Sir,

Congenital lobar emphysema (CLE) is a rare congenital abnormality that is characterized by overdistension and air trapping of one or more lobes of lung, leading to a compression atelectasis of ipsilateral lung and subsequently of contralateral lung. Similar to other developmental lung anomalies, this condition is usually diagnosed in the neonatal period or during early childhood. However, we report a case of CLE that was detected in late childhood.

A 15-year old boy presented with progressive exertional breathlessness since 5 months. There was no other significant history. Patient denied any history of recurrent chest infection. According to his mother, the achievement of developmental milestones was normal but he did not gain adequate weight. His immunization history was up-to-date. On examination, he was of thin built with 13.7 kg/m² BMI. There was no cyanosis, clubbing, oedema, icterus, or lymphadenopathy, although pallor was present. Vitals were within the normal limit. On inspection, right hemithorax appeared enlarged. Movement of chest was restricted on the right side with mediastinum shifted to the left. Breath sound, vocal fremitus and vocal resonance were grossly reduced on the right side along with a resonant percussion note. Liver was palpable at 4 cm below the costal margin. A clinical diagnosis of right-sided pneumothorax was made on admission.

But chest X-ray in PA view [Figure 1] revealed hyperlucency of the entire right lung with leftward mediastinal shift without any visible visceral pleural lining. Attenuated but maintained vascularity was noted in the hyperlucent right lung. ABG analysis and other blood investigations were normal except Hb-10.4 gm/dl. Electrocardiography showed only left axis deviation without any evidence of left ventricular hypertrophy. Two-dimensional echocardiography did not reveal any cardiac abnormality. Spirometry showed a mixed ventilatory pattern with poor bronchodilator reversibility. Computed tomographic scan of thorax [Figure 2] showed hyperlucent and hyperexpanded right middle and lower lobe with collapsed right upper lobe along with mediastinal shift to left. Pulmonary vasculature on the right side appeared attenuated and displaced. Herniation of lung tissue was noted in retrocardiac space and in anterior mediastinum. The left lung was normal except for compression atelectasis. No evidence of pneumothorax or intraluminal foreign body was found. Subsequent fibreoptic bronchoscopy [Figure 3] showed a hypoplastic right upper lobe bronchus along with a normal trachea and left main bronchus.

Thereby, a final diagnosis of congenital lobar emphysema of right middle and lower lobe with right upper lobe bronchial hypoplasia was made on the basis of radiological and bronchoscopic findings. Patient was treated conservatively because of clinically less severe disease. The patient had been advised for a close monitoring and regular follow-up. He is currently being followed up since last one year and no clinical deterioration has been noted so far.

CLE is an uncommon congenital anomaly of respiratory system with a prevalence of one per 20,000 to 30,000 deliveries and the incidence is estimated to be one in 70,000.
Figure 1: Chest X-ray shows hyperlucent and hyperexpanded right lung with mediastinal shifting to the left side along with attenuated but maintained vascularity of the right lung.

Figure 2: CT scan of thorax showing marked overinflation of right middle and lower lobe along with collapsed right upper lobe with mediastinal shifting to the opposite side. There is attenuation of vascularity on the right side and herniation of right lung tissue to anterior mediastinum and retrocardiac space.

Figure 3: Fibreoptic bronchoscopy showing hypoplasia of right upper lobe bronchus, visible as a slit-like opening.

This disorder mostly presents within few hours after birth to six months. Usually CLE involves a single lobe; however, multilobar as well as bilateral involvement has also been recorded. The most common affected lobe is the left upper lobe, followed by the right upper lobe and right middle lobe. But multilobar involvement including a lower lobe is very uncommon and no such case was recorded among 30 cases as mentioned in a case series by Ozcelik et al. Such multilobar involvement (including lower lobe) was found in only three out of 28 cases as mentioned in another case series by Lincoln et al.

Breathlessness is the most common symptom associated with cough, wheezing, grunting or recurrent infection. Patients who present later in life usually have history of recurrent chest infections and frequent antibiotic use, which was not the scenario in this case. Exact aetiology is not known and no apparent cause is found in 50% cases. Congenital cartilage defect is found in 25% of cases that causes bronchial collapse at expiration, leading to overinflation of the alveoli. Other bronchial obstructions (redundant mucosal fold, mucus plugging, bronchial hypoplasia/stenosis etc.) are responsible for remaining 25% of cases. In our patient, although hypoplastic bronchus was present in association with emphysematous right middle and lower lobe, it was not related causally and we could not find such an unusual association in the literature.

Clinically CLE may mimic pneumothorax but they can be differentiated from the chest X-ray and CT finding. This differentiation is absolutely essential as because insertion of an intercostal tube in CLE cases in suspicion of pneumothorax may be deleterious to the patient. Bronchoscopy is done to look for any congenital or acquired tracheobronchial abnormality and to exclude foreign body obstruction. Cardiological evaluation must be done as concomitant congenital heart disease and CLE has been found in 12-20% cases in the literature.

Treatment of CLE is mainly surgical. Lobectomy is the standard surgical procedure that allows expansion of compressed normal lung tissue. Long-term outcome was found to be excellent with complete cure in over 85% of cases. Recently endoscopic parenchymal sparing resection has been performed in a case of CLE with suspicion of pneumothorax may be deleterious to the patient. Bronchoscopy is done to look for any congenital or acquired tracheobronchial abnormality and to exclude foreign body obstruction. Cardiological evaluation must be done as concomitant congenital heart disease and CLE has been found in 12-20% cases in the literature.

In conclusion, CLE presenting in late childhood or adulthood for the first time may lead to a diagnostic challenge to the physician and it can be detected only if a high diagnostic suspicion is maintained. The diagnosis and its differentiation from pneumothorax are important, so that unnecessary intervention for pneumothorax can be avoided and the patient can be taken up for a more definitive surgical management. Asymptomatic and less severe patients may be considered for a non-operative conservative approach with strict advice regarding regular follow-up.
Case Letters

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Sir,

A 44-year-old chronic alcoholic male was brought to the hospital after he suffered a head injury due to fall under the influence of alcohol. The patient had had repeated and excessive vomiting episodes following a drinking binge just before the fall. He had a 15-year history of alcohol abuse without any other comorbidities. At the time of admission, the patient was drowsy but arousable and disoriented to person and place. The patient was afebrile with a pulse rate of 105 per minute and blood pressure of 110/70 mm Hg. Physical examination was otherwise unremarkable. The stretch reflexes were normal and Babinski sign was not elicited. Pupils were equal, round, and reactive to light. Laboratory parameters were remarkable for the levels of serum glutamate oxaloacetate transaminase (SGOT) 258 U/L, serum glutamate pyruvate transaminase (SGPT) 140 U/L, and serum sodium level 133 mEq/L. Nasogastric tube was inserted to decompress the stomach, and intravenous normal saline along with pantoprazole 40 mg was administered. Computed tomography (CT) scan of brain was normal. The patient’s clinical condition had improved significantly on the fifth day when he was discharged. At the time of discharge, the patient had the sole complaint of mild epigastric pain. Oral pantoprazole 40 mg daily and abstinence from alcohol was advised. He presented 15 days later with high-grade fever and severe dysphagia for solids as well as liquids. A CT scan of the neck and thorax revealed presence of gas in the retropharyngeal region extending from C2 to D4 level [Figure 1] and across the midline [Figure 2]. Extension of the gas collection across the midline suggested the involvement of danger space. The pharyngeal airway

Remarkable computed tomography findings in Boerhaave’s syndrome and trachea were partially compressed and displaced anteriorly [Figure 1]. In addition, collection was seen in the superior mediastinum [Figure 3]. Presence of free air was also seen in the posterior mediastinum. The findings were suggestive of acute mediastinitis secondary to esophageal perforation, likely to be due to Boerhaave’s syndrome. Surgical exploration of the mediastinal extension of the prevertebral collection yielded 30 ml of pus. Presence of a small transmural perforation was noted along the posterior wall of upper thoracic esophagus. It was surgically repaired. Patient developed postoperative complications in the form of sepsis, bilateral pleural effusion, and collapse of the left lung, to which he finally succumbed. The pus culture grew a highly resistant strain of Klebsiella. Boerhaave’s syndrome is a transmural perforation of the esophagus, usually associated with forceful emesis. It occurs due to sudden increase in the intramural esophageal pressure produced during vomiting, as a result of neuromuscular incoordination causing failure of the cricopharyngeus muscle to relax. The condition is usually seen in alcoholics. The commonest site of esophageal tear in Boerhaave’s syndrome is the left posterolateral wall of the lower third of esophagus, just proximal to the gastroesophageal junction. The second most common site of rupture is the subdiaphragmatic or upper thoracic area. [1,2] The diagnosis of Boerhaave’s syndrome can be difficult due to lack of specific symptoms, and hence, delays in presentation and medical care are not uncommon. [3] Patients may present with symptoms such as chest pain, difficulty in swallowing, and cough. Symptoms due to