Unusual association of diseases/symptoms

Complete right lung agenesis presenting with bronchial asthma and allergic rhinitis

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Summary

A 26 year-old lady presented with episodic breathlessness, chest tightness, recurrent nasal obstruction and excessive sneezing, mainly during change of season along with opacity of the right hemithorax on chest x-ray. Further detailed work-up including spirometry, high-resolution CT scan of the thorax and fibre-optic bronchoscopy confirmed complete right lung agenesis in patients with bronchial asthma and allergic rhinitis. Complete control of symptoms was achieved with formeterol 6 µg and mometasone 200 µg (via dry powder inhaler) and intranasal fluticasone 50 µg (nasal spray) 2 puffs twice daily and oral montelukast 10 mg with levocetirizine 5 mg once daily.

BACKGROUND

Unilateral lung agenesis is extremely rare. These patients usually present in infancy or childhood with recurrent respiratory infection and cardiopulmonary insufficiency. Here, we report a case of bronchial asthma and allergic rhinitis whose investigation showed complete right lung agenesis.

CASE PRESENTATION

A 26-year-old lady presented to our department with episodic breathlessness, chest tightness and recurrent nasal obstruction and sneezing since 15 years of age. She revealed that her symptoms occurred mostly during change of season. Her mother was a known asthmatic. She was born out of non-consanguineous marriage with normal growth and developmental milestones. No other significant history was obtained. General physical examination revealed tachycardia (114 beats per minute) and tachypnoea (19 cycles per minute) with no other abnormalities. Respiratory system examination showed reduced chest movement on right side and trachea deviated slightly to right. Dull note was heard on percussion in anterior lung field on the right side. Heart sounds were heard in the right parasternal region. Breath sounds were diminished on the right side and diffuse polyphonic rhochi heard all over the left hemithorax.

INVESTIGATIONS

Chest x-ray (CXR) posterior–anterior (PA) view (figure 1) showed homogeneous opacity with signs of volume loss on the right side. CT scan of the thorax (figures 2 and 3) showed trachea continuing beyond T4 vertebral level and dividing into left lobar bronchi at a lower level. CT scan images (figures 4 and 5) showed absent right lung with hyperinflation and herniation of the left lung. Fibre-optic bronchoscopy showed constriction at the T4 vertebral level where normally trachea divides into right and left main bronchi (figure 6) and division of trachea into left lobar bronchi at a lower level (figure 7). Spirometry

showed that forced vital capacity was 1.22 l (46% of predicted), prebronchodilator forced expiratory volume in 1 second (FEV₁) was 760 ml (31% of predicted) and post-bronchodilator FEV₁ after four puffs of 200 µg of salbutamol was 1040 ml (44% of predicted), post bronchodilator change in FEV₁ was 280 ml and 13% indicating positive acute bronchodilator response and hence confirming bronchial asthma. Cardiac evaluation and skeletal survey showed no abnormality.

OUTCOME AND FOLLOW-UP

The patient was completely asymptomatic when reviewed after 1 month and she was advised to stop

Figure 1 Chest x-ray posterior–anterior view showing right-sided opacity with signs of volume loss.

Figure 2 HRCT thorax showing absent right lung with mediastinal shift to right and division of trachea directly into left lobar bronchi.

intranasal fluticasone and to continue formeterol and mometasone inhaler with oral montelukast and levocetirizine preparation.

DISCUSSION

Congenital anomalies of the lung are rare disorders. They occur during 4th and 5th weeks of intrauterine life.¹ Genetic, teratogenic, mechanical factors, viral infections and vitamin A deficiency are possible aetiologies.² Developmental pulmonary anomalies have been classified by Schneider and Schwalbe³ into three groups;

1. Agenesis—both lung and bronchus absent.
2. Aplasia—rudimentary bronchus present but limited to blind pouch without lung parenchyma.
3. Hypoplasia—hypoplastic bronchus with variable of lung parenchyma is present.

Unilateral lung agenesis was first described by De Pozze in 1673 during autopsy of an adult female. Lung agenesis has no sex predilection. Parental consanguinity through an autosomal recessive mode of inheritance may be a risk factor.⁴ Fifty per cent of children born with lung agenesis

Figure 4 HRCT thorax showing absent right lung with complete shift of mediastinum into the right hemi thorax.

are stillborn or die within few days due to concomitant anomalies^{5 6} and associated congenital anomalies occur in about 50% of the cases. However, there are reports of people with lung agenesis living well over 40 years of age.^{7 8} Left lung agenesis is more common and has a longer life expectancy. Right lung agenesis is associated with high mortality due to increased risk of respiratory infections, which is attributed to greater mediastinal shift that leads to distortion and malrotation of the main bronchus and the great vessels obstructing the drainage of functioning lung.^{9 10} Rarely can lung agenesis remain asymptomatic and may be diagnosed incidentally when patients present with symptoms of other illness.^{9 10} In our case, the patient presented with bronchial asthma and allergic rhinitis.

Clinically, the condition is indistinguishable from lung collapse. Other differential diagnoses include diaphragmatic hernia, adenoid cystic malformation and intrathoracic lobar sequestration. However, in the presence of opaque hemithorax with intact diaphragm lung agenesis should always be considered. CXR PA view may initially

Figure 3 HRCT thorax showing division of trachea into left lobar bronchi (black arrow).

Figure 5 HRCT thorax showing herniation of mediastinum and left lung into right hemi thorax.

Figure 6 Showing a constriction ring (double arrow) and no division of trachea into right and left main bronchus at T4 vertebral level.

Figure 7 Showing trachea dividing at a lower level into left lobar bronchi.

suggest the diagnosis. CT of the thorax very well delineates pulmonary vasculature, parenchyma and bronchial structures. Hence, it has been regarded as an optimal

non-invasive investigation for diagnosis. Fibre-optic bronchoscopy allows direct visualisation of bronchial tree and confirms the diagnosis.¹¹ The best method to diagnose lung agenesis is pulmonary angiogram. In our case, high-resolution CT of the thorax and bronchoscopy confirmed the diagnosis.

Thus, we conclude that a high index of suspicion is necessary to diagnose the condition. Simple treatment of asthma with usual doses of inhaled medication can alleviate the symptoms.

Learning points

- ▶ Congenital anomalies of the lung are rare events. Right lung agenesis is associated with more complications than left side.
- ▶ Patients present with recurrent respiratory illness or due to complications during infancy or childhood. Rarely remain asymptomatic till adulthood.
- ▶ Opaque hemithorax is not always synonymous with destroyed lung of tuberculosis even in high burden countries.
- ▶ Regular asthma medications are good enough to relieve symptoms even in patients with unilateral lung agenesis for control of symptoms.

Competing interests None.

Patient consent Obtained.

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