

Autobullectomy in Idiopathic Giant Bullous Lung Disease

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CLINICAL SUMMARY

A 26-year-old male with a previous history of five pack-years tobacco smoking presented with progressive dyspnoea and cough of four years duration. His vital parameters were normal with pulse oximetry saturation of 95% on room air. Chest auscultation showed reduced breath sounds and rhonchi.

INVESTIGATIONS

Chest radiograph (postero-anterior view) showed a giant bulla on the right side (Figure 1). High resolution computed tomography (HRCT) (Figures 2 A and B) showed bullae affecting both upper lobes with a giant right upper lobe bulla occupying significant volume of the hemithorax. The intervening lung parenchyma was normal. Two-dimensional echocardiography was within normal limits. Spirometry showed forced expiratory volume in the

first second (FEV_1) 42% predicted; forced vital capacity (FVC) 82% predicted; and FEV_1/FVC ratio 44% suggestive of obstructive abnormality with a

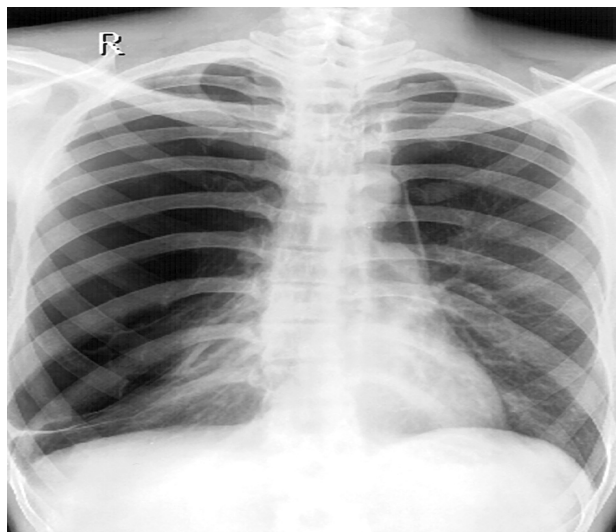


Figure 1. Initial chest radiograph (PA view) showing right upper lobe giant bulla.

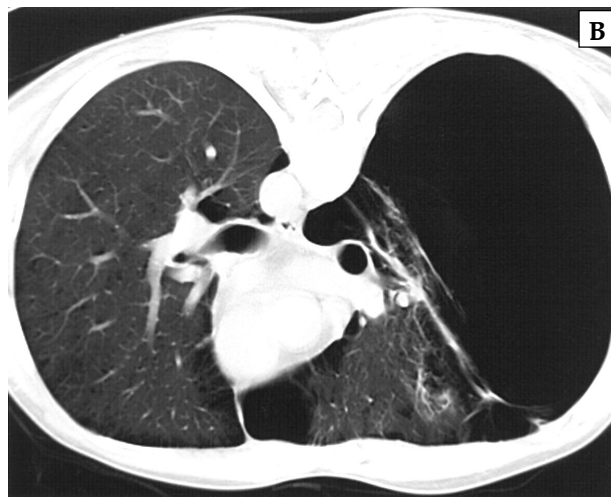
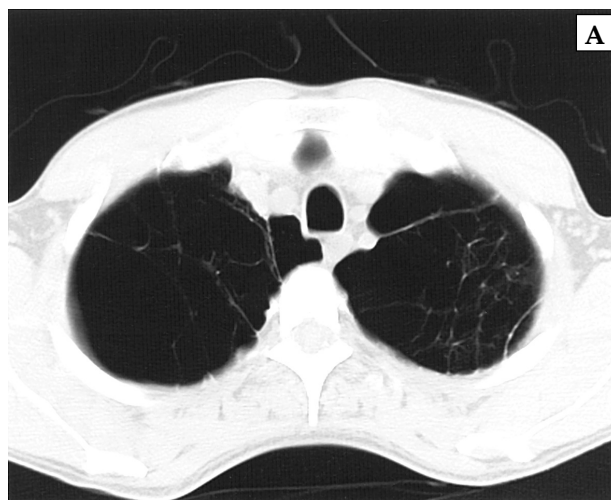


Figure 2 A and B. HRCT chest of the same patient showing bilateral upper lobe bullae with normal parenchyma in the non-bullous area of the lung.

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good post-bronchodilator reversibility. Diffusion capacity of the lung for carbon monoxide (DLCO) was normal. Lung perfusion studies showed diminished inhomogeneous uptake in the left upper lobe and absence of uptake in the right upper lobe. Haemogram and serum chemistry were normal. Serum immunoglobulin (Ig) E was 2107 IU/L (normal up to 150 IU/L) but specific IgE against *Aspergillus fumigatus* was negative. Rheumatoid factor, antinuclear antibody (ANA) and α_1 -antitrypsin were negative. The patient was treated with inhaled corticosteroids and oral bronchodilators. Surgery was planned for a later date. However, in the subsequent months he showed improvement in symptoms and repeat chest radiograph after one year of follow-up (Figure 3) showed significant reduction in the size of the right upper lobe bulla with improvement in FVC by 870 mL and FEV₁ by 420 mL.

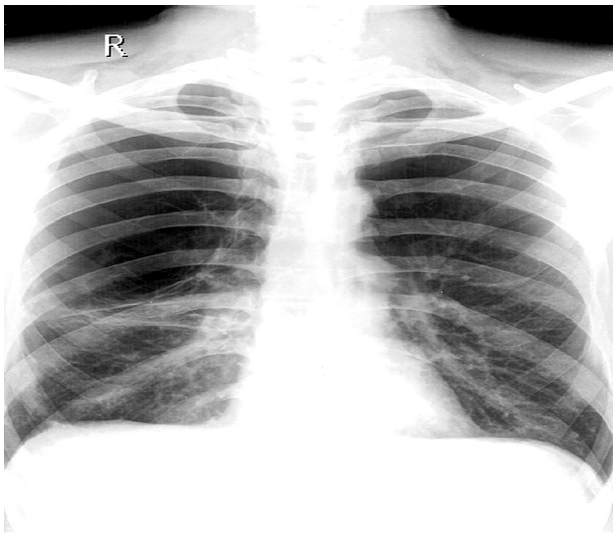


Figure 3. Follow up chest radiograph (PA view) showing marked reduction in the size of the right upper lobe giant bulla.

DIAGNOSIS

Autobullectomy in idiopathic giant bullous lung disease.

DISCUSSION

Bullous lung disease, an idiopathic clinical syndrome is characterised by the presence of bullae in one or both the lung fields with normal intervening lung, originally described by Bruke in 1937.¹ On the other hand, bullous emphysema is the presence of bullae in a patient with chronic obstructive pulmonary disease and is characterised by the presence of centrilobular emphysema in the non-bullous lung.² Giant bullous lung disease (*vanishing lung syndrome*) is a distinct clinical syndrome, characterised by large bullae that occupy a significant volume of hemithorax and are often asymmetrical, compressing the surrounding parenchyma.³ The radiographic criteria for vanishing lung syndrome⁴ include the presence of giant bullae in one or both upper lobes, occupying at least one-third of the hemithorax and compressing surrounding normal lung parenchyma. Bullectomy or lung volume reduction surgery is the treatment of choice for giant bullous lung diseases even in asymptomatics. However, autobullectomy may occur rarely. Inflammation, tumour, mucous plug or blood clot may obstruct an already compromised bronchial communication with the bulla resulting in a closed space. Gradually, air reabsorbs leading to shrinkage and spontaneous regression of the giant bulla.^{5,6} Autobullectomy has been reported to have resulted in improvement in pulmonary function as in our case and is on the same premise as lung volume resection surgery.⁷

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