

Bilateral Basal Bronchiectasis or End-Stage Interstitial Lung Disease? A Clinical Diagnostic Dilemma

Rumi Khajotia*

Consultant Pulmonologist, Hospital Tuanku Ja'afar and International Medical University, Seremban, Malaysia

***Corresponding Author:** Rumi Khajotia, Consultant Pulmonologist, Hospital Tuanku Ja'afar and International Medical University, Seremban, Malaysia.

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Distinguishing bilateral basal bronchiectasis from end-stage interstitial lung disease (advanced fibrotic interstitial lung disease) can pose a serious challenge and a diagnostic dilemma in clinical practice.

However, it is important for the discerning physician to be able to distinguish between the two with a fair degree of accuracy as the treatment options would vary.

Basically the two conditions, while having a similar history and physical findings, have some subtle distinguishing features which the astute clinician would do well to realize.

Clinical history

In a case of bronchiectasis, the history would typically be one of multiple infective exacerbations over a period of many years, accompanied by cough with large quantities of purulent sputum which is more easily expectorated out when the patient lies in a particular position (postural) and also by chest physiotherapy. These patients usually require treatment with oral or intravenous antibiotics off and on whenever they experience an infective exacerbation.

In end-stage interstitial lung disease too, the above symptoms of cough and purulent expectoration would be present, but in these patients, while delving deeper into the history, it would be noted that the patient indeed had a long history of dry cough with progressively increasing breathlessness for many years prior to the present symptoms.

This critical past history would certainly lead to suspicion of interstitial lung disease (ILD) rather than bilateral basal bronchiectasis, in such cases.

Physical examination

In both, bilateral basal bronchiectasis and end-stage interstitial lung disease, patients may have breathlessness and may be tachypnoeic. Clubbing is usually present in 70% cases of both bronchiectasis and interstitial lung disease, but it can also be absent in a significant number of patients (30%). In both cases, chest wall movements may either be normal or bilaterally diminished. Percussion findings in many cases may be non-specific, with areas of dullness or even hyper resonance, depending on size of cavities, their proximity to the chest wall and whether they are filled with secretions or not. However, auscultatory findings can be quite useful for the discerning physician. Patients with bronchiectasis would have coarse, bubbling or 'leathery' rales, very similar to those present in end-stage interstitial lung disease where there is widespread honey-combing and fibrosis. However, in patients with interstitial lung disease 'velcro' rales too would be heard in some areas, as patients with ILD have the disease progressing in different stages at different parts of the lungs. So, while some parts of the lung may exhibit ground-glass changes, others may have predominant reticulonodular changes while at other parts of the lung predominantly honey-combing and fibrosis may be seen.

Therefore, while coarse crepitations (rales) which are quite indistinguishable from those of bronchiectasis may be heard in areas of honey-combing and fibrosis (end-stage/burnt-out ILD), 'velcro' rales too would be heard in areas with active interstitial lung disease (ground glass and reticulonodular changes), in the same patient.

Hence, unlike bronchiectasis, in patients with end-stage ILD the auscultatory findings may vary in different parts of the lungs, depending on the stage of the disease process there. This is an important indicator for the astute physician when trying to distinguish bilateral basal bronchiectasis from end-stage (advanced fibrotic/burnt-out) ILD.

Non-invasive investigative findings

Plain chest radiography can also help distinguish between the two conditions. Patients with bronchiectasis would typically show multiple areas of cavitations, while patients with end-stage ILD could show a myriad of radiographic findings such as areas of ground-glass shadowings, interspersed with areas of reticulonodular changes and honey-combing and fibrosis, depending on the stage of the disease process in different parts of the lung. This would lead the physician to suspect ILD rather than bilateral basal bronchiectasis in the patient. High-resolution computed tomography (HRCT) chest scan would help confirm the above radiographic findings. In case of ILD, ground-glass and reticulonodular changes interspersed with areas of honey-combing and fibrosis are distinctly seen and can be clearly distinguished from bronchiectatic changes. Lung function testing would show an obstructive impairment in patients with bronchiectasis while patients with ILD would show a predominant restrictive impairment with a reduced diffusion capacity for carbon monoxide (DLCO).

Confirmatory diagnosis of end-stage ILD

End-stage ILD can be confirmed by bronchoalveolar lavage (BAL) and an open-lung biopsy. In such a patient, BAL would show a paucity or complete absence of cellular content in the lavage fluid while an open-lung biopsy done under direct vision would show areas of fibrosis and scarring, thereby confirming the diagnosis of burnt-out/end-stage/advanced fibrotic ILD.

Conclusion

It is of vital importance to be able to distinguish bilateral basal bronchiectasis from interstitial lung disease, as the treatment options vary.

While the treatment for both, bilateral basal bronchiectasis and end-stage ILD are similar (oxygenation, antibiotics, antifungals, bronchodilators, treatment of hemoptysis, chest physiotherapy and supportive care), patients with ILD may also need to be treated with immunosuppressants (such as azathioprine), corticosteroids, N-acetylcysteine (mucomyst) and medications such as pirfenidone (Esbriet) and nintedanib (Ofev) which may slow the rate of disease progression, as areas of active disease may be interspersed with areas of burnt-out disease in the patient with ILD.

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