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Case report

An unusual case of unilateral hyperlucent lung

A 38-year-old male, clerk by profession, presented to our institute with complaints of cough without expectoration and difficulty in breathing for 3 years. The dyspnoea was persistent, exertional and had progressively worsened over this period of 3 years, it was currently grade 2 on the modified Medical Research Council (mMRC) grading. He was a never-smoker and there was no history of environmental tobacco smoke or exposure to noxious particles or history of illicit drug abuse. Past medical and surgical history was not significant. There was no history of significant respiratory infections during childhood and there was no history of anyone in his family with similar complaints.

He was tachypnoeic and oxygen saturation was 94% on room air. There were decreased chest movements on the left side with a hyperresonant note on percussion in the mammary, axillary and infra axillary areas anteriorly and in all the areas of the chest posteriorly. On auscultation, decreased air intensity on the same side was heard. Chest radiography was carried out (figure 1).

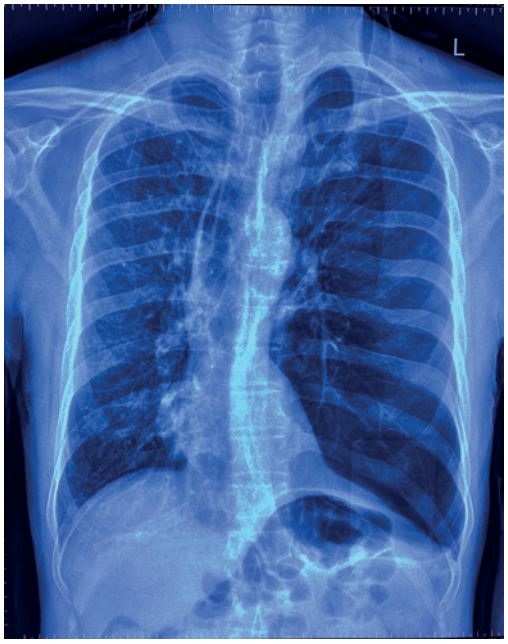

Figure 1 *Posteroanterior chest radiograph showing increased translucency of the left hemithorax with displacement of mediastinal structures to the contralateral side.*

Task 1
What is the main site of abnormality seen in the chest radiograph?

a) Pleural abnormality
b) Parenchymal abnormality
c) Chest wall abnormality
d) Airway abnormality

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Differential diagnoses of unilateral hyperlucent lung are expansive, ranging from soft tissue to pulmonary parenchyma. A systematic approach to interpretation of radiography guides us in quick localisation of the anatomical site of involvement. https://bit.ly/3fNy4vT

Answer 1

b. Parenchymal abnormality. The chest radiograph shows unilateral hyperlucent lung with mediastinal shift. There are attenuated lung markings in the left hemithorax with evidence of hyperinflation (low lying and flattened diaphragm) and shift of mediastinum to the right side. In contrast to pleural abnormality (pneumothorax), there is no line of pleural demarcation. There is presence of vascular markings, although attenuated, in the hyperlucent area.

A parenchymal defect manifesting as unilateral hyperlucent lung is usually due to two processes, *i.e.* obstructive and compensatory hyperinflation [1]. The findings in the current chest radiograph points towards obstructive hyperinflation as there is a large size hyperinflated lung field with narrowed vascular markings, the cause of which can be segregated based on the size of the diseased hemithorax. A small hilar shadow along with small sized affected lobes is characteristically seen in Swyer–James–MacLeod syndrome (SJMS), whereas large lung volumes are seen in cases of congenital lobar emphysema (CLE), bronchial atresia, and bullous emphysema [2].

Further evaluation was undertaken, and pulmonary function tests were suggestive of severe airflow limitation: forced vital capacity (FVC) 2.35 L (61% predicted), forced expiratory volume in 1 s (FEV_1) 1.17 L (36% predicted), with a FEV_1/FVC ratio of 49.7%. Arterial blood gas analysis revealed hypoxaemia (arterial oxygen tension 71.3 mmHg).

Task 2

How would you like to proceed?

a) Computed tomography (CT) scan of the chest
b) Bronchoscopy
c) Ventilation perfusion scans
d) Pulmonary function tests with lung volumes

Answer 2

a and b. Both a CT scan of the chest and bronchoscopy. Chest radiography in the current case showed signs of obstructive hyperinflation, but there are various conditions that can cause this such as foreign body aspiration, inflammatory bronchial strictures, endobronchial tumours, bronchial atresia, SJMS, congenital lobar emphysema, unilateral bullous emphysema or cystic lung diseases, and extrinsic bronchial compression due to mediastinal lesions. Common to all these conditions are the characteristic radiographic finding, *i.e.* the presence of a hyperlucent lung, with spreading and narrowing of the pulmonary vessels and on expiratory films there is no appreciable change in the volume of the affected lung, whereas the contralateral normal lung decreases in volume [3]. Thoracic CT is essential for diagnosis of the underlying cause and the majority of these conditions can be ruled out by performing bronchoscopy in addition to a CT scan. For example, evidence of blind-ending bronchi and mucoceles seen characteristically in bronchial atresia, visualisation of any intrinsic or extrinsic compression of the bronchus or presence of any strictures or bronchomalacia which can lead to signs of obstructive hyperinflation. Another significant differential is SJMS which is secondary to recurrent bronchiolitis in infancy and childhood and is characterised by diminished vascularity but a small hemithorax with associated abnormalities, such as bronchiectasis [2].

Apart from the abovementioned conditions of unilateral hyperlucent lung associated with air trapping, another important differential of CLE is congenital pulmonary airway malformation (CPAM), which can be differentiated by the presence of multiple cysts within the hyperlucent lung. Bullous emphysema is usually seen in smokers with concomitant marijuana abuse and commonly has bilateral involvement. Finally, CLE can mimic pneumothorax as both the conditions have similar findings on clinical examination and radiographically there is shift of mediastinum to the side opposite to the hyperlucent lung. The two can be distinguished by the presence of vascular markings inside the hyperlucent area in the former [1, 3].

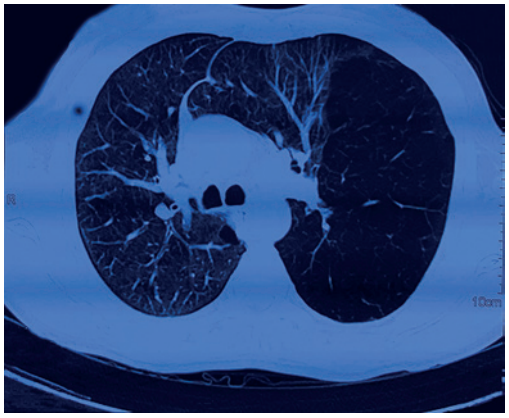

Figure 2 *Axial section of the chest CT at the level of the main carina showing shift of mediastinum to right side and hyperinflation of left lower lobe and compression of adjacent parenchyma.*

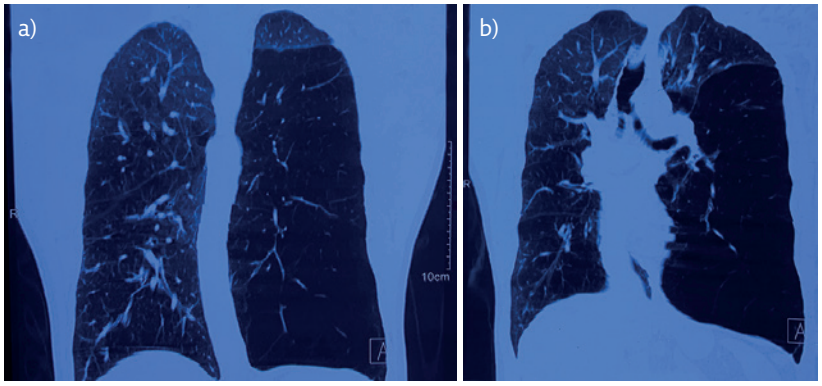

Figure 3 *a and b) Coronal sections of the chest CT showing hyperinflation of left lower lobe with displacement of oblique fissure and compression of adjacent upper lobe. Vascular markings are noted inside the hyperinflated lungs and are attenuated.*

Hence to evaluate the anatomy and associated abnormalities of the affected lobe, CT scan of chest was carried out, which showed emphysema of the left lower lobe with mediastinal shift to the right side and compression of left upper lobe, with rest of the parenchyma being normal (figures 2 and 3). Fibre-optic bronchoscopy was carried out to rule out any intraluminal obstruction and it revealed normal bronchial anatomy and patent airways. α_1-antitrypsin level was normal. Echocardiography showed no valvular lesion or shunts, and ejection fraction was 55%.

A definitive diagnosis of CLE of the left lower lobe was made as clinical and radiological features were absolute for the disease and normal bronchoscopy additionally ruled out other causes of obstructive hyperinflation.

Task 3
Which is the most common lobe involved in CLE?

a) Right upper lobe
b) Right middle lobe
c) Left upper lobe
d) Left lower lobe

Answer 3
c. Left upper lobe. The left upper lobe is most often affected (43%) followed by right middle lobe (32%) and right upper lobe (20%). Lower lobe involvement is very rare. Typically, it is unilateral with only a couple of cases reported to have multilobar or bilateral involvement [4, 5]. As remarked previously, this report describes an atypical presentation of CLE with its first manifestation in fourth decade of life and involvement of the left lower lobe, which is also known to be very rare.

Task 4
What is the best treatment option for this condition?

a) Bronchodilators
b) Inhaled corticosteroids
c) Endobronchial valve placement
d) Surgical resection/lobectomy

Answer 4
d. Surgical resection/lobectomy. In infants with respiratory distress, the treatment of choice remains resection, usually lobectomy with promising results [4, 6]. Newer and nonsurgical options, such as an endobronchial valve, have been tried in a few cases and appears to be a potential option in selected patients [7]. Evidence of adults presenting with CLE is sparse and hence management of such patients remains unclear and is mainly based on severity of symptoms and radiological features.

The patient was given a trial of inhaled bronchodilators. He had relief of symptoms and there was no limitation of activity, therefore the patient was continued on conservative management. The patient is in routine follow-up, with no clinical progression of his symptoms to date.

Discussion

CLE is a rare bronchopulmonary malformation characterised by overdistension of the affected lobe due to massive air trapping and accompanying damage to septa leading to compression and displacement of adjacent normal lung tissue [4]. It was first reported by Gross and Lewis in 1954 [8]. It has a prevalence of 4.5 per 100000, with one third of cases presenting in neonatal period itself and almost all by 6 months of age [9]. First presentation in adulthood is much more uncommon [4, 10].

The aetiology of this abnormality is still unknown, although a variety of causes have been proposed which cause obstruction of the developing airway, leading to ball valve obstruction and air trapping causing emphysema, such as cartilaginous dysplasia of bronchi, vascular anomalies, kinking of the bronchioles, compression by large lymph nodes, mucus plugs, alveolar disease and bronchial stenosis [11, 12]. The most accepted among the theories are bronchial cartilage defects like hypoplasia, flaccid tissue, or absence of cartilage. It occurs twice as often in males as in females [12]. In most cases, the left upper lobe is affected followed by right middle lobe, right upper lobe and lower lobes.

On physical examination, a hyperresonant note on percussion in the afflicted lobe is noted and breath sounds are diminished in that part of the lung. Although rare, wheezing and rhonchus can also be heard.

The most common abnormality seen on spirometry is an obstructive pattern, although it can be normal in mild or asymptomatic cases [10]. Fundamental to diagnosis are the radiological features which are markedly increased volume of the affected lobe, depression of the ipsilateral diaphragm, compression of the surrounding lung parenchyma and displacement of mediastinum to contralateral side suggesting over-inflation and air trapping in the affected lobe. Despite the attenuation of vascular markings in the affected lobe, they are well maintained and can be traced to the periphery of the lung. Chest radiography and CT scan are required to establish the diagnosis. Other adjunctive investigations are a ventilation/perfusion scan, which depicts obstructed and delayed ventilation and slightly decreased perfusion of the affected lobe, and magnetic resonance imaging, which can demonstrate any vascular lesion causing external compression [10, 13].

Diagnosis of CLE can pose a challenge for physicians as there are various conditions that can cause unilateral hyperlucent lung associated with air trapping (also known as obstructive hyperinflation). A review of the literature indicates that some patients were erroneously diagnosed with pneumothorax initially and therefore a careful reading of the radiograph is warranted [14]. A unilateral hyperlucent radiograph can itself be due to a wide variety of reasons and a structured approach can help us to narrow the list of differentials. First and foremost, we must look for the technical quality and rotation of film as it can falsely lead to hyperlucent lung on the side to which the patient is rotated. A systematic look into the details of the radiograph, moving from outside to inside can help us in localising the anatomical site of the abnormality, a summary of which is detailed in table 1.

CLE is known to be associated with congenital heart diseases in about 20% cases. Most common cardiac anomalies are left to right shunts (mainly ventricular septal defects), tetralogy of Fallot, and patent ductus arteriosus. Hence, two-dimensional echocardiography is an essential component in baseline evaluation of CLE patients [15, 16]. A strong suspicion, detailed history, chest CT and bronchoscopy are required to clinch the diagnosis and rule out the resembling diseases.

The treatment of choice for CLE depends on severity of disease [16]. The medical management of the disease includes inhaled bronchodilators and surgical treatment usually involves lobectomy [4, 6, 17]. Recently nonsurgical approaches such as endobronchial valve placement have been attempted in a few cases with promising results and this appears to be a potential future option with a less invasive approach [7]. Fierro *et al.* [18] performed a retrospective study on six infants to determine if surgical removal of the lobe would improve pulmonary function by inserting a balloon catheter in the affected lobe with bedside bronchoscopy. They postulated that temporary balloon occlusion of the lobe can assist in decision making for a lobectomy in symptomatic infants by monitoring oxygenation, gas exchange and radiological parameters after the occlusion [18].

Similar results were seen when surgical and medical management were compared in

Table 1 *Differential diagnosis of unilateral hyperlucent lung*

	Anatomical site	Radiological and supportive findings	Abnormalities
1.	Technical issue	Hyperlucency on the side to which patient is rotated	Rotation
2.	Chest wall	Normal vasculature of hyperlucent side	Poland syndrome (hypoplasia of pectoralis muscle) Mastectomy
3.	Pleural spaces	Thin sharp pleural line with no vascular markings peripheral to it	Pneumothorax
4.	Parenchymal defect	Obstructive hyperinflation: hyperlucent lung with spreading and narrowing of pulmonary vessels Characteristic appearance: at full expiration normal lung volume decreases whereas the diseased side volumes remain fairly similar Compensatory hyperinflation: small or normal sized hyperlucent lung, spreading of the vascular markings and displacement of the ipsilateral hilum with normal decrease in volume of lung in the expiratory films	Congenital lobar emphysema Swyer–James–MacLeod syndrome Unilateral bullous emphysema or cystic lung diseases Atelectasis/lobectomy
5.	Vascular defect: congenital	Hyperlucent lung, a small hilum, poor vascularisation, and ipsilateral shift of the mediastinum Normal reduction in lung size on expiration	Aplasia of a pulmonary artery (its proximal interruption) and pulmonary hypoplasia
	Vascular defect: acquired	Unilateral hyperlucent lung with no evidence of air trapping on both inspiratory and expiratory radiographs Normal sized lung and the ipsilateral hilum is normal size/ enlarged	Pulmonary thromboembolism, fibrosing mediastinitis, sequelae of mediastinal irradiation
6.	Airway defect	Abnormally large affected side even in expiratory film Obstruction in spirometry	Unilateral obstruction: bronchial atresia, extrinsic and intrinsic causes of bronchial compression (such as a foreign body), endobronchial tumours

a long-term follow-up study of CLE patients [16]. Hence, a severely symptomatic patient or progression of disease should be managed with surgical intervention whereas conservative management can be tried with close follow-up for mild or asymptomatic disease [16, 19, 20].

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Conflict of interest

None declared.

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