CASE REPORT

VANISHING LUNG SYNDROME

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Abstract: 'Vanishing Lung Syndrome' is a rare disorder, in which emphysematois bullae appear in the lung, which progressively enlarge, causing compression of the adjacent lung. The paucity of the disorder in the literature and rarity of the condition, prompts us to report this case.

KEY WORDS: Emphysema, Emphysematous bullae, vanishing lung.

INTRODUCTION:

In this disorder, emphysematous bullae appear in the lung, the alveolar walls gradually disintegrate to form large air spaces, with eventual atrophy of the lung and compression of the adjacent side. The patients have little or no evidence of cough or respiratory infection, and finally die in respiratory failure.

This is a very rare condition. To our knowledge, our case is the first reported in the Indian literature.

CASE REPORT:

An 18 years old female from Srinagar city was evaluated for complaints of low grade fever and non-productive cough of one week duration. There were no symptoms pertaining to the involvement of any other system of body, and in her past there was no history suggestive of measles or tuberculosis or, any illness with particular reference to respiratory system or, any ailment requiring hospitalization or long term treatment. On examination she appeared to be of average built with pulse rate of 84 beats per minute, respiratory rate of 18 per minute and normal blood pressure (110/70mmHg). Overall, her general physical examination was normal. On chest examination, the left side of chest appeared bulging, and shift of trachea to right side. Percussion note was hyperresonant on the left, and cardiac dullness was absent at its usual location. On the right side, anteriorly, the percussion note appeared dull up to the costal margin. The breath sounds were diminished on left side anteriorly and posteriorly. Apex beat was not localized and the heart sounds were more clearly heard on right side of the sternum in the forth and fifth intercostal space. Rest of the systemic examination was normal.

On investigating, her hemoglobin was 11g/dL, and ESR, 05mm/hour. Her hemogram and all other biochemical investigations were within normal range. Profile for tuberculosis and systemic lupus erythematosus was negative. Chest radiograph (Fig. 1) revealed decrease of right hemithorax with shift of trachea, mediastinum and heart to the same side. Left side showed increase of volume and translucency. Lung parenchyma and vasculature were not visible on left. There appeared thin walled emphysematous

Figure 1: Chest radiograph showing hypertranslucent left side and shift of mediastinum to the right.

Figure 2: CT chest of same patient showing left hemithorax wider than the right, with overinflation and emphysematous air spaces and thickened septae in between.

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bullae on whole of the left side and the diaphragm on the same side was pushed down. Computed tomography (Fig.2), revealed the two halves of chest asymmetrical, left half more wider than the right because of overinflation of the lung; and shift of the mediastinum to the right side. On left side there were emphysematous spaces and reticular pattern between the overinflated air-spaces, and thickened septae in between. On CT, there was no evidence of any foreign body, hilar, parahilar or pleural pathology. Fibreoptic bronchoscopy revealed normal study Pulmonary function tests revealed restrictive pattern with reduced vital capacity (2.9 litres), residual volume (900ml), total lung capacity (4.3 litres) and normal FEV1/FVC (0.78). Ultrasonography did not show any cystic changes in abdominal viscera, and the facility for estimation of serum α1-antitrypsin was not available.

DISCUSSION:

The disorder starts with appearance of emphysematous bullae where alveolar walls disintegrate, and large air spaces are created leading to lung atrophy, and progression occurs till death due to respiratory failure. The basic pathogenesis is not exactly known, however it has been attributed to possible respiratory myositis. There is evidence of diaphragmatic weakness, and the condition is associated with systemic lupus erythematosus in one third of cases. Lung function tests show a restrictive pattern. The disorder needs to differentiate from few other similar conditions. Infantile lobar emphysema is an obstructive distension of one lobe in an infant, often giving rise to severe dyspnea and necessitating surgical removal. There is a strong male preponderance and about a half have congenital cardiac abnormalities; and left upper or middle lobe is usually involved, and the disorder manifests in less than 6 weeks age. In lobar emphysema with bronchial atresia, there is complete atresia of the proximal bronchus, which may be patent peripherally. The left upper lobe is transradiant and hypoplastic. Macleod's syndrome comprises of unilateral emphysema of a lung or lobe due to localized bronchiolitis or bronchitis. The pulmonary artery on the affected side is often small and there is irregular dilatation of bronchi with failure to fill the peripheral airways. The condition often occurs six months to five years after viral bronchitis, or follows tuberculosis in the childhood.

The presence of clinical and radiological features and restrictive pattern on pulmonary function testing favours the diagnosis of 'vanishing lung syndrome' in our case, who is under our follow up for future course, and may need further evaluation.

REFERENCES: