A 5 months old male infant presented with cough and increased chest activity since 4 days. He had similar symptoms at 2 months of age for which he was hospitalized and had required mechanical ventilation for 10 days. Thereafter he had similar episodes at the ages of 3 months and 4 months, each episode being severe enough to require hospitalization. During each hospitalization he was given IV antibiotics and nebulization for 5-7 days. In between the episodes of tachypnea, he remained asymptomatic. He was on exclusive breast feeds. There was no history of cyanosis, poor feeding, prolonged fever, recurrent skin infections, prolonged diarrhea or weight loss. Anthropometry revealed a weight of 6.8 kg, length 63 cm. Chest examination revealed slightly reduced movement of left chest with reduction in air entry and slight tracheal shift to the right. There was also bilateral scattered wheeze. Chest X-ray is depicted in Figure 1. Echocardiography was normal.

What is the diagnosis?

Congenital lobar emphysema (CLE). X-Ray chest of the infant shows volume loss left lung with mediastinal shift to the right suggestive of congenital lobar emphysema. CLE is a rare congenital malformation of lung development with a prevalence of 1 in 20,000 to 1 in 30,000. (1) It occurs when partial bronchial obstruction causes hyperinflation of the lung lobe which in turn compresses the normal lung parenchyma causing contralateral mediastinal shift. (2) The causes of the bronchial obstruction can be multifactorial. In 50 percent of cases there is decrease in the bronchial cartilage tissue producing a ball valve effect with consequent overinflation. In the rest of the cases bronchial obstruction may be due to vascular abnormalities, bronchial stenosis, bronchogenic cysts or endobronchial obstruction due to viscid secretions, mucus plugs or granulation tissue. (3) A chest radiograph helps in the diagnosis but is not definitive. The characteristic appearance is that of pressure expansion of a lobe of lung causing compression of adjacent lobes. The mediastinum may be displaced away from the emphysematous side. In severe cases the over expanded lung may be herniated into the contralateral hemithorax. (4) A CT scan confirms the diagnosis and may rule out associated anomalous vascular slings. The most commonly accepted form of treatment is surgical removal of the affected lobe. However conservative management may be considered in those patients with mild disease who have no respiratory distress and are able to feed and grow. (5).

References