Basilar Hyperlucency in a Patient With Emphysema Due to Hypocomplementemic Urticarial Vasculitis Syndrome

Ziad Ghamra MD and James K Stoller MD MSc FAARC

Disproportionate emphysematous involvement of the lung bases, compared with the apices, sometimes called basilar hyperlucency, is an unusual radiographic pattern that has been reported primarily in patients with alpha-1 antitrypsin deficiency, but also in individuals with emphysema caused by intravenous injection of methylphenidate. We present a patient with emphysema associated with hypocomplementemic urticarial vasculitis syndrome and whose chest radiograph demonstrated basilar hyperlucency. To the extent that basilar hyperlucency has not been well recognized as a feature of hypocomplementemic urticarial vasculitis syndrome, this report extends the spectrum of causes of this unusual radiographic pattern of emphysema. *Key words: emphysema, hypocomplementemic urticarial vasculitis syndrome, alpha-1 antitrypsin deficiency, methylphenidate, basilar hyperlucency.* [Respir Care 2003;48(7):697–699. © 2003 Daedalus Enterprises]

Introduction

Disproportionate emphysematous involvement of the lung bases compared with the apices, sometimes called basilar hyperlucency (BH), is an unusual radiographic pattern that has been reported primarily in patients with alpha-1 antitrypsin deficiency (AATD), but also in individuals with emphysema caused by intravenous injection of methylphenidate. The present report extends the spectrum of conditions associated with BH by describing a patient with emphysema due to hypocomplementemic urticarial vasculitis syndrome (HUVS) and whose plain chest radiograph showed BH.

Case Summary

A 57-year-old white woman was admitted to The Cleveland Clinic Hospital with an acute exacerbation of chronic obstructive pulmonary disease. She had smoked half a pack of cigarettes per day for 30 years, having stopped smoking several years before her initial presentation with dyspnea, 10 years earlier. Because of a history of arthritis and recurrent urticarial skin lesions, the possibility of HUVS was entertained and confirmed by low serum C3 and C4 levels (53 mg/dL and 4 mg/dL, respectively) and decreased C1q activity with positive immunoglobin G antibodies to C1q.[1] There was no history of intravenous methylphenidate use, and a serum alpha-1 antitrypsin level was normal at 157 mg/dL (1 mg/dL = 0.184 μmol/L [alpha-1 antitrypsin]).

On an out-patient visit preceding her admission, her forced expiratory volume in the first second (FEV_1) was 0.68 L (26% of predicted) and the ratio of FEV_1 to forced vital capacity was 0.27.

Her plain chest radiograph (Fig. 1) demonstrated preferential distribution of hyperlucency in the lung bases. Computed tomography (CT) sections from the lung apices and bases (Fig. 2) also suggested BH, which was confirmed by densitometric analysis, using $<$ –900 Hounsfield units as the emphysema threshold. Specifically, we calculated the ratio of lung pixels with density $<$ –900 Hounsfield units (deemed emphysematous) to those more dense than –900 Hounsfield units, by 2-mm sections separated by 10 mm, from apex to base. For example, in the patient's left lung the mean percentage of emphysema for 5 contiguous apical sections was 56.8%, versus 73.4% for 5 contiguous basal sections.

Discussion

The present report extends the spectrum of conditions associated with BH by presenting a patient with this pattern of emphysema on the basis of HUVS. Indeed, though

Ziad Ghamra MD and James K Stoller MD MSc FAARC are affiliated with the Department of Pulmonary and Critical Care Medicine, The Cleveland Clinic Foundation, Cleveland, Ohio.

Correspondence: Ziad Ghamra MD, Department of Pulmonary and Critical Care Medicine, A90, The Cleveland Clinic Foundation, 9500 Euclid Avenue, Cleveland OH 44195. E-mail: ghamraz@ccf.org.

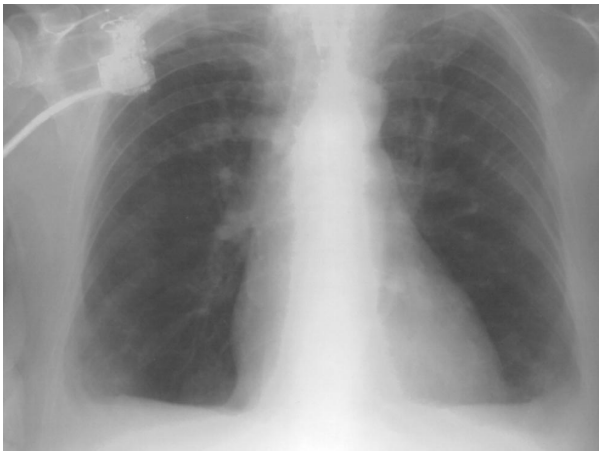

Fig. 1. Antero-posterior chest radiograph of the patient, demonstrating a pattern of basilar hyperlucency.

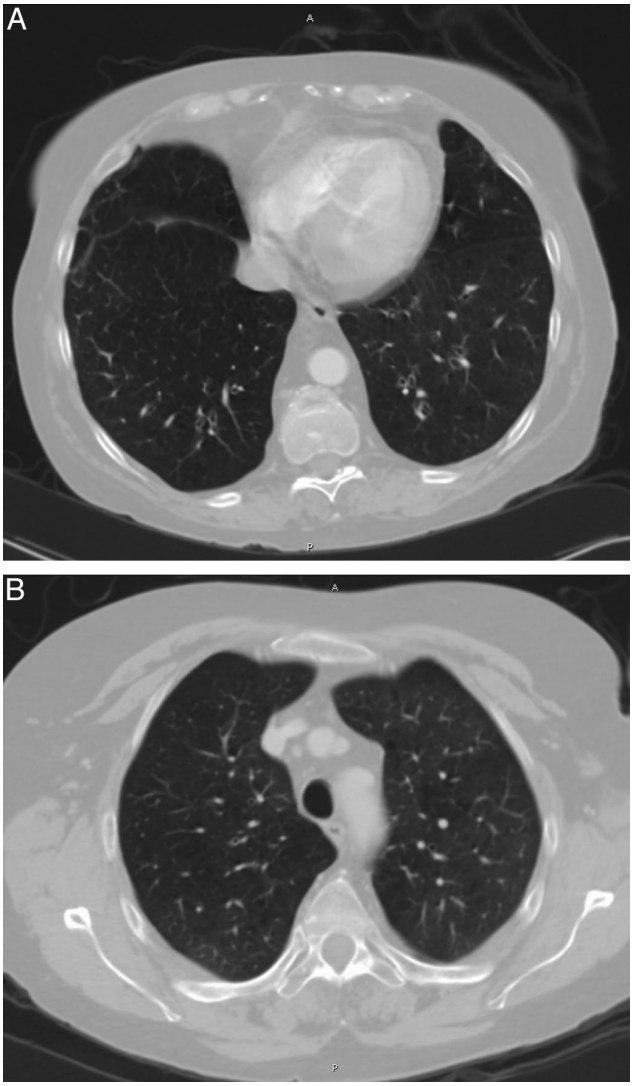

Fig. 2. Chest computed tomograms from the base (A) and apex (B) of the patient's lung, suggesting disproportionate emphysematous involvement of the base, compared with the apex. This pattern was born out by a densitometric analysis in which the percentage of pixels with densitometry < –900 Hounsfield units (indicating emphysema) showed a greater proportion of emphysema at the base than at the apex.

available studies describe BH in association with other uncommon causes of emphysema (namely AATD and intravenous injection of methylphenidate), the present report is the first, to our knowledge, to explicitly describe BH in association with HUVS. Though the patient's history of smoking raises the possibility that her emphysema was the result of cigarette smoking, factors that suggest an alternative cause were the severity of her air flow obstruction in the face of only minimal (ie, 15 pack-years) smoking and the pattern of BH, which is atypical for smoking-related emphysema.

BH has been extensively described in patients with PI*ZZ (protease inhibitor genotype ZZ) AATD, by Gishen et al.[2] In that series of 165 patients, 140 (85%) had chest radiograph evidence of emphysema (vascular attenuation or bullae), of whom 87 (62%) had preferential emphysematous involvement of the lung bases and sparing of the upper lung zones.

This pattern of disproportionate involvement of the lung bases in AATD has been confirmed by later CT studies. For example, in reviewing the CT images of 17 patients with AATD and severe obstructive airway disease (mean FEV_1 24% of predicted, range 10–45% of predicted), Guest and Hansell[3] assigned an emphysema score to each of the upper, middle, and lower zones (a higher score indicates more severe emphysema). Overall, an average distribution score of 2.7/2.8/4.5 (upper/middle/lower zone) was observed, and a lower zone predominance was confirmed in over two thirds of the cases.

A pattern of BH on both plain chest radiographs and chest CT examinations has also been described in patients with emphysema due to intravenous methylphenidate injection. In the largest available series of such patients characterized by radiographic features, Stern et al observed BH universally among the 21 patients examined (mean FEV_1 29% of predicted, range 16–79%).[4] The basilar emphysema was mild in 4 patients, moderate in 3, and severe in 14 (mean score 2.5 out of 3, where ratings represented subjective assessments of the severity of hyperinflation and vascular paucity from 0 [absent] to 3 [severe]). Available chest CT images from 3 patients confirmed this pattern, and in one patient who underwent a high-resolution densitometric chest CT, basilar predominance was confirmed by lower mean density at the lung bases than at the apices (ie, mean –900 Hounsfield units vs –825 Hounsfield units). Finally, in 3 post-mortem lung specimens that were imaged after inflation and fixation, severe panlobular emphysema was worse at the bases than at the apices. Plots of

lung attenuation in these 3 patients' lungs showed high inverse correlations (ie, $r = -0.88$ to -0.94) between the distance from the base of the lung and Hounsfield unit attenuation.

The radiographic presentation in our patient invites consideration of why the emphysematous changes disproportionately affect the lung bases. Though, to our knowledge, no definitive explanation has been advanced for BH, one proposed mechanism is the greater blood flow per unit volume in the lungs bases, allowing greater elastase burden and, at least in the case of AATD, a greater unopposed elastolysis at the lung bases.[5] Whether this explanation, if true, explains the findings in HUVS, is speculative.

As in patients with emphysema due to severe AATD or intravenous injection of methylphenidate, our patient's presentation demonstrates that emphysema due to HUVS can occur predominately at the lung bases. In the 2 largest available series of HUVS patients, which together report only 34 individuals, BH has been described in only 2 patients. In the earliest series reporting the association of HUVS and emphysema, Schwartz et al found evidence of fixed air flow obstruction in 8 of 13 patients and radiographic evidence of emphysema in 5 patients, 2 of whom were described as having "generalized decreased peripheral vasculature most prominent in lower lung fields."[6] Also, a chest CT of one of those individuals was described as showing "decreased peripheral vasculature in the middle and lower lung fields, compatible with anatomic emphysema" and with "bullae present at the lung bases." In a later, larger series of 18 HUVS patients Wisniseski et al reported emphysema in 13 of the 18 patients, though the radiographic features were not described.[1] In the other 2 reports[7,8] that, to our knowledge, describe emphysema in patients with HUVS, radiographic features were also not discussed.

In conclusion, in describing the third patient with a pattern of BH complicating HUVS, we suggest that clinicians should include HUVS, along with AATD and intravenous methylphenidate use, among the possible causes of this unusual radiographic pattern.

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