

Asymptomatic Unilateral Pulmonary Hypoplasia/Agenesis in Adults: A Report of Six Cases

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ABSTRACT

Bilateral pulmonary agenesis is incompatible with extrauterine life. Unilateral pulmonary hypoplasia or agenesis is rare, the vast majority of cases presenting in the newborn, infancy, or early childhood, with respiratory symptoms. Pulmonary hypoplasia or agenesis presenting in adults is extremely rare, and the initial chest radiographs may present significant interpretation challenges. We report six patients encountered incidentally in a 10-year period, during which approximately 400,000 routine adult chest radiographs were reported. In all six cases, the chest X-ray films were equivocal, and multidetector computed tomography (CT) was required to settle the diagnosis. The cases are presented to draw the attention of radiologists and physicians to this rarity and to avert the kind of initial errors that we made in interpreting their chest radiographs. The invaluable role of multidetector CT in resolving this potential confusion is emphasized. A brief review of the relevant literature is undertaken.

Key words: Adult chest; chest X-ray; computed tomography; congenital pulmonary anomalies

Introduction

The chest X-ray film remains the most common radiographic examination requested as a part of routine medical fitness assessment, in particular, to exclude communicable diseases such as pulmonary tuberculosis. We report approximately 150 routine adult chest radiographs daily, performed for preemployment, visa, military service, or school admission purposes.

All cases with equivocal chest X-ray findings were subjected to further imaging including computed tomography (CT).

Of the six cases which turned out to have pulmonary hypoplasia or agenesis, their chest X-ray films mimicked confusing features such as lobar collapse, pleural effusion, hydatid disease, and post tuberculous destroyed lung. Multi-detector CT, with coronal and sagittal reformation, was critical to making the correct diagnosis in each case.

Case Reports

Case 1

A 19-year-old male had a chest X-ray done for visa purposes. It showed a large, rounded mass over the right hemidiaphragm [Cases 1 and 2]. In view of the patient's country of origin and being from a sheep-rearing community, the possibility of a right basal pulmonary hydatid cyst was considered. He had no significant past or current health issues.

Contrast-enhanced CT of the chest and abdomen [Case 3a and b] confirmed a large, almost pedunculated, herniation of the liver into the thorax, presumably resulting from a hypoplastic right lower lobe.

Case 2

An asymptomatic 28-year-old male had a preemployment chest X-ray which showed a left pleural opacity with a positive

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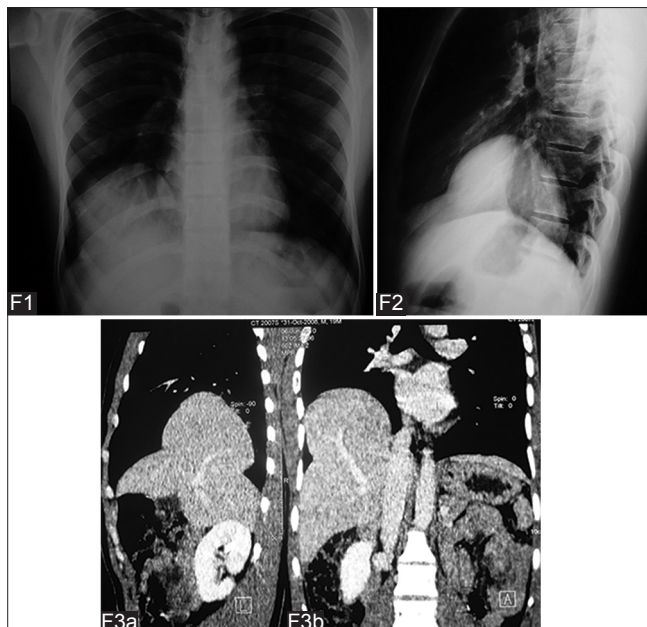
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meniscus sign, suggesting pleural effusion [Case 1]. This was accompanied by typical layering on the lateral decubitus film [Case 2], as expected of pleural fluid.

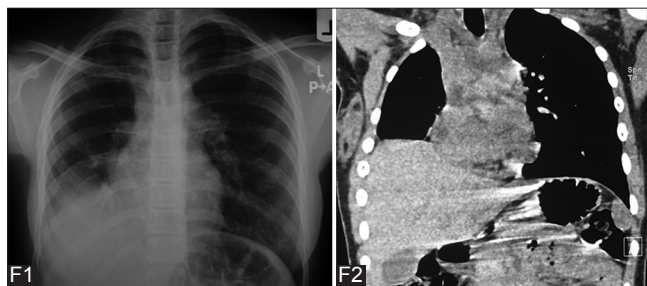
Two repeated attempts at percutaneous diagnostic pleural aspiration yielded no fluid. A subsequent ultrasound study also demonstrated no fluid. CT of the chest and upper abdomen [Case 3] demonstrated replacement of most of the left lung by herniated omentum and intestinal loops, and no pleural fluid. A diagnosis of left lung hypoplasia was made.

Case 3

A 13-year-old girl had a routine pre-high school chest X-ray. The film showed opacification of the right base with



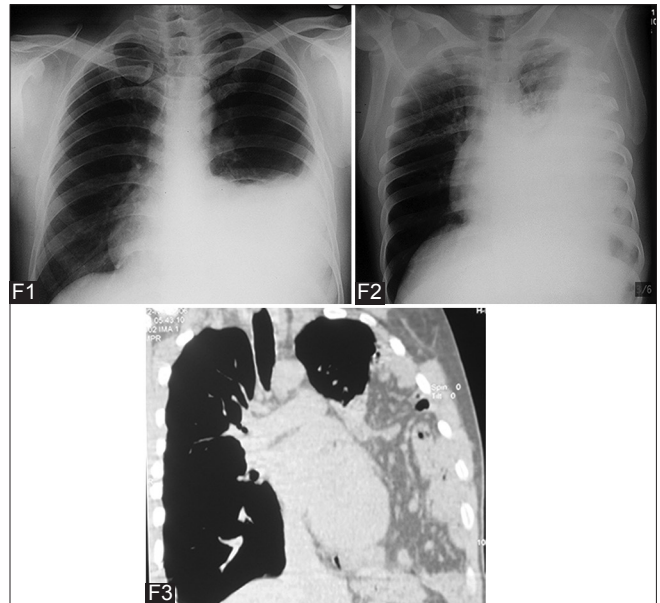
Case 1: Fig. 1: 19-year-old male: routine posterior-anterior chest X-ray showing a rounded mass lesion in the right lower chest. Note obliteration of the right hemi-diaphragm. Fig. 2: Right lateral chest film, showing the right base mass with smooth outlines. Fig. 3a&b: Contrast computed tomography chest, with sagittal and coronal reformatted images. The liver dome is herniated into the thorax. Note the portal vein extending into the herniated segment



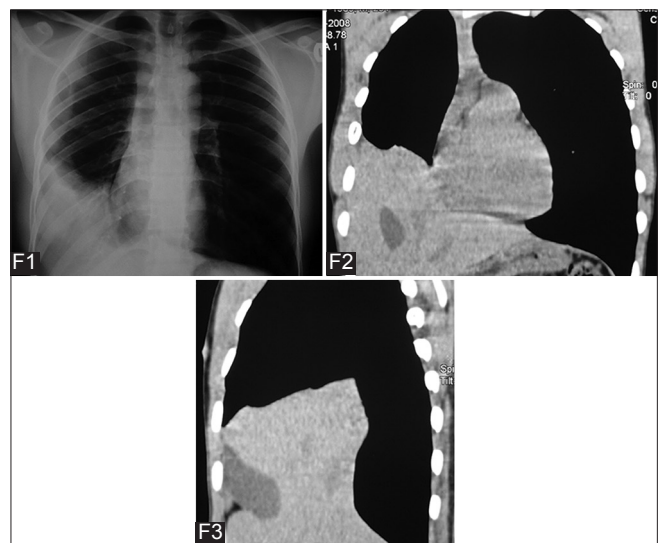
Case 3: Fig.1: 13-year-old girl: routine posterior-anterior chest radiograph shows obliteration of the right hemi-diaphragm and slight cardiac shift to the right. Right lower lobe collapse was considered a possibility. Fig.2: Coronal reformatted computed tomography chest shows the liver markedly elevated into the right chest, with cardiac shift to the right

obliteration of the right hemidiaphragm and slight shift of the heart to the right [Case 1]. The features suggested right lower lobe collapse/consolidation, but she had no symptoms and demonstrated no clinical signs.

CT of the chest showed elevation of the liver into the right hemithorax and cardiac shift to the right [Case 2]. There was



Case 2: Fig.1: 28-year-old male: routine posterior-anterior chest X-ray showing an opacity in the left base, with a positive meniscus sign, suggesting pleural fluid. Fig. 2: Left lateral decubitus film shows layering of the pleural "fluid". Fig.3: Coronal reformatted computed tomography chest shows the left hemi-thorax occupied by omentum and loops of bowel, sparing the left upper lobe



Case 4: Fig.1: 25-year-old male, routine chest radiograph showing a right basal opacity with obliteration of the right hemi-diaphragm and cardiac shift to the right. Fig. 2: Coronal reformatted computed tomography chest shows elevation of a deformed liver into the right thorax. Fig. 3: Sagittal reformatted computed tomography chest, showing the elevated liver with deformity of its fundus. Note the unusual location of the gallbladder

no collapse or consolidation. The changes were considered as due to the right lower lobe agenesis or segmental hypoplasia.

Case 4

A 25-year-old male had a preemployment chest X-ray which showed a poorly defined, elevated right hemidiaphragm and cardiac shift to the right [Case 1]. A right lower lobe collapse was considered a possibility although the patient exhibited neither symptoms nor clinical signs.

A CT study [Cases 2 and 3] revealed that the liver dome was deformed and elevated into the lower chest. The gallbladder was malpositioned. Since there was no history of previous chest trauma or significant chest disease, the changes were considered due to segmental right pulmonary hypoplasia.

Case 5

A 28-year-old female had a preemployment chest X-ray. The chest film [Case 1] showed a marked mediastinal shift to the left and a largely opaque hemithorax. Although a destroyed left lung from old TB was considered, the patient denied any history of significant chest disease.

Chest CT showed a hypoplastic left bronchus and lung [Case 2]. The right lung showed compensatory hypertrophy, extending across the midline and occupying most of the left hemithorax.

Case 6

A 32-year-old male had a preemployment chest X-ray which showed a largely opaque left hemi thorax, with severe deviation of the mediastinal structures to the left [Case 1]. He denied any significant chest symptoms.

CT of the chest showed the complete absence of the left lung, with compensatory hypertrophy of the right lung, extending to occupy the left hemithorax [Cases 2 and 3]. The left bronchus was completely absent.



Case 5: Fig. 1: 28-year-old female with routine posterior-anterior chest radiograph showing severe mediastinal shift to the left. The right lung is hyper-inflated with a flattened hemidiaphragm. Fig.2: Axial computed tomography chest showing a hypoplastic left bronchus and lung, with compensatory hypertrophy of the right lung, extending across the midline to occupy most of the left hemithorax

An anomalous right upper lobe bronchus was noted arising directly from the trachea, high above the level of the carina [Case 3]. The left pulmonary artery was also completely absent [Case 4]. Features were considered as those of complete left pulmonary agenesis. The cases are summarized in Table 1.

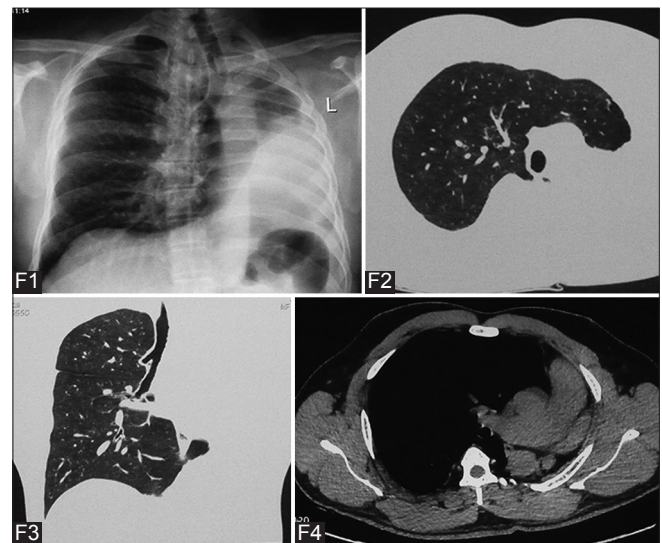
Discussion

The incidence of pulmonary hypoplasia is estimated as approximately 1 in 15,000 live births, with no gender prevalence. Most cases present in neonates, infants, and childhood, but a few cases may remain asymptomatic until adolescence and adulthood and are discovered incidentally.^[1-4]

Pulmonary underdevelopment represents a broad spectrum of abnormalities which are classified as:

1. Pulmonary agenesis (complete absence of the lung, bronchus, and pulmonary artery)
2. Pulmonary aplasia (rudimentary blind-ending bronchus, but absent lung and pulmonary artery)
3. Pulmonary hypoplasia (hypoplastic bronchus with variable lung tissue reduction).^[5-7]

When symptoms occur, they are usually related to the severity of the hypoplasia and may present as recurrent episodes of wheezing or pneumonia, as the underdeveloped lung is more susceptible to bacterial or viral proliferation. The function of the other lung which has undergone compensatory



Case 6: Fig.1: 32-year-old male with routine posterior-anterior chest radiograph, showing severe mediastinal shift to the left and hyper-inflated right lung. Fig.2: Axial computed tomography of the chest with lung window showing the right lung crossing the midline to the left. Fig. 3: Coronal reformatted computed tomography chest with lung window showing the absence of left bronchus. Note an anomalous early branch of the right upper lobe bronchus. Fig.4: Axial computed tomography chest with mediastinal window. Note a large single (right) pulmonary artery and absence of the left pulmonary artery

Table 1: Summary table pulmonary hypoplasia/agenesis in adults

Case	Age (years)	Sex	Chest X-ray indication	Side	Chest X-ray findings/presumptive diagnosis	CT findings
1	19	Male	Residence visa	Right	Large, smooth margin lower lobe mass overlying elevated right diaphragm ? Hydatid cyst	Sessile anomaly of hepatic dome extending into the right lower chest
2	28	Male	Preemployment	Left	Left basal opacity with positive meniscus sign and layering on decubitus film ? Pleural effusion	Herniated omentum and bowel Grossly reduced left lung volume
3	13	Female	School admission	Right	Obliterated, elevated right hemidiaphragm. Mild cardiac shift to the right ?right lower lobe collapse/consolidation	Markedly elevated liver dome with slight cardiac shift to the right, and reduced right lung volume
4	25	Male	Preemployment	Right	Right basal opacity, poorly defined, elevated right hemidiaphragm, and cardiac shift to the right ? Lower lobe collapse ? Pleural fluid collection	Deformed hepatic dome elevated into right chest, with a malpositioned gallbladder. Reduced right lung volume
5	28	Female	Preemployment	Left	Opaque left hemithorax with severe mediastinal shift to the left Hyperinflated right lung ? Post-TB destroyed left lung ?? Pulmonary hypoplasia	Compensatory emphysema of right lung, extending across the midline. Rudimentary left bronchus and lung. Complete cardiac shift to the left
6	32	Male	Preemployment	Left	Opaque left hemithorax with severe mediastinal shift to the left Hyper-inflated right lung ? Destroyed lung from TB ?? Pulmonary hypoplasia	Compensatory emphysema of right lung, crossing the midline to occupy most of the thorax Aplastic left main bronchus Anomalous right upper lobe bronchus from the trachea. Absent left pulmonary artery

TB – Tuberculosis, CT – Computed tomography

hypertrophy may be seriously impaired by pneumothorax or pulmonary hypertension.^[6,7]

The diagnosis of pulmonary hypoplasia may be difficult on the regular chest radiograph. Two of our cases were wrongly diagnosed as lower lobe collapse, an experience previously reported.^[6]

Pulmonary hypoplasia is well known to be associated with diaphragmatic hernia.^[7] This was present in two of our cases (Cases 1 and 2). An elevated liver dome with diaphragmatic elevation and ipsilateral cardiac shift was observed in two other cases (Cases 3 and 4).

It is difficult to ascertain if pulmonary hypoplasia is a consequence of the restriction of normal pulmonary development by the presence of a hernia, or if the hernia and mediastinal shift observed are secondary reactions to a reduced volume of the hemithorax engendered by pulmonary hypoplasia.

Contrast-enhanced CT remains the best modality for the evaluation of pulmonary hypoplasia because of its capacity for multiplanar reformation and the excellent demonstration of the vascular system.^[8-10]

Intravenous contrast was not administered in some of our cases because in the context of routine chest examination and the absence of symptoms, once the diagnosis was established

on plain CT, contrast administration was considered superfluous and an additional cost.

Apart from CT, other ancillary procedures such as bronchoscopy, bronchography, ventilation- perfusion scintigraphy, and magnetic resonance imaging have also been employed in the evaluation of pulmonary hypoplasia.^[9-11]

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Conflicts of interest

There are no conflicts of interest.

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