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An Unusual Presentation of Congenital Cystoadenomatoid Malformation of Lung

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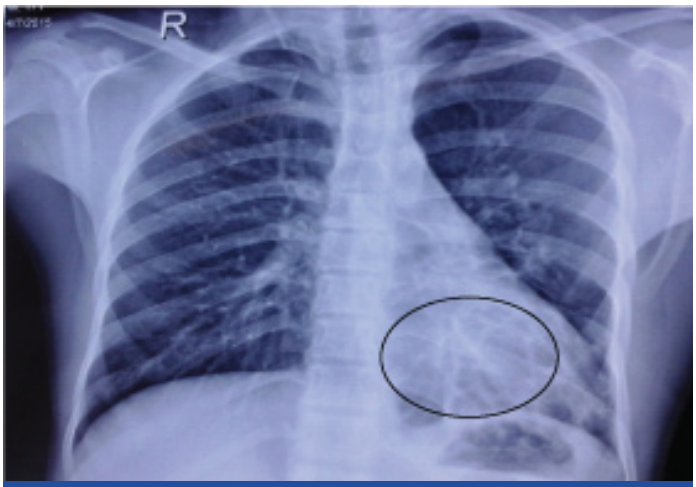
A 14-year-old male presented with dyspnoea and recurrent respiratory tract infection since two years, managed with conservative management, but symptoms aggravated and he developed three episode of left lower lobe pneumonia in last six month. This episode was not associated with persistent fever, weight loss, haemoptysis, immunocompromised condition or any cardiac illness since last one week. Patient developed left sided chest pain which got worse on deep inspiration along with fever and lower respiratory tract infection.

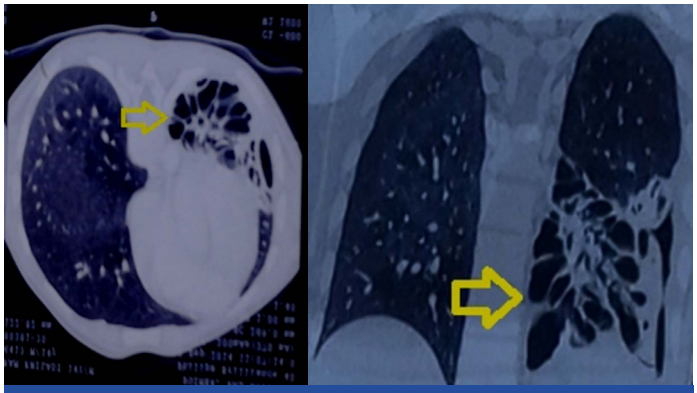
Chest radiograph PA view showed homogenous opacity in left lower zone of lung with incomplete obscuring of left costo-phrenic angle and dome of diaphragm [Table/Fig-1].

Contrast Enhanced Computed Tomography (CECT) chest revealed multiple well-defined cystic radiolucencies of variable sizes communicating with bronchi in left lower lobe, which were nearly completely replacing it. Broncheiactatic changes were observed in adjacent area and patchy areas of consolidation were noted in superior segment of left lower lobe [Table/Fig-2a,b].

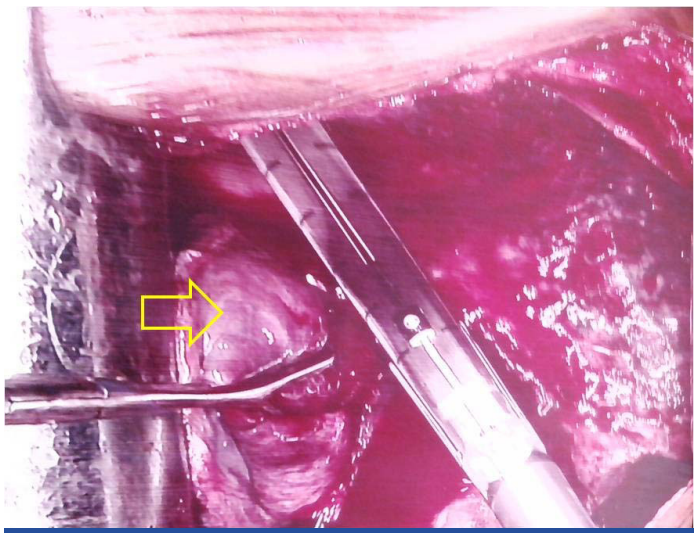
Left lower lobectomy was done. Intraoperatively large, fleshy, cystic, vascular mass involving whole left lower lobe with dense adhesion between lung mass and parietal pleura was observed [Table/Fig-3]. Resected specimen showed large cyst wall lined by necrotic debris along with multiple small cysts [Table/Fig-4]. Histopathological evaluation showed cysts lined by pseudostratified ciliated columnar epithelium with cystic and malformed bronchi and bronchioles. The findings confirmed the diagnosis of late onset left lower lobe Congenital Cystoadenomatoid Malformation (CCAM). On follow up satisfactory clinical and radiological improvement occur in nine month.



[Table/Fig-1]: Chest radiograph (Postero-anterior view) showing uniform homogenous ill-defined opacity in the lower left lung field circle. There was no evidence of pleural/pericardial effusion or pneumothorax.

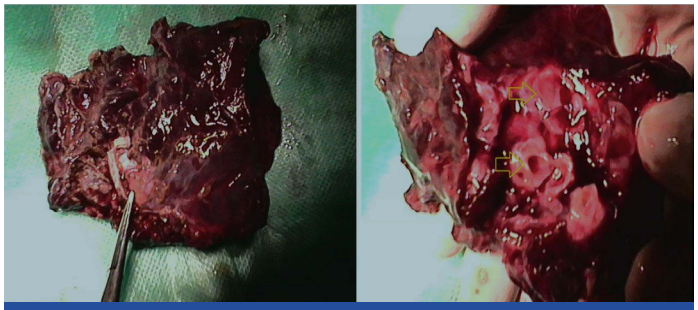


[Table/Fig-2]: (a) Contrast enhanced axial computed tomography of the chest demonstrating pneumatocele of the left lower lobe of the lung (arrow). There was presence of multilocular cystic lesions with thin walls of variable sizes on the left side compared to the normal lung parenchyma on the right side; (b) Contrast enhanced coronal computed tomography of the chest showing multiple large cystic areas in the left lower lobe of the lung (arrow). Air fluid levels were not evident.



[Table/Fig-3]: Intraoperative photograph showing large, fleshy, cystic, vascular mass involving left lower lobe.

CCAM is a developmental abnormality of lung development which was originally described by Ch'in KY and Tang MY in 1949 [1]. The three subtypes traditionally associated with this congenital thoracic malformation have now been expanded into 5 types with a revised nomenclature of (CPAM) [2,3]. Majority of cases are diagnosed in the first two years of life. Clinical features include tachypnea, grunting, retraction, and cyanosis [4]. CCAM occurring in an adult is rare and presents as incidental finding or secondary to repeated infection [5]. Only few authors have previously reported CCAM occurring in adult or adolescent patients [5-8]. Differential diagnosis includes congenital pneumonia, pleural effusion, haemothorax, congenital diaphragmatic hernia, pediatric pneumothorax, pneumatocele, sequestration of lung.



[Table/Fig-4]: Photograph of the gross resected specimen showing large widely spaced and irregular cystic structures (arrow).

Healthy individual with recurrent episode of pneumonias in same lobe of lung should be evaluated for acquired and congenital anatomical lesion of lung. Though CCAM is rarely present with adult patient, it should be kept in differential diagnosis of radiologically proven cystic lesion of lung parenchyma, for its small but definitive risk of malignant transformation [9]. Early diagnosis and surgical resection is mainstay of management. Close clinical and radiological follow up required for malignant change even after resection of the lesion.

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