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Case Report

Pulmonary Aplasia: A Case Report

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Abstract

Developmental malformations of the lung are common but aplasia of the lung is extremely rare and usually the patients presented in early childhood with recurrent lower respiratory tract infection and respiratory distress. A 2 years and 6 months old girl presented with complaints of respiratory distress since birth and recurrent episode of cough since six month of age ,with opacity of the right hemithorax on X-ray, CT scans shows aplasia of the right lung. Patient was managed conservatively and his parents were counseled about the anomaly. The child is doing well and is in a regular follow-up with us. Aplasia of the lung should be suspected in children with recurrent respiratory distress with opacity of the hemithorax on X-ray and herniation of the opposite lung across the mediastinum.

Keywords: Aplasia, agenesis, congenital, opaque hemithorax.

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Introduction

Pulmonary agenesis means complete absence of a lung. Agenesis differs from aplasia by the absence of a bronchial stump or carina that is seen in aplasia. Bilateral pulmonary agenesis is incompatible with life, manifesting as severe respiratory distress.[1] Unilateral aplasia can have few symptoms and nonspecific findings. Symptoms tend to be related to associated central airway complications of compression, stenosis or tracheobronchomalacia. Pulmonary agenesis is thought to be an autosomal recessive trait. Lung agenesis or aplasia is a very rare and exact incidence is not known. The prevalence of agenesis of lung is reported to be 34 per 1,000,000 live births.[2] Pulmonary agenesis can occur between the 4th and 5th week of gestation in the embryonic phase, before the pseudoglandular period, when primitive lung is forming as a diverticulum protruding from the foregut. The etiology is suggested to be related to duplication of the distal part of the upper arm of chromosome 7.[3] Bilateral lung agenesis is life threatening and about 50% patients with unilateral lung agenesis die within 5 years of life, with left lung agenesis having better prognosis than the right one. However, some reports have shown patients with right lung agenesis living upto old age because of absence of critical anomalies in other organs.[4,5] Pulmonary agenesis is often seen in association with other congenital anomalies such as the VACTERL sequence.[6,7,8] CT scan of chest is diagnostic.[9,10,11] Conservative treatment is usually recommended, although surgery has offered.

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Because of rarity of this clinical entity we are interested to report such a case who was admitted in Rajshahi Medical College Hospital, Rajshahi, Bangladesh.

Case Report

A 2 years and 6 months old girl was admitted in Rajshahi Medical College Hospital on April 2017 with complaints of respiratory distress since birth and recurrent episode of cough since six month of age. She had respiratory distress not associated with bluish coloration of lip, tongue and nail bed. She has no history of choking, foreign body impaction, contact with TB patient. Cough not aggravated after feeding. For this she was diagnosed as pneumonia by local doctor and treated with several course of i/m, oral antibiotics with mild improvement of cough for a time being. She was born by NVD at term. Immunized as per EPI schedule, development was age appropriate. She was the only issue of lower class non-consanguineous parents. On general physical examination she was ill looking, dyspnoeic, no cyanosis, clubbing, BCG mark was present, lymph nodes not enlarged, neck vein not engorged. ENT examination was normal, HR 120/min, respiratory rate 66 breaths/min, BP 80/50 mm of Hg temperature101°, height 84 cm (above 10th centile), weight 11 kg (on 5th centile), OFC 47 cm (above10th centile) WHZ score- 1.76 SD. Respiratory system examination revealed drooping of the right shoulder, reduced chest movement on right side, chest indrawing present, there was no visible apical impulse, trachea sifted to right side, apex beat located in right 4th ICS just medial to the MCL, chest expansibility reduced on right side, vocal fremitus absent on right side, percussion note dull in all areas of right lung field, on auscultation breath sound was absent on right side. There was crepitation in left mid zone. Cardiovascular system examination revealed apex beat on right 4th inter costal space, medial to mid clavicular line. Other system examination showed no abnormality

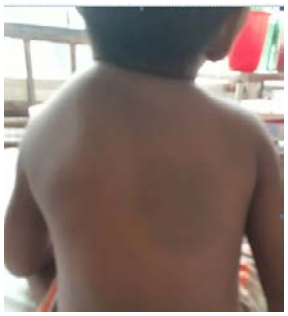

Fig-1: A 2 years and 6 months old girl with right sided lung aplasia showing drooping of right solder.

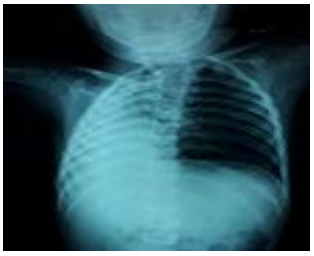

Fig-2: CXR AP view Showing hyper translucency of left learn trachea shift to right, homogenous opacity on right lung field, no lung marking in on the right side.

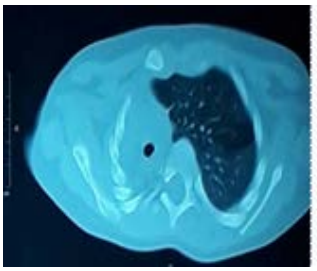

Fig-3. HRCT- shows right lung tissue is not visualized, gross ipsilateral shifting of mediastinum with herniation of the contra-lateral lung,right principal bronchus was partially visualized.

WBC- 15000/cu mm, N- 35%, L -62%, E-02%, M-01%, B-00%, Hb 11.40gm/dl, ESR 08mm in 1st hour. PBF- leucocytosis with increase roulaux formation., MT-negative. echocardiogram: dextroversion with normal 2D echo**.** HRCT-showed right lung tissue is not visualized, gross ipsilateral shifting of mediastinum with herniation of the contra-lateral lung, right principal bronchus

was partially visualized, left lung was hyperinflated suggesting aplasia of right lung with gross ipsilateral shifting of mediastinum structures and herniation of the contra lateral lung associated compensatory hyper inflated left lung. All these clinical and radiological finding we diagnosed as aplasia of right lung with bronchopneumonia. Parents were properly counselled about the disease. The patient was treated with i.v ceftriaxone and flucloxacilline for 10 days. She became afebrile after four days, respiratory distress improved. We discharged the patient after ten days.

Discussion

Pulmonary aplasia has been reported at different ages newborn, infants, children and adults, even at 90 years of age.[12] More common are unilateral aplasia which may has few symptoms and non-specific findings, among which only one-third are diagnosed during life.[13] Embryologically, these malformations correspond to a failure of development of the respiratory system from the foregut. Arrest at the stage of the primitive lung bud produces bilateral pulmonary agenesis. The respiratory anlage at a later stage may develop only unilaterally and lead to lung agenesis. Lobar agenesis results when developmental arrest on one side occurs in an older embryo. Pulmonary hypoplasia may occur during the last trimester of pregnancy with failure of final alveolar differentiation.[14] They are generally sporadic, with only a few reports of these conditions occurring in siblings in an autosomal recessive pattern. They occur with equal frequency in both sexes and involve both lungs equally. There is high incidence (>50.0%) of associated cardiac, gastrointestinal, genitourinary, skeletal, central nervous system malformations and VACTERL sequence. Diagnosis should be suspected when respiratory difficulty occurs with tracheal deviation, in the presence of clinically symmetric chest and Chest X-ray suggestive of massive atelectasis with mediastinal shift.[2] No treatment is required in asymptomatic cases. Treatment is necessary for lower respiratory tract infections. Patients having stumps may require surgical removal of the stump if postural drainage and antibiotics fail to resolve the infection.[9] Corrective surgery of associated congenital anomalies, wherever feasible, may be undertaken. Prognosis depends on two factors. Firstly, the Severity of associated congenital anomalies and secondly, involvement of the normal lung in any diseaseprocess.[15] If patient survives the first five years without major infection, an almost normal life span can be expected.[16]

Conclusion:

Pulmonary aplasia is very rare condition which should be suspected in children with recurrent respiratory distress with opacity of the hemithorax on X-ray.

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