROGRESSIVELY INCREASING BREATHLESSNESS IN A 65-YEAR OLD MAN

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R Khajotia. Dry cough with progressively increasing breathlessness in a 65-year old man. Malaysian Family Physician. 2009; 4(2&3):103-4

CASE REPORT

A 65-year old man presented to the chest clinic with history of progressively increasing breathlessness since the past 2 years. He was apparently alright 2 years ago when he noticed that he was getting breathless on climbing a flight of steps. The breathlessness has progressively increased since then and he now also gets breathless while performing his normal daily household activities. Since 10 months he has developed a dry cough which is especially worse at night and disturbs his sleep. He is a non-smoker, non-alcoholic. He was an office worker but is now retired since 7 years.

He has been visiting his family physician since the past 6 months for the breathlessness and cough. He is being treated with metered-dose inhalers and has also twice been given a course of antibiotics. He claims he has not gained any significant relief from the treatment. The dry cough has persisted and so has the breathlessness.

On general examination, the patient appears to be mildly dyspnoeic with a respiratory rate of 28 breaths per minute. He is afebrile with a pulse rate of 78 beats per minute. His fingernails reveal a 'parrot-beaked' appearance (grade III clubbing). On examination of his chest, there are coarse rales heard bilaterally at both lung bases (left > right). These rales are akin to the sound heard on separating two pieces of velcro, hence termed *"velcro" rales.* A chest radiograph is taken (Figure 1). On reviewing the chest x-ray it is decided to further thoroughly investigate the patient.

QUESTION

1. How would you interpret the chest radiographic findings?
2. What is the likely diagnosis in this patient?
3. What further investigations would you perform in this patient in order to confirm the diagnosis?
4. What is the likely aetiology of this condition?
5. What are the treatment options available?
6. What is meant by a "burnt-out" condition in this disease?

ANSWER

1. The chest radiograph shows a typical "ground-glass" appearance in the mid-and-lower zones of both lung fields bilaterally (Figure 1). On closer screening of these areas, it is observed that there are also multiple "reticulo-nodular" shadowings seen (Figure 2, marked by arrows).

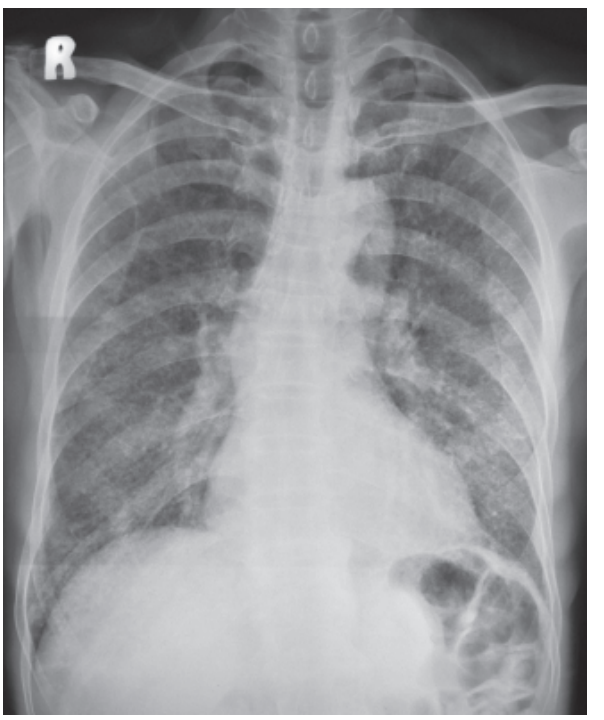

Figure 1.

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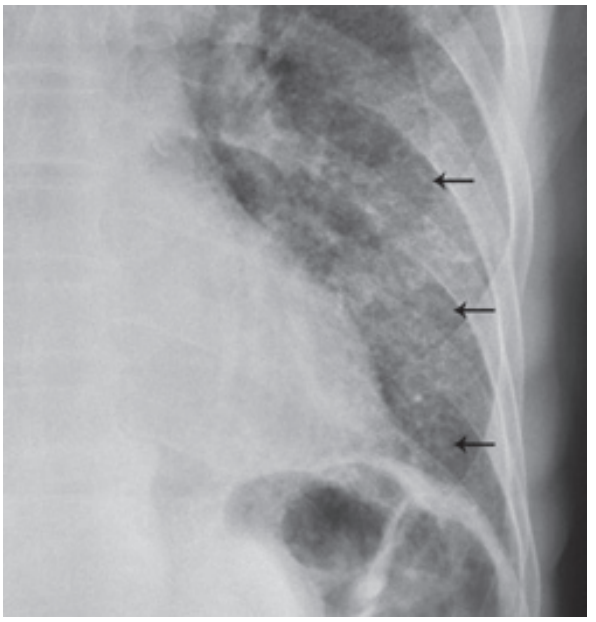

Figure 2. Chest radiograph showing ground-glass appearance and reticulo-nodular shadowings in the mid-zone.

2. Interstitial lung disease (ILD) / idiopathic pulmonary fibrosis (IPF) / cryptogenic fibrosing alveolitis (CFA) (all three terminologies are synonymous with each other).

3. In order to confirm the diagnosis the following investigations are necessary:

   (a) High-resolution CT scan of the chest: It would confirm the presence of ground-glass appearance and reticluo-nodular shadowings in the lung parenchymal tissue and interstitium, and would also determine their exact location.

   (b) Lung function tests, which include spirometry, body plethysmography and diffusion studies for carbon monoxide (DLCO): Spirometry would show a restrictive impairment as evidenced by a fall in the FVC, while the FEV1 and FEV1/FVC ratio would both be normal. There would be a fall in the DLCO indicating involvement of the lung interstitium and consequent diffusion impairment.

   (c) Arterial blood gas (ABG) investigations, to determine the level of hypoxia, carbon dioxide retention and thus assess diffusion impairment and arterio-venous shunting (usually in end-stage disease).

   (d) Blood investigations: ESR is usually high in these patients. Autoantibodies such as rheumatoid factor (RA), anti-nuclear antibodies (ANA) and anti-neutrophil cytoplasmic antibodies (ANCA) may also be present.

   (e) Fibreoptic bronchoscopy: To confirm the diagnosis by performing a transbronchial lung biopsy (TBLB) and to determine the activity of the disease process by performing a broncho-alveolar lavage (BAL).

   (f) Open lung biopsy if required, for a definitive diagnosis.

4. Interstitial Lung Disease (ILD) is basically an autoimmune disorder of unknown aetiology. Hence, it must be remembered that multiple organ systems may also be involved such as the kidneys, joints, liver, thyroid, etc.

5. Treatment options available include:

   (a) Steroids: oral and intravenous

   (b) Immmunosuppressants such as azathioprine, cyclophosphamide

   (c) Supportive therapy such as bronchodilators and continuous oxygen therapy, in severe cases. Antibiotics are used to combat infection, when required.

6. When interstitial lung disease has been long-standing, untreated and has progressed significantly, therefore develop large areas of fibrosis with honey-combing (cavity formation) in the lung tissue. This is termed, “burnt-out” interstitial lung disease as this is basically destroyed lung and heralds a poor prognosis in the patient. Treatment is usually ineffective in such cases. This is also known as “end-stage” interstitial lung disease.

Through this case report we have attempted to help family physicians make a clinical and radiological diagnosis of interstitial lung disease with a fair degree of accuracy, thereby enabling them to advise such patients on further investigations and treatment options available.

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