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Giant Pulmonary Bulla with Mediastinal Shift in a 12 ½ year old girl
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Abstract
Pulmonary bulla in children represent interesting entities. Mostly congenital bronchopulmonary foregut malformations and acquired cysts like pneumatoceles have also been described. We present a case of a 12 ½ years old girl with acute onset respiratory distress symptoms harbouring a huge pulmonary cyst exhibiting mass effects, resulting in mediastinal deviation. Following initial workup, cyst excision was carried out which revealed presence of fungal hyphae that was susceptible to Fluconazole therapy post operatively. The case points out how a pulmonary cyst can present in older children with symptoms of respiratory distress and mass effects.

Keywords: Pulmonary cyst/bulla, Giant cyst/bulla, Mediastinal shift.

Introduction
Cystic anomalies of the lung in the paediatric population represent fascinating entities with several differentials. Mainly divided into congenital and acquired lesions,1 their manifestations remain constant with features of respiratory distress and mass effects predominantly. We present an interesting case of a giant pulmonary bulla exhibiting a mass effect in a 12 ½ year old girl.

Case Presentation
A 12 ½ years old girl presented with pain in the left hypochondrium for three days, breathlessness and left sided chest pain for two days with associated pain in left side of the back and left upper limb, especially at elbow region for the same time period. She had no associated complaints of fever, or any other urinary, bowel systemic symptoms. Past medical, family and drug histories were unremarkable. Physical examination at the time of admission revealed her to be alert, oriented and vitally stable. Abdominal examination was positive for tenderness in left hypochondrium and gut sounds were audible. Other systemic examinations were within normal limits. Her initial investigations, which included a complete blood count, electrolytes, SGPT and serum amylase, and an ultrasound of the abdomen, were normal. Chest X-Ray revealed increased lucency over the left hemithorax with visible lung markings and fluid level, suggestive of a large cystic structure, along with mediastinal shift to the right.

Subsequent CT examination showed a giant cyst with fluid level measuring 20.1 x 8.6 x 13.8 cm in lower lobe of the left lung exhibiting mass effects on the surrounding lung parenchyma and mediastinal shift to the right (Figure-1).

Following this, surgical excision of cyst via thoracotomy was planned; however, on the second day of admission, patient became hypotensive with a blood pressure of 85/45, unresponsive to fluid resuscitation, with decreased left sided chest movement and absent breath sounds in the left upper and middle lung zones on auscultation. Therefore, an emergency left posterolateral thoracotomy was done. Intraoperatively, a large cyst was identified arising from the superior/posterior segments of the left lower lobe which contained purulent fluid (Figure-2). Cyst walls were excised, fluid was drained and edges of the lung parenchyma were oversewn. The wound was closed in layers and two chest tubes were placed. Fluid drained was subjected to bacterial and Acid Fast Bacilli culture and cyst was sent for histopathological analysis.

Post-operatively, the patient remained stable. Histopathological analysis, revealed features of chronic granulomatous inflammation and abscess formation along with presence of septate fungal hyphae, on special staining with Periodic acid Schiff (PAS). Therefore, fluconazole 100 mg once daily and ciprofloxacin 250 mg Q12H were initiated to which the patient responded and was subsequently discharged on the fourth post-op day. Medicines were continued for two days.
weeks post-operatively and chest tubes were subsequently removed in the follow-up visits to the clinic. She was followed as outpatient for 2 months and was completely stable.

Discussion

In children, etiology of cystic lesions differs from that in adults, with congenital lesions being more prominent. Congenital lesions, also known as bronchopulmonary foregut malformations, include the discrete entities of pulmonary sequestration, congenital cystic adenomatoid malformation (CCAM), congenital lobar emphysema, and bronchogenic pulmonary cysts.

The patients are known to present with significant respiratory distress and mediastinal shift; however, the larger cysts mostly present in infants or young children due to their obvious compressive effects on the affected lung. Mostly diagnosed with help of chest radiography, CT examination has greatly added to the appropriate diagnosis as well as management of such cysts. The most prominent late complication of these malformations include repeated infections, haemorrhage, erosion into adjacent structures or spontaneous pneumothorax. Surgical removal of the symptomatic cyst is the rule; however, observation is being carried out by pulmonologists in asymptomatic individuals or those with mild symptoms in Congenital Lobar Emphysema on the premise that spontaneous resolution has been documented. Long term follow-up indicates that some compensatory growth does occur after lung resection and this mechanism is more active in infants and children than in older subjects.

In acquired malformations, pneumatoceles and pseudocysts may be inflammatory, as a sequel to pneumonia due to plugging of smaller bronchi and cystic dilatation of the lung parenchyma or trauma. Mostly, they spontaneously resolve and dissipate; however, may enlarge in some cases to cause tension pneumothorax or rupture spontaneously causing pneumothorax. Infective cavities most frequently occur due to bacterial organisms, while a variety of systemic disorders like hyper immunoglobulin E syndrome, hydatid cysts, Langerhans cell histiocytosis, Marfan syndrome, proteus syndrome, and neurofibromatosis may give rise to pulmonary cystic changes.

This case represents how a pulmonary cyst can manifest itself with sudden onset of symptoms in a child, because of subclinical infections in the cyst. The symptoms were not consistent with infectious processes due to absence of fever and leukocyte abnormalities, but were in line with the mass effect caused by the cyst itself. Post-surgical histopathological analysis revealed fungal infection in the pulmonary cyst, which although has been described previously in hydatid cysts and bronchopulmonary malformations, is still a rare entity to be seen clinically. Although the finding of an inflammatory pseudocyst without an obvious or past medical cause has also been reported in literature, it has not been described with an additional fungal pathology of the lungs, since septate fungi like Aspergillus appear in pre-existing pulmonary cavities in their noninvasive forms.

Based on the above experience, surgical excision of such cysts appears to have a favorable prognosis without a need to extend to a lobectomy approach. Hence, if these cysts are found symptomatic, surgical excision should not be delayed so as to avoid decompensation of the patient following mass effects of the cyst.

References