Bronchiolitis obliterans — an illustrative case following toxic fume exposure

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Abstract

Bronchiolitis obliterans is a condition where irreversible narrowing of small airways leads to chronic airflow obstruction. Usually an associated underlying condition can be identified. The diagnosis depends upon this together with the appropriate lung function abnormalities and radiological features. We describe a case of bronchiolitis obliterans that developed after toxic fume inhalation and which demonstrates the typical clinical, physiological and radiological features on which the diagnosis of this condition is made.

Introduction

Bronchiolitis obliterans is a condition where chronic irreversible airflow obstruction develops in the small airways in response to some associated noxious stimulus. Making the diagnosis depends upon a constellation of diagnostic features including: (*i*) the appropriate history of an associated underlying causative pathology or noxious stimulus; (*ii*) clinical evidence of diminished lung function; and (*iii*) the appropriate correlative radiological features.

Case report

A previously healthy 26-year-old male presented with shortness of breath and wheezing. An industrial accident had involved toxic fume inhalation some 5 months prior to this. An obstructive picture was obtained on lung function testing. A high-resolution CT scan of the chest was requested to investigate the lung abnormality further.

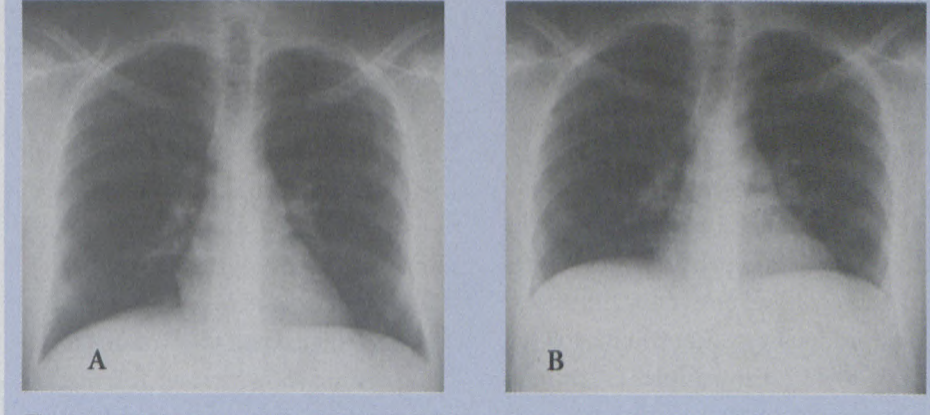
Inspiratory and expiratory chest radiographs were normal (Fig. 1).

High-resolution CT images of the lungs were obtained using 1.5 mm collimation and 10 mm spacing during both end-inspiration and end-expiration. The images were initially viewed at a window level of –645 Hounsfield units (HU) and a window width of 1 660 HU, and the appearance suggested a mosaic attenuation pattern accentuated with end-expiration compatible with areas of air trapping (Fig. 2). This appearance was further exaggerated by decreasing the window level to –902 HU and the window width to 1 023 HU (Fig. 3). Bronchiectasis was present involving the lower lobe segmental bronchi (Fig. 4). There were no areas of consolidation, and no reticulonodular opacities were noted.

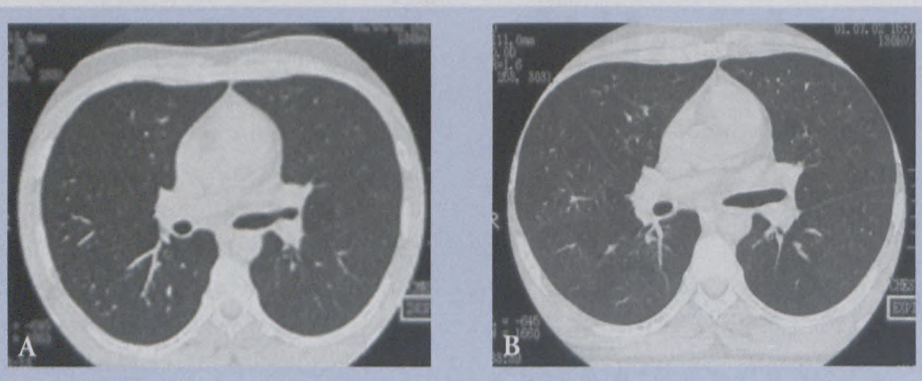
On the basis of the lung function testing and the CT appearance, and given the history of toxic fume exposure, a diagnosis of bronchiolitis obliterans was made.

Discussion

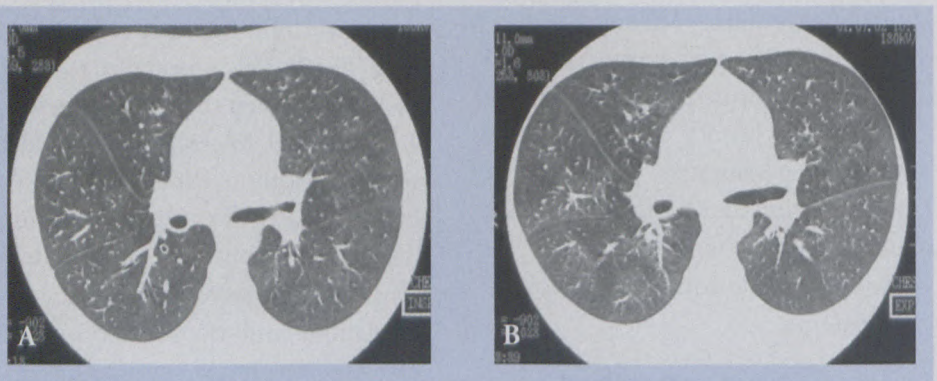
Bronchiolitis obliterans (also termed constructive bronchiolitis, or obliterative bronchiolitis) is charac-



Figs 1a and 1b. Inspiratory (1a) and expiratory (1b) frontal chest radiographs appear normal, particularly with a normal pulmonary vascular appearance and no evidence of air trapping.



Figs 2a and b. High-resolution scan slices taken during inspiration (2a) and expiration (2b). Areas of mosaic attenuation are apparent on the expiratory phase scan.



Figs 3a and b. The same scan slices as in Fig. 2 taken at different window settings that further accentuate the mosaic attenuation pattern. The darker areas are the normal ones indicating areas of air trapping and hypoperfusion.

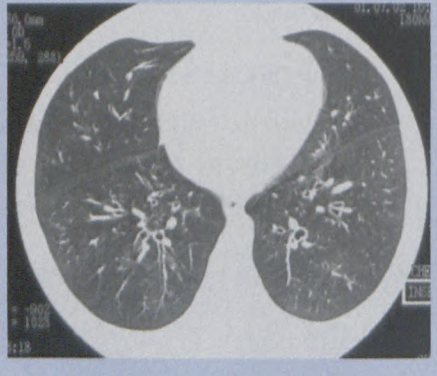


Fig. 4. Bronchial dilation and wall thickening are also present.

terised histologically by fibrosis of the submucosal and peribronchial tissues of terminal and respiratory bronchioles producing concentric narrowing of the bronchiolar lumen. Characteristically there is very little associated inflammation. The clinical severity is dependent upon the number of bronchioles involved as well as the degree of narrowing of their lumina. It is non-uniform in distribution. No inflammatory or infiltrative changes are seen in the adjacent parenchyma.

The development of bronchiolitis obliterans is closely related to a number of associated pathologies (Table I). It is very rarely truly idiopathic.

The diagnosis is extremely difficult to make at open lung biopsy, and relies more upon the combination of clinical features, lung function testing and the radiographic findings. Usually there is a history of some relevant associated pathology. Lung function testing shows irreversible airflow obstruction with a forced expiratory volume in 1 second (FEV1) of less than 60% of the predicted value in the absence of any other cause of airway obstruction.

Plain film chest radiography may show either no abnormalities or evidence of mild air trapping or hyperin-

Table I. Aetiological associations of bronchiolitis obliterans

1. Post infective
   - Viral
     - Adenovirus
     - Respiratory syncytial virus
     - Influenza
     - Parainfluenza
     - Cytomegalovirus
     - HIV
   - Non viral
     - Mycoplasma pneumonia
     - Pneumocystis carinii
     - Bacterial pneumonia
2. Toxic fume inhalation
   - Nitrogen dioxide (silo-filler's disease)
   - Sulphur dioxide
   - Ammonia
   - Chlorine
   - Phosgene
3. Drug reaction
   - Penicillamine
   - Gold
   - Lomustine
4. Connective tissue disorders
   - Rheumatoid arthritis
   - Sjögren's syndrome
   - Polymyositis
5. Transplantation
   - Bone marrow transplant (graft-versus-host rejection)
   - Lung transplant
   - Heart and lung transplant
6. Miscellaneous
   - Inflammatory bowel disease
   - Pulmonary neuroendocrine cell hyperplasia/carcinoid microtumours
   - Bronchopulmonary dysplasia
   - S. androgynus ingestion
   - Bronchiectasis, cystic fibrosis
   - Cryptogenic — very rare

flation and mild peripheral pulmonary vascular attenuation.

The high resolution CT (HRCT) findings include the following:

1. **The 'mosaic attenuation' or 'mosaic perfusion' pattern.** This consists of multifocal areas of decreased lung attenuation, i.e. 'darker areas', which may be either poorly or sharply defined. These areas of decreased attenuation result from a combination of air-trapping secondary to the bronchiolar narrowing as well as reduced perfusion due to secondary hypoxic vasoconstriction in these poorly ventilated areas. As a result there is a preferential shunting of blood to the normally ventilated areas of the lung which accentuates the mosaic appearance. With more extensive lung involvement the mosaic pattern is replaced by a more homogeneous hypoattenuation pattern. The term 'mosaic attenuation' is somewhat of a misnomer as the pathological areas of the lung parenchyma are actually the darker ones, i.e. the areas of reduced beam attenuation. The mosiac pattern becomes even more exaggerated on expiratory HRCT images as the areas of air trapping remain dark whereas the normally perfused areas (which are relatively hyperperfused) become relatively denser and smaller in cross-sectional area.

2. **Decreased calibre of the pulmonary vasculature in affected areas.** There is a reflex hypoxic vasoconstriction in the areas where air trapping is found. With larger confluent areas of involvement this results in the narrowing of macroscopic pulmonary arterial branches. Eventually this vascular constriction becomes permanent.

3. **Bronchiectasis and bronchial wall thickening.** In many cases both bronchial dilation and wall thickening are seen in the affected areas. Occasionally the obstructed bronchioles can fill with inspissated secretions creating small centrilobular branching opacities or the 'tree-in-bud' appearance.

Finally it is important to note the following points concerning bronchiolitis obliterans. Firstly, if the areas of air-trapping are extensive enough the characteristic mosaic pattern may not be seen and the lungs may appear homogeneously dark on end-expiratory scans. On the other hand, subtle changes may require manipulation of the image window levels and widths to show the affected areas adequately.

Secondly, the signs mentioned above are not unique to bronchiolitis obliterans and may be seen in other obstructive pulmonary diseases. Therefore the diagnosis is made based upon a combination of the clinical features, HRCT findings, and history or clinical evidence of any relevant associated cause or underlying condition.

Thirdly, the Swyer-James or Macleod's syndrome is probably the same pathological process occurring in the relatively immature lungs of infants and young children, usually in response to a viral lung infection.

And lastly, as the anatomical changes related to bronchiolitis obliterans are permanent due to bronchiolar fibrosis with little or no associated inflammatory component there is generally a poor clinical response to steroid therapy.

Conclusion

The diagnosis of bronchiolitis obliterans cannot be made on the radiological features alone but requires correlation with the lung function testing and relevant clinical information as well. HRCT scanning is useful in showing areas of air-trapping, secondary circulatory shunting and vascular attenuation, but requires meticulous attention to the acquisition and presentation of the images so as to optimise and accentuate the mosaic attenuation pattern and other associated features.

Recommended reading

1. Webb WR, Müller NL, Naidich DP. Airway's diseases. In: Webb WR, Müller NL, Naidich DP, eds. *High Resolution CT of the Lung.* 3rd ed. Philadelphia: Lippincott, Williams and Wilkins, 2001: 467-546.
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