Bullous Lung Disease or Bullous Emphysema?

Ritesh Agarwal and Ashutosh N Aggarwal

Introduction

Bullous lung disease is an entity characterized by the presence of bullae in one or both the lung fields, with normal intervening lung.[1,2] On the other hand, bullous emphysema is the presence of bullae in a patient with chronic obstructive pulmonary disease (COPD), and is characterized by the presence of centrilobular emphysema in the nonbullous lung.[3–5] To select patients who are more likely to benefit from bullectomy (bullous lung disease), a proper preoperative assessment is essential.

Computed tomography (CT) can locate the bullae with considerable accuracy, even when their presence is not suspected on the basis of clinical and radiographic data. It also helps in assessing the extent and localization of bullae and associated diffuse nonbullous emphysema. Pulmonary function tests (PFTs) are also important tools in the assessment of these conditions. In addition to helping with the diagnosis, PFTs can help make an objective assessment of the severity of the underlying disease and monitor the response to treatment. The case we describe below illustrates the preoperative evaluation of a patient with giant bullous lung disease and the role of CT and PFT in the management of a patient presenting with giant bullae.

Case Report

A 50-year-old male nonsmoker presented with history of progressive exertional dyspnea of 2 years duration. At presentation to our institute the patient had Medical Research Council grade III dyspnea. There was no history of cough, chest pain, or hemoptysis. General physical examination was normal. Examination of the respiratory system revealed absent breath sounds in the whole of the right hemithorax. A chest radiograph showed multiple air-filled spaces in both the lungs, with the largest in the right hemithorax, occupying the whole of the right lung (Fig. 1). CT chest scan confirmed this finding, and also showed absence of emphysema in the nonbullous lung (Fig. 2). A provisional diagnosis of bullous lung disease was made. PFTs showed a restrictive defect and confirmed the presence of noncommunicating air spaces, as evidenced by a 3.12-L difference between the total-lung-capacity (TLC) value measured via body plethysmography and the TLC value measured via the helium-dilution technique (Table 1). In view of his symptoms, the patient underwent bullectomy. The postoperative chest radiograph showed complete expansion of the right lung (Fig. 3). PFTs repeated 3 months postoperatively showed a difference of 0.76 L between the TLC measured via body plethysmography versus via the helium-dilution technique; that difference was due to small bullae in the left lung (Table 2). A family screening to look for similar abnormalities was negative. The patient has been asymptomatic on regular follow-up for the last 2 years.

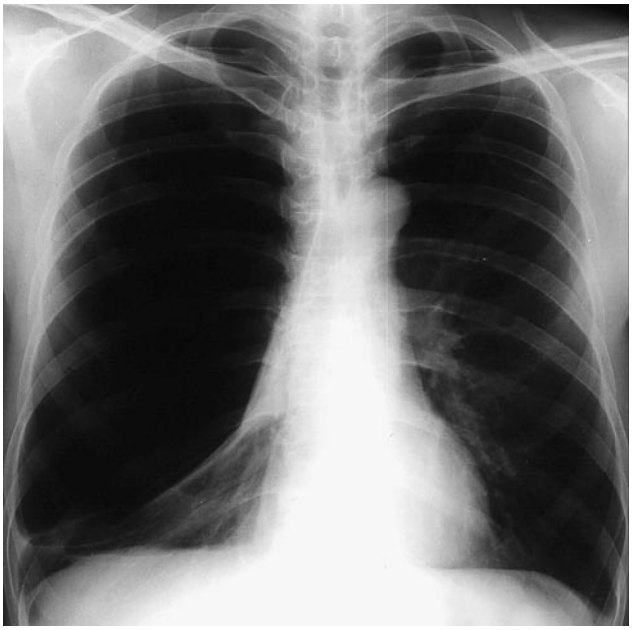

Fig. 1. Chest radiograph showing a large bulla in the right hemithorax.

Ritesh Agarwal and Ashutosh N Aggarwal are affiliated with the Department of Pulmonary Medicine, Postgraduate Institute of Medical Education and Research, Chandigarh, India.

Correspondence: Ritesh Agarwal, Department of Pulmonary Medicine, Postgraduate Institute of Medical Education and Research, Sector 12, Chandigarh 160012, India. E-mail: riteshpgi@gmail.com; ritesh@indiachest.org.

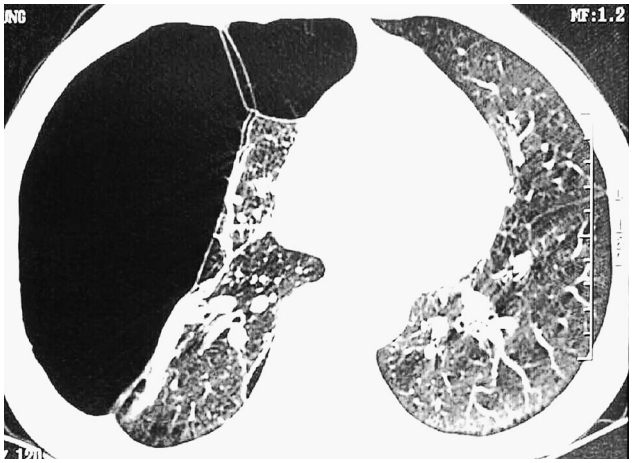

Fig. 2. Computed tomography chest scan confirming the presence of a large bulla in the right hemithorax. There is no centrilobular emphysema in the contralateral lung.

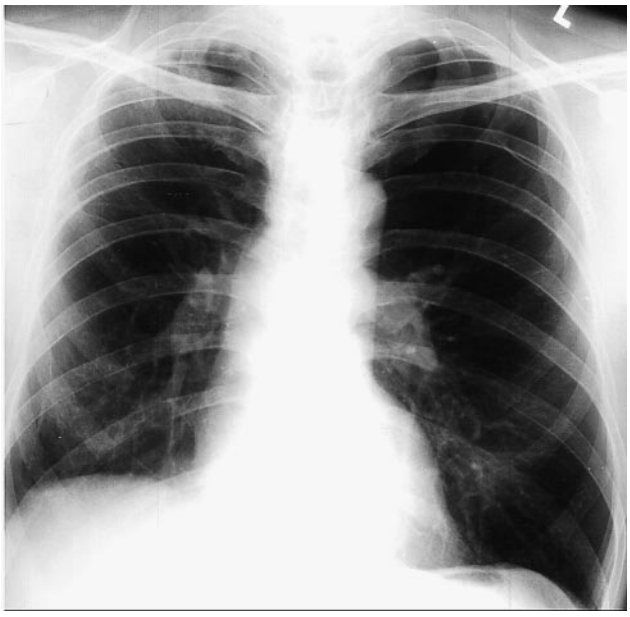

Fig. 3. Postoperative chest radiograph. The right lung is fully expanded.

Discussion

Bullous lung disease is different from bullous emphysema in that bullous lung disease has bullae with structurally normal intervening lung, whereas bullous emphysema has bullae associated with more diffusely abnormal lung parenchyma because of COPD. Giant bullous lung disease, as seen in our patient, is said to be present if the bullae occupy at least one third of the hemithorax and compress the surrounding lung parenchyma.[3] Bullectomy, either via videothoracoscopy or conventional thoracotomy, is the treatment of choice for giant bullous lung disease, even if asymptomatic.[6] Bullectomy is indicated for symptomatic patients who have incapacitating dyspnea or chest pain, and who have complications related to bullous disease such as infection or pneumothorax.

Bullectomy needs to be differentiated from lung-volume reduction surgery (LVRS), which is surgical removal of 20–30% of nonbullous emphysematous lung from each side. The recently published National Emphysema Treatment Trial[7] showed that LVRS benefits selected subgroups of COPD patients who have upper-lobe disease and poor exercise capacity. Specifically, LVRS improves 6-min-walk distance, forced expiratory volume in the first second (FEV_1), dyspnea score, and quality-of-life score, and decreases residual volume and the need for supplemental oxygen. However, patients with $FEV_1 < 20\%$ of predicted and either homogenous emphysema or carbon-monoxide-diffusion capacity $< 20\%$ of predicted do not benefit from

Table 1. Preoperative Pulmonary Function Test Results

Variable	Predicted	Measured via Helium Dilution		Measured via Body Plethysmography	
		Observed	% of Predicted	Observed	% of Predicted
FVC (L)	4.33	3.01	69.5	3.06	70.7
FEV_1 (L)	3.41	2.1	61.6	2.01	58.9
FEV_1/FVC (% of predicted)	78	69.8	89.5	65.7	84.2
TLC (L)	5.89	4.87	82.7	7.99	135.7
RV (L)	1.82	1.68	92.3	4.87	267.6

FVC = forced vital capacity
FEV_1 = forced expiratory volume in the first second
TLC = total lung capacity
RV = residual volume

Table 2. Postoperative Pulmonary Function Test Results

Variable	Predicted	Measured via Helium Dilution		Measured via Body Plethysmography	
		Observed	% of Predicted	Observed	% of Predicted
FVC (L)	4.33	4.02	92.8	4.05	93.5
FEV_1 (L)	3.41	2.9	85	3.11	91.2
FEV_1/FVC (% of predicted)	78	72	92.3	77	98.7
TLC (L)	5.89	6.13	104	6.89	116.9
RV (L)	1.82	1.98	108.8	2.77	152.2

FVC = forced vital capacity
FEV_1 = forced expiratory volume in the first second
TLC = total lung capacity
RV = residual volume

LVRS and have unacceptable perioperative mortality.[7] Thus, taking a corollary from the National Emphysema Treatment Trial, some patients with bullous emphysema may also benefit from bullectomy. However, LVRS has distinct indications applicable only to a subset of patients, and with different expectations and outcomes than bullectomy.

Patient selection remains one of the most important aspects of successful surgery, since bullous lung disease is associated with excellent postoperative outcomes, whereas surgery for bullous emphysema is not very rewarding, except probably in a select group of patients.[4,8] In general, the freedom from long-term return of dyspnea is proportional to the quality of the remaining lung after bullectomy.

High-resolution CT is an important tool for preoperative assessment, because it can identify underlying centrilobular emphysema, which is synonymous with a diagnosis of bullous emphysema. Moreover, high-resolution CT also allows assessment of associated diseases such as bronchiectasis, infected cysts, pleural disease, and pulmonary hypertension.[4,5] PFTs can also differentiate between the 2 entities. PFT values from a patient with bullous lung disease typically show a restrictive defect, whereas those from a patient with bullous emphysema show an obstructive defect. In addition to helping in differential diagnosis, PFTs also help in quantifying the size of bulla and objectively documenting postoperative improvement.[6]

There is a difference between the lung volume measured via the helium-dilution technique and that measured via body plethysmography. In the former, the subject rapidly breathes in and out of a reservoir that contains a known volume of gas and a trace amount of helium (an inert gas, very little of which absorbs into the pulmonary circulation). The helium is diluted by the gas that was previously present in the lung. With the knowledge of the gas in the reservoir and the initial and final helium concentrations, the functional residual capacity and the TLC can be calculated. The helium-dilution technique may underestimate the exact volume of gas in the lung because of inadequate time to equilibrate with slowly communicating and noncommunicating air spaces such as bullae. However, lung volume can be more accurately measured and should be measured in these cases, with body plethysmography, which measures the total volume of the thorax. In fact, the difference in TLC between the 2 techniques (body plethysmography minus helium-dilution) approximates the volume of the bullae. In our case, there was substantial clinical improvement with decrease in bulla volume, documented on postoperative PFT (bulla volume fell from 3.12 L to 0.76 L).

In conclusion, this case exemplifies the importance of selecting the correct pulmonary function test (body plethysmography) for measuring lung volume, and the utility of CT in the evaluation of patients with bullous lung disease. The presence of bullae and the etiology (in this case, bullous lung disease) was confirmed by high-resolution CT. PFTs suggested the etiology and confirmed physiologic improvement after surgery.

REFERENCES

1. Roberts L, Putman CE, Chen JTT, Goodman LR, Ravin CE. Vanishing lung syndrome: upper lobe bullous pneumopathy. Rev Interam Radiol 1987;12:249–255.
2. Murphy DM, Fishman AP. Bullous diseases of the lungs. In: Fishman AP, Elias JA, Kaiser LR, Fishman JA, Senior RM, Grippi MA, editors. Fishman's pulmonary diseases and disorders. New York: McGraw-Hill; 1998:849–866.
3. Stern EJ, Webb WR, Weinacker A, Muller NL. Idiopathic giant bullous emphysema (vanishing lung syndrome): imaging findings in nine patients. AJR Am J Roentgenol 1994;162(2):279–282.
4. Morgan MD, Strickland B. Computed tomography in the assessment of bullous lung disease. Br J Dis Chest 1984;78(1):10–25.
5. Carr DH, Pride NB. Computed tomography in pre-operative assessment of bullous emphysema. Clin Radiol 1984;35(1):43–45.
6. Greenberg JA, Singhal S, Kaiser LR. Giant bullous lung disease: evaluation, selection, techniques, and outcomes. Chest Surg Clin N Am 2003;13(4):631–649.
7. Fishman A, Martinez F, Naunheim K, Piantadosi S, Wise R, Ries A, et al; National Emphysema Treatment Trial Research Group. A randomized trial comparing lung-volume-reduction surgery with medical therapy for severe emphysema. N Engl J Med 2003;348(21):2059–2073.
8. Fitzgerald MX, Keelan PJ, Cugell DW, Gaensler EA. Long-term results of surgery for bullous emphysema. J Thorac Cardiovasc Surg 1974;68(4):566–587.