Vanishing lung syndrome

Laleng M. Darlong · Ranendra Hajong · Rubul Das · Noor Topno

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It is a rare condition in which emphysematous bulla develop in the lung with the alveolar spaces gradually disintegrating to form large air sac and finally all the alveolar air sac combining to form a large or giant bulla which eventually replaces a major part of the lung parenchyma resulting in vanishing lung syndrome, originally described by Burke in 1937 [1]. The term is also used when a bulla occupies more than one-third of the haemithorax. Most of the patients affected are young male, smokers. High-resolution computed tomographic (HRCT) scan is an important tool for assessment [2, 3]. Bullous disorder of the lung is primarily a medical problem, although surgical intervention using video-assisted thoracic surgery (VATS) is beneficial in reducing chronic and disabling dyspnoea and in the management of specific complications such as pneumothorax, infection, haemoptysis or chest pain [1, 2]. Most authors agree that preventive surgery is indicated when the bulla occupies half or more of the haemithorax, compresses the normal lung, or has enlarged over a period of years [1, 4]. The natural history of such untreated giant bulla is usually dead due to respiratory failure [1].

A 20-year-old boy was detected during a routine chest X-ray to have a bullous lung disease on the right side (Fig. 1). There was no history of smoking, chest pain, recurrent cough or fever, he had mild dyspnoea medical research council (MRC) dyspnoea scale of 2. Physical

examination revealed a tall lean individual with prominent right chest ballooned out as a barrel and hyper extensible joints at the wrist and fingers. Routine haemogram and biochemistry examination was normal. Chest X-ray showed a complete loss of the right lung with wide intercostals spaces, flattened right haemidiaphragm, mediastinal shift to

Fig. 1 Chest X-ray PA view showing a right side vanishing lung

L. M. Darlong · R. Hajong · R. Das · N. Topno
Department of Surgery,
North East Indira Gandhi Regional Institute of Health and
Medical Sciences (NEIGRIHMS),
Shillong, Meghalaya, India

L. M. Darlong (✉)
E-mail: lmdarlong@gmail.com

Fig. 2 HRCT thorax showing a giant bulla with compressed right middle lobe and left mediastinal shift

left side, elongated heart with scoliosis of the dorsal spine. ECG showed sinus rhythm with left axis deviation. Pulmonary function test revealed a restrictive pattern. A HRCT scan of the thorax confirmed the chest X-ray finding of giant bulla and additionally showed a compressed right middle lobe lung herniated beyond midline and normal left lung parenchyma (Fig. 2). Magnetic resonance angiography done revealed a hypoplastic/diminished flow on the right pulmonary artery. The patient was also evaluated for other anomalies

keeping in view the likelihood of Marfan's syndrome [5], Ehlers-Danlos syndrome [6]. An echocardiography done showed normal cardiac valve and ejection fraction. An important differential diagnosis of giant bulla or vanishing lung syndrome is a pneumothorax. The “double wall” sign seen on CT scan due to air outlining both sides of the bulla wall parallel to the chest wall in bullous disease, and the “Deep sulcus” sign with radiolucency along the costophrenic angle seen in pneumothorax in supine chest X-ray helps in the differentiation [1–3]. Treatment of such cases is removal of the bulla or total pneumonectomy in the absence of functioning lung. Such procedure is now routinely done using VATS or thoracotomy [1, 2, 4].

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