Isolated Left Lung Aplasia with Bronchial Asthma

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Congenital lung anomalies are categorised as pulmonary agenesis, aplasia and hypoplasia with distinct clinical implications. An 8-year-old boy was referred for an “opaque left hemithorax” for which he had received antituberculous therapy. A detailed evaluation including flowing contrast computed tomography of the thorax and fiberoptic bronchoscopy led to a diagnosis of left lung aplasia. He also had wheezing dyspnea, which was confirmed as bronchial asthma. Congenital lung defects with associated asthma was reported only twice till date. A high index of suspicion is required to recognise such a patient.

Key words: Bronchial asthma, Congenital lung anomalies, Lung aplasia.

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Congenital malformations of the lung are rare disorders occurring with variable degree of severity. These are the result of insult to the developing embryo during the fourth and fifth weeks of intrauterine life(1). Boyden clearly categorised these congenital anomalies as pul-monary agenesis, aplasia and hypoplasia(2). This categorisation is widely accepted as each condition has distinct and important clinical implications. The clinical presentation being variable, diagnostic errors often occur.

Although congenital lung anomalies were sporadically documented from the sub-continent(3-7), the occurrence of asthma in such patients is extremely rare. This association was reported only twice before, both of whom were adults when documented with pulmonary agenesis and associated asthma(7,8). The paucity of such a report in children in the literature prompted this description of an 8-year-old boy with pulmonary aplasia who also had asthma.

Case Report

An 8-year-old boy was referred to our institute for evaluation of a left-sided “opaque hemithorax”. Since early childhood, he had experienced paroxysmal wheezing dyspnea along with dry cough, which had aggravated during change of season. However, there were no associated nasal symptoms. He was the
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first-born child of a non-consanguineous marriage with uneventful antenatal and postnatal periods. He had normal milestones and had received complete immunisation. A family history of atopy was elicited. His younger brother aged 5-years had a history of repeated colds suggestive of allergic rhinitis. There was no family history of congenital anomalies. A year prior to referral, a chest radiograph done for these complaints showed a left opaque hemithorax. This resulted in the patient being prescribed antituberculous therapy for six months but without relief.

Physical examination revealed a boy in no acute distress. There was no clubbing or cyanosis. Flattening of the chest was observed on the left side along with an ipsilateral shift of the mediastinum. Chest excursion too was reduced on the same side. Normal vesicular breath sounds were audible on the right side along with polyphonic rhonchi, but the intensity of breath sounds on the left side was vastly reduced. Heart sounds were normal in character and intensity.

A review of three chest roentgenograms done over a period of 6 months demonstrated an opaque left hemithorax with ipsilateral shift of mediastinum along with herniation of the right lung (Fig.1). There was no change in the radiological picture. The complete hemogram was normal. Spirometry showed a forced vital capacity of 1.1L (80% of predicted), and forced expiratory time in one second (FEV₁) of 870 mL (70% of predicted). There was a significant reversibility (210 mL) in FEV₁ after inhaled salbutamol. Fiberoptic bronchoscopy visualised a short left main bronchus, which ended in a blind pouch. The tracheobronchial tree on the right side was normal. Electrocardiogram showed low voltage normal waves with clockwise rotation of the heart. An echocardiogram demonstrated a normal pulmonary artery pressure of 20 mmHg with normal lumen; however, the bifurcation of pulmonary artery could not be seen. Atrial and ventricular functions were essentially normal. Serial sections of computed tomography (CT) of the thorax revealed a short left main bronchus with an absent left lung. CT-thorax with flowing contrast, confirmed an absent left pulmonary trunk with a normal right pulmonary artery (Fig. 2). Preaortic and precardiac herniation of the normal lung with left and posterior displacement of the heart was also seen. A skeletal survey and an ultrasonographic examination of the abdomen were normal. A diagnosis of left lung aplasia with bronchial asthma was made. The patient’s symptoms of wheezing dyspnea and dry cough were abolished after initiation of combination of inhaled long acting bronchodilator and steroid.

![Fig. 1. Chest roentgenogram (posteroanterior view) showing an opaque left hemithorax with ipsilateral shift of mediastinum and herniation of the right lung.](image)
This was given in the form of a dry powder capsule (formetrol 6 µg and budesonide 100 µg inhaled twice a day through a device (Rotahaler®)).

**Discussion**

The association of congenital lung anomalies with asthma is yet to be highlighted. An extensive search of the literature revealed only two reports (7,8) of lung agenesis with associated asthma. Both these reports are of recent origin. The first was a 23-year-old white woman in whom right-sided pulmonary agenesis was detected at 6 months of age when she presented with an upper respiratory tract infection. Symptoms of asthma and allergic rhinitis were present since early childhood. For these symptoms she had received 34 months of immunotherapy during her college days. Her father too had asthma and allergic rhinitis(8). The second was a 35-year-old Indian male who was referred for evaluation of a left ‘opaque hemithorax’ that led to a diagnosis of agenesis of left lung along with Klippel-Feil syndrome and an absent gall bladder. On evaluation, a history suggestive of asthma and allergic rhinitis was also elicited. He had an atopic background as his mother and sister also had asthma and allergic rhinitis. His spirometry demonstrated an improvement of 230 mL in FEV$_1$ after administration of 200 µg of inhaled salbutamol. His symptoms were abolished after receiving a combination of inhaled long acting bronchodilator and steroid(7). Our patient, who had an isolated left lung aplasia, too had a remarkable response after initiation of a similar inhaled combination. To our knowledge, this is the first case of asthma with isolated left lung aplasia reported in the paediatric age group from India.

Associated congenital malformations are present in almost half of the patients with congenital lung anomalies(9). These associated systemic congenital defects are a major cause of morbidity and mortality. They are more commonly seen with right-sided malformations while left-sided defects are
usually isolated(9). Right-sided congenital lung defects are reported to have mortality twice that of the left side(10). This is attributed to a greater mediastinal shift towards the right side, leading to distortion and malrotation of the main bronchus with great vessel disturbances.

Non-invasive imaging techniques have now emerged as the diagnostic modality of choice, especially CT-thorax. It helps to delineate pulmonary parenchyma, pulmonary vasculature and the bronchus, while CT-angiography and echocardiography detail the pulmonary vasculature. With the advent of these techniques, the exact diagnosis can be established without opting for invasive procedures like bronchography and pulmonary angiography(4). In our patient, contrast enhanced CT-thorax showed a short left main bronchus, an absent left lung as well as compensatory overinflation and herniation of the normal right lung. Both CT-angiography and echocardiography demonstrated complete absence of left pulmonary trunk. This confirmed the diagnosis of left lung aplasia.

Most patients present with repeated chest infections and, radiologically, an opaque hemithorax. In our country, an opaque hemithorax is often mistaken for fibrotic lung disease subsequent to pulmonary tuberculosis. This often results in patients erroneously receiving antituberculous therapy, as was seen in our patient. The cough and wheezing dyspnea are usually thought to be due to obstructive airways disease caused by distorted bronchi.

Our report highlights the fact that a high index of suspicion is required to diagnose congenital lung anomalies, and that asthma can occur in such patients. Appropriate anti-asthma therapy can alleviate symptoms and reduce morbidity.

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