CONGENITAL LOBAR EMPHYSEMA: A DIAGNOSTIC CHALLENGE BUT TREATABLE CONGENITAL DISORDER OF LUNGS

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Article

INTRODUCTION

Lobar emphysema is a rare abnormality and usually congenital. Its incidence is 1 per 20,000 to 30,000 live births and is characterised by marked overinflation of a single pulmonary lobe usually upper left, and less commonly right middle lobe. Lower lobes are rarely affected1. There is gross overinflation of the affected lobe and mediastinal shift to opposite side. It can often present as a diagnostic problem initially confusing with pneumonia and other causes of respiratory distress2. Diagnosis is usually based on clinical examination and plain radiography but it is not always straightforward3,4. In these cases computed tomography and ventilation perfusion (V/Q) scintigraphy may be helpful. This report describes a case, initially diagnosed as severe pneumonia/disease but eventually proved to be suffering from congenital lobar emphysema and required surgical intervention.

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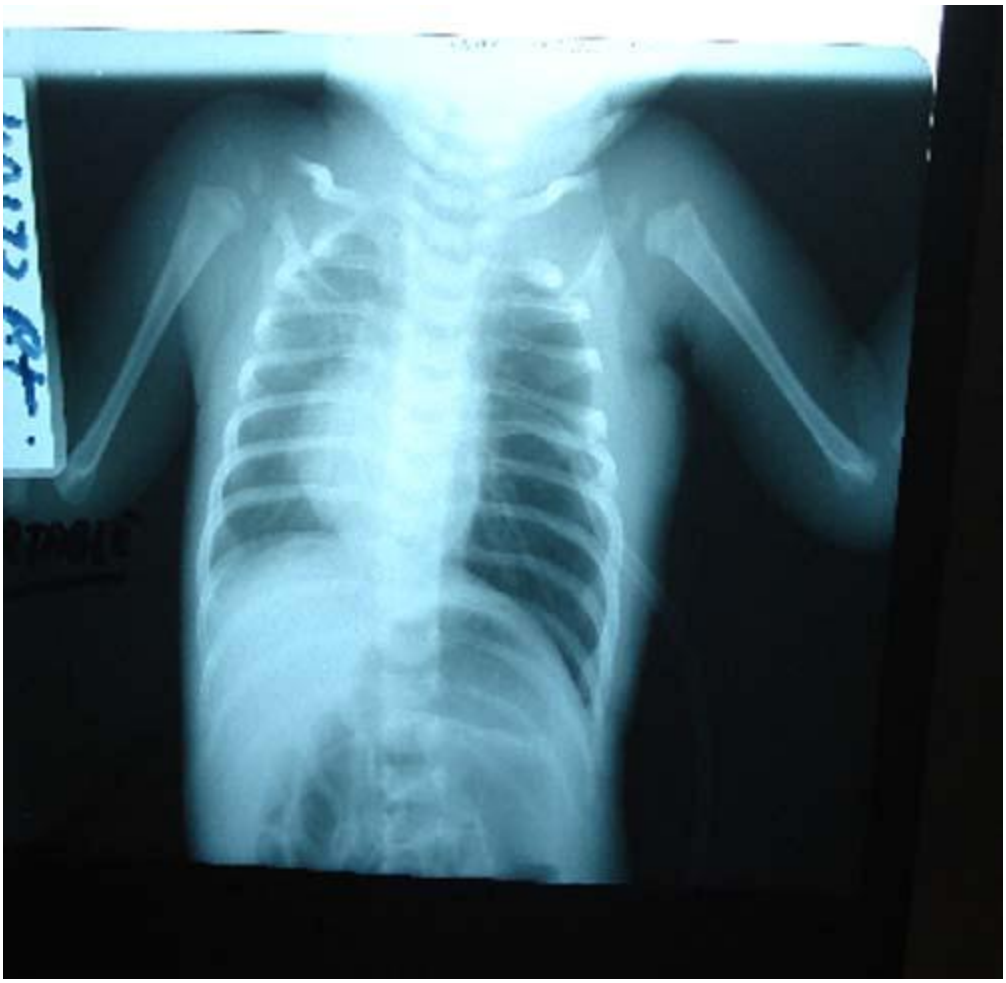
Case Report

A 3 weeks old neonate was brought to CMH Multan with c/o fever, inability to take feeds, vomiting, cough, and progressive difficulty in breathing for the last 3 days. Baby was cyanosed with SaO2 of 80%. He was tachypnoeic with severe chest indrawings. Trachea was shifted to the right, and chest movement was minimal with respiration inspite of marked tachypnea. Percussion note was relatively hyperesonant over left anterior thoracic region. Air entery was comparatively reduced on left hemithorax but there were bilateral scattered ronchi and crepitations on auscultation. Radial pulse was regular with rate of 150/min. Heart sounds were normal in intensity and character and no murmur was audible. Abdomen was soft, non protuberant. Liver and spleen were displaced 3 cm below the costal margin. Neurological examination was unremarkable. He was diagnosed as a case of severe disease according to WHO protocol of ARI(Acute Respiratory Infections). He was managed accordingly with broad spectrum antibiotics, O2 inhalation and i/v fluids. Condition of baby was stabilized within few hours. He became active, peripheral cyanosis settled and he started taking feeds orally. But his respiratory distress persisted. He remained tachypnoeic and air entry was markedly reduced on left lower chest.

Chest radiograph showed a grossly hyperinflated left hemithorax with downward shift of left hemidiaphragm and mediastinal shift to right hemithorax (Fig.1).



Cardiac shadow was unremarkable. No structural defects were seen on echocardiography. CT Scan of chest confirmed grossly emphysematous left lower lobe but with no anatomical abnormality of the bronchial wall.

Other investigations revealed TLC 9800/mm3, lymphocytes 58%, granulocytes 32%. Hb was 14.6g/dl and the platelet count 360,000/ mm3. Arterial Blood Gases revealed pH 7.37, pCO2 60.2 mmHg, pO2 120 mm Hg, HCO3- 33.2 mmol. Serum urea electrolytes and serum calcium were within normal limits.

Baby remained well for 4 days but condition deteriorated again. He became febrile, off feeds and tachypnea worsened. Chest radiograph revealed consolidation of left lower lobe and blood culture showed growth of methicillin resistant Staph aureus (nosocomial infection). Inj vancomycin was added to treatment protocol and baby responded well. Treatment was continued for 14 days and repeat chest radiograph confirmed complete resolution. Although there was marked constitutional improvement but tachypnea and intermittent bouts of cough persisted. Ventilation perfusion(V/Q) scan was planned but could not be done due to nonavailability of facilities.

A final diagnosis of congenital lobar emphysema involving left lower lobe was made and surgical resection of involved lobe was planned. Surgery was done at 1.5 months age using left postero-lateral thoracic incision. Pleura and vascular anatomy were found to be normal. Left lower lobe was found to be emphysematous and grossly enlarged. It filled left hemithorax and extended to the right displacing the mediastinum. There were no tension cysts and upper lobe was normal. Left lower lobectomy was done. Patient,s condition improved dramatically after surgery and postoperative period was uneventful. He was discharged after 5 days. Histological examination of resected lung tissue showed normal bronchial cartilage of major airways and distended alveoli.

Discussion

Congenital lobar emphysema is a rare condition, however a number of case reports and few case series have been published in international medical literature. Upon reviewing medical literature in our own country we could find only one case of congenital adenomatoid malformation mimicking lobar emphysema in a 15 months old baby reported in 20035. This may be due to failure in making diagnosis, lack of reporting habits or true reflection of prevalence of disease.

Respiratory distress is the commonest mode of presentation in CLE as witnessed in our case also. Similar symptoms may occur in bronchopneumonia, cyanotic congenital heart diseases, and several congenital abnormalities of the lung6,7. Most of the time these cases are initially diagnosed as pneumonia, aspiration syndromes or foreign body inhalation as in our case also. In more than 50% of cases, the left upper lobe is involved and there is a shift of the mediastinum. It was different in this case as this time involvement was confined to left lower lobe.

Several etiological factors have been associated with the development of CLE. In 50% of cases there is decreased bronchial cartilage tissue. This defect produces a ball valve effect with consequent overinflation1,2. Vascular abnormalities that produce compression, bronchial stenosis, bronchogenic cysts, and congenital cytomegaloviral infection have also been associated. Our patient had a normal vascular anatomy and bronchial cartilage on gross and histological examination nor were there any other lung abnormalities. Concurrent congenital heart disease (usually patent ductus arteriosus, or ventricular septal defect, occurs in 15% of cases8. Our patient had neither of these on echocardiography.

Chest radiograph is the basic investigation in CLE from which a diagnosis can be made and is readily available also. But unfortunately hurried and careless interpretatios usually result in delayed diagnosis. In an Indian series, a correct diagnosis on chest radiography before referral was made in only 4 out of 10 cases even though the diagnosis could be arrived at in all 10 cases using the initial radiographs8. A CT scan, bronchoscopy and angiopulmonography are also used in the diagnosis. CT scan is helpful in demonstrating anatomical details and rulingout different possibilities in differential diagnosis. In this child CT scan was done to exclude an extrinsic mass effect as the cause of over inflated lung and to demonstrate the bronchial anatomy before surgery.

Surgery remains definitive and reliable modality for ultimate treatment of CLE4 In over 85% of cases, long term outcome after surgery is excellent with complete cure9 Our patient improved immediately after surgery with normalization of the SaO2 within few hours.

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