Congenital Pulmonary Cyst

Congenital pulmonary cyst is a rare condition, although intrathoracic cysts are not infrequent in children(1,2). We report a case of congenital pulmonary cyst which simulated tension pneumothorax following air trapping and infection.

An 8-year-old boy developed acute respiratory distress with tachypnea-dyspnea, fever and tachycardia. He was taken to a District Hospital where a chest roentgenogram showed a large cystic space with collapse of right lung. A diagnosis of tension pneumothorax was made and an intercostal drain was inserted anteriorly in the second intercostal space. However, the radiological changes persisted and there was a marginal improvement in the clinical state of the patient. He was then referred to us for further management. There was no air entry in the right hemithorax. X-ray chest showed radiolucent right hemithorax, without any shifting of mediastinum to the left with herniation of the lesion across the anterior mediastinum (Fig. 1A). A diagnosis of congenital pulmonary cyst was made and pre-operative lung functions (bed side tests, acid-base studies and blood gas analysis) showed compromise of function of moderate degree. A standard right posterolateral thoracotomy revealed a cyst of the lung arising from the periphery of the lower lobe. Pulmonary cystectomy was performed and the child made an uneventful recovery with expansion of lung (Fig. 1B). The histology of the excised cyst wall showed tall ciliated columnar epithelium, and secondary infection with squamous changes at other sites. He is asymptomatic and with normal pulmonary functions at 2 year follow-up.

Congenital pulmonary cysts are considered to arise as a result of abnormal bronchial budding at relatively distal levels of the tracheo-bronchial tree(3). These cysts, which communicate with the bronchial tree, can be unilocular or multi-locular and are lined with respiratory epithelium. Secondary infection can destroy the columnar epithelium making pathological differentiation from empyema or lung abscess impossible. They usually produce signs early in life, either by enlarging rapidly as a result of air trapping within the cyst or more frequently, by becoming secondarily infected. Communication of the cyst with the bronchial tree and secondary infection may result in rapid expansion of the cyst because of a ball valve obstruction in the narrow communicating channel(5). Radiological features depend on the extent and site of the lesion; the chest X-ray is an important investigation for diagnosis. More recently ultrasound, computed tomography and magnetic resonance imaging are used.
increasingly to accurately diagnose the condition. The treatment consists of pulmonary cystectomy or segmental resection in older child, but lobectomy is the treatment of choice in the neonates. Lobectomy is well tolerated and complication rate following segmental resection is unacceptably high(1,4,5). In the present case, intercostal drainage failed to result in a collapse of the cyst (a characteristic feature of congenital pulmonary cyst). However, in the presence of dangerously over-expanded cyst, intercostal drainage may be useful to gain time for pre-operative preparation as done in our case.

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REFERENCES


Fig. 1 A. Preoperative X-ray chest showing air trapping and herniation of the cyst with collapsed right lung. B: Satisfactory expansion of right lung following pulmonary cystectomy.