CASE REPORT

Unilateral pulmonary hypoplasia: a rare clinical presentation

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SUMMARY

The clinical presentation of an adolescent man with left lung hypoplasia is described in detail including the history and clinical examination findings. On further investigation, the chest x-ray and CT scan of the chest reveal interesting findings. Generally, such conditions are brought to notice in the early years of life but the presentation of this condition at the age of 18 years make it even more interesting and unique. This is a discussion of the management of such a patient and the different types of lung hypoplasia which can be encountered in clinical practice.

BACKGROUND

Although many abnormalities of the lung and bronchial tree have been described (eg, blind-ending trachea with absence of lungs and agenesis/aplasia of one lung), most of these gross abnormalities are rare.¹ In aplasia of trachea, there is a rudimentary airway stump, but absence of any distal lung or a hypoplastic lung. Tracheal agenesis or aplasia is often associated with other ipsilateral congenital malformations.² Congenital malformations associated with pulmonary hypoplasia may be present in any system. Central nervous system (anencephaly or hydrocephaly),³ cardiovascular system (Tetralogy of Fallot),⁴ diaphragm, musculoskeletal systems are a few of them. Pulmonary hypoplasia at birth is often associated with severe congenital urinary obstruction and oligohydramnios, for example, Potter's syndrome.²

CASE PRESENTATION

An 18-year-old boy presented to the medical clinic with exertional chest pain, dyspnoea during sleep, cough with expectoration and intermittent fever for 5 days. He was a labourer and came from Jharkhand. He gave a history of antitubercular therapy taken for around 3 months. There was no history of any diurnal variation in fever, chills/rigors associated, haemoptysis or anxiety/palpitations.

General examination revealed mild pallor and a thin built. On respiratory system examination, mild scoliosis was present in chest towards the left side and drooping of the left shoulder (figure 1A,B).

On inspiration, chest expansion was asymmetrical with reduced expansion on left side. On palpation, apex beat was found to be markedly shifted to the left side to lie in the mid-axillary line in fifth intercostal space, trachea was shifted to the left side, vocal fremitus was increased. On percussion, the note was resonant over the left precordial region and dull in left axillary region. On auscultation,

bronchial breathing was heard over the left lung in the axillary and infrascapular region. Other systems were normal except that liver dullness was pushed downwards to be present at sixth intercostal space.

From the history and examination, it was found that there was some degree of consolidation in the left lung with mediastinal shift to the left side.

INVESTIGATIONS

Haemogram and ELISA for HIV was normal. ECG revealed no abnormality. Sputum staining (Gram's stain and Ziehl-Neelsen stain) showed only Gram-positive cocci in pairs and no acid-fast bacilli were found. MGIT (mycobacterium growth indicator tube) also revealed absence of acid-fast bacilli. Chest x-ray (figure 2) revealed markedly shifted trachea (towards left), crowding of ribs on the left side and compensatory hyperplasia of the right lung.

CT scan (figure 3) of the chest revealed complete collapse of the left lung with loss of volume of hemithorax, fibronodular lesions in the right lung with 'tree in bud appearance' on the right. There was compensatory emphysema of the right lung and a marked mediastinal shift to the left.

Ultrasound of the whole abdomen was also normal.

DIFFERENTIAL DIAGNOSIS

Initially, from the history and clinical examination, tuberculous fibrosis, Swyer-James syndrome and pulmonary hypoplasia were kept as differentials. The absence of any radiological findings excluded the likelihood of tuberculous fibrosis. In Swyer-James syndrome, the affected lung parenchyma appears hyperlucent, a result of air trapping owing to bronchiolar obstruction and blood vessels within the affected lung parenchyma are decreased in number and calibre. It is excluded from the

Figure 1 (A) Mild scoliosis (B) drooping of left shoulder.

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Figure 2 Chest x-ray.

radiological findings of the CT scan of the patient as it is the left lung which is affected here but the normal right lung is hyperlucent (probably owing to compensatory emphysema) and blood vessels are also normal in the right lung as seen on contrast-enhanced CT scan. The radiological findings of x-ray and CT scan indicated towards pulmonary hypoplasia since birth.

TREATMENT

The supportive treatment was given to the patient. As per the sensitivity report of the culture-sensitivity investigation, amoxicillin was administered orally for 5 days. Symptomatic treatment was also given for other symptoms.

During follow-up, his condition improved. He was recommended proper dietary intake of nutrients. He was also advised to go for surgical referral but owing to his poor socioeconomic status, he refused to undergo the surgery for his condition.

DISCUSSION

Bronchial buds are formed at fifth week of intrauterine life and all the 17 segments are formed by the end of 6 months.¹ The complete development of the bronchial tree takes place during the initial days of postnatal life. Pulmonary hypoplasia is characterised by reduced and/or less-developed lung tissue to varying

Table 1 Categories of developmental disorders of lung

Group I	No bifurcation of trachea
Group II	Only rudimentary main bronchus
Group III	Uncompleted development after main bronchus bifurcation
Group IV	Incomplete development of small segment bronchi of any lobe

degrees.^{5 6} In cases of premature rupture of membranes at 15–28 weeks' gestation, the reported incidence of pulmonary hypoplasia ranges from 9% to 28% (13% in most studies). Death rates of 71–95% have been reported during the perinatal period in patients with pulmonary hypoplasia.⁷ According to Boyden,⁸ developmental disorders of lungs can be found to be either of the following three:

1. Agenesis (complete absence of the lung tissue);
2. Aplasia (no lung tissue, but there is a rudimentary bronchus);
3. Hypoplasia (all lung tissues exist but underdeveloped).

However, Monaldi⁹ has categorised developmental disorders of the lung into four categories (table 1).

The patient presented here belongs to group III according to Monaldi and third category according to Boyden.

There are primary (idiopathic) and secondary causes in the aetiology of the pulmonary hypoplasia. It usually occurs secondary to the intrauterine diseases impairing normal lung development. Deformities of the chest wall, pleural effusion associated with hydrops fetalis, cystic adenomatoid malformation and diaphragmatic hernia physically limits the development of the lung.¹⁰ Furthermore, conditions leading to oligohydramnios such as early rupture of the membranes or fetal renal insufficiency also impair development of the lungs.¹¹ Hypoplasia may be bilateral in thoracic dystrophies and oligohydramnios. In such cases, development of the airways and arterial truncation is inhibited and thus, surface area required for alveolar–capillary gas exchange becomes limited.¹¹

Clinical course (severity of symptoms and their time of occurrence) is decided by the degree of hypoplasia present in the patient.¹² The child may present with respiratory distress at birth or a reduced Apgar score requiring mechanical ventilation but babies with normal Apgar scores have also been seen.^{13 14} As with our patient, some of the patients have a silent childhood, the disease being manifested only at adulthood.⁶

In most cases, the contralateral lung is enlarged, with a greater number of alveoli, but has a normal bronchial pattern. Individuals with aplasia and hypoplasia may present with recurrent infection, dyspnoea and reduced exercise capacity, and there may be pooling of bronchial secretions, with secondary infection and overspill of infected secretions into the

Figure 3 CT scan of the chest (A) non-contrast and (B) contrast.

contralateral normal lung.² On the same grounds, our patient also presented to us with an acute illness, ie respiratory infection that is non-tubercular. The fibronodular lesions in the CT scan also suggests some degree of infectious pathology in this case.

Learning points

- ▶ Unilateral pulmonary hypoplasia can present in late adolescent life as an infective pathology when pulmonary function is decompensated.
- ▶ Totally shifted mediastinum with congested ribs and absence of breath sounds on affected side gives a hint towards this condition upon clinical examination.
- ▶ Such cases need to be immediately managed to prevent any fibrosis following infection in the only functional lung.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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