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Aplasia – Congenital Lung Abnormality with Non-Development

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INTRODUCTION

The following case presentation is of a 75-year-old male who presented to the emergency department with chronic obstructive pulmonary disease (COPD) exacerbation, secondary to pneumonia, and a history of a congenital left lung abnormality with non-development. Since the first report by de Pozzis in 1673, more than 200 cases of lung abnormalities with non-development have been recorded.[1,2] The first proposed classification of underdevelopment of the lung was introduced by Schneider in 1912: Class I Agenesis - total absence of bronchus and lung, Class II Aplasia - rudimentary bronchus without lung tissue, and Class III Hypoplasia - bronchial hypoplasia and variable but reduced amount of lung tissue.[1,2] The rarity of reports and applicable literature involving congenital lung abnormalities with non-development in the elderly, such as pulmonary aplasia, prompted this case presentation.

CASE PRESENTATION

A 75-year-old male patient presented to the emergency department complaining of increasing dyspnea and symptoms of fatigue for three days after developing an upper respiratory infection with a non-productive cough. His dyspnea became worse at night, and he denied any fever or pain. He had not used his home oxygen or continuous positive airway pressure (CPAP) machine for the past month while traveling from Louisiana to Georgia to visit family. His past medical history includes hypertension, high cholesterol, COPD, obstructive sleep apnea, 30-year history of 2 to 3 cigars/day, and a congenital left lung abnormality with non-development.

In the emergency room, the patient was administered two aerosol treatments with 1.25 mg Xopenex (levalbuterol) and normal saline via a small volume nebulizer for right upper lobe wheezing and started on 500 mg Levaquin (levofloxacin) intravenously to treat his suspected pneumonia. After receiving treatments in the emergency room the patient was admitted to the pulmonary medicine floor for further observation. Admission diagnosis was COPD exacerbation secondary to pneumonia.

The patient's chest radiograph upon admission revealed almost complete opacification of the left hemithorax, possibly due to completely atelectatic left lung and/or dense left pulmonary air space consolidation, with a right pleural effusion. (See Figure 1)

Figure 1. Admission Chest Radiograph

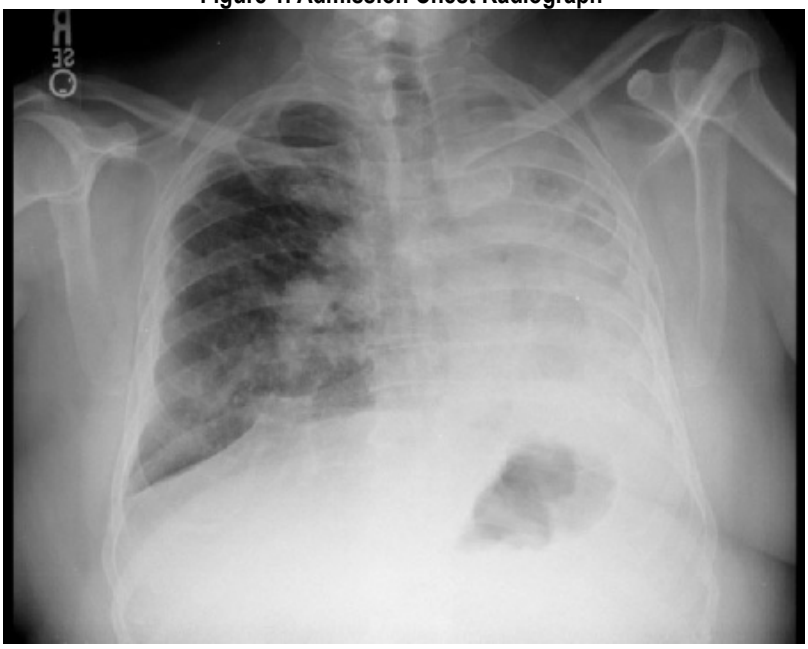
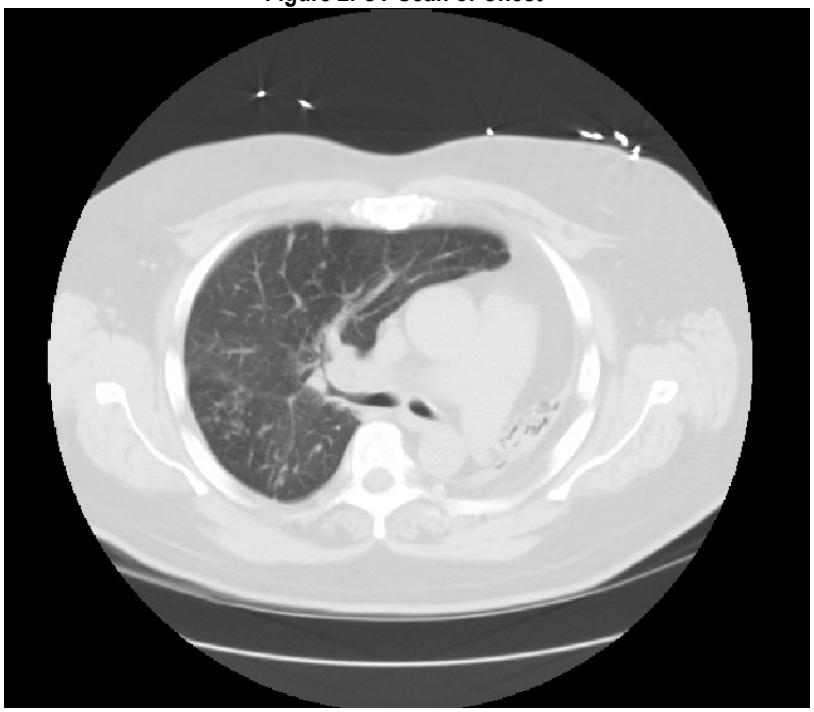


Figure 2. CT Scan of Chest

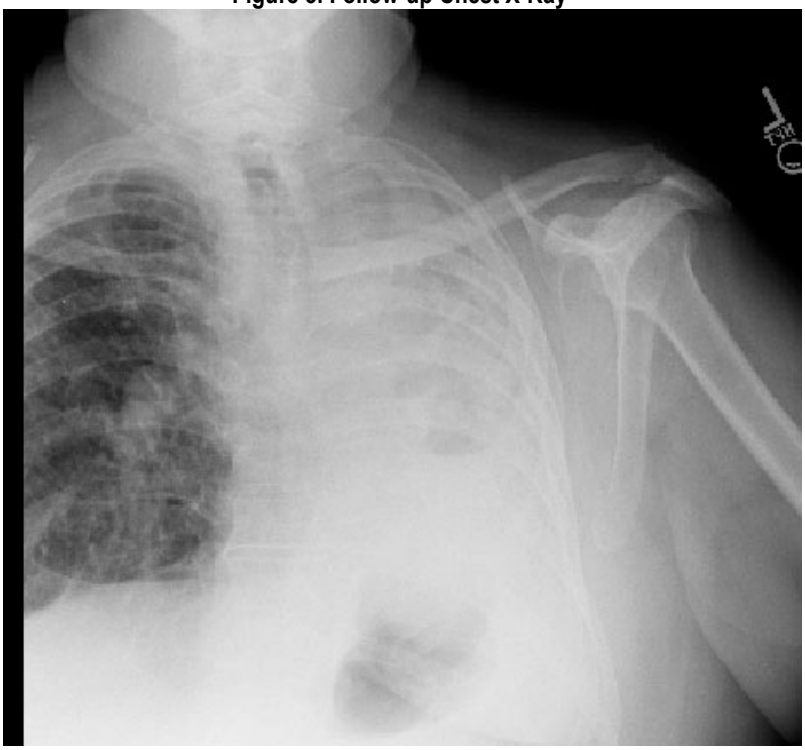


Computed tomography (CT) scan revealed a left lung abnormality with non-development, bronchiectatic changes in the left lower chest, and a small cavitary lesion noted with a small amount of fluid. CT scan also revealed a leftward shift of the mediastinal structures with hyperexpansion of the right lung. No further lung tissue existed below the left mainstem bronchus.

Oxygen was delivered at 2LPM via nasal cannula to treat hypoxemia, dyspnea, and per patient's use of oxygen at home. Consistent with COPD, the arterial blood gases at seven days after admission on 2 LPM of oxygen via nasal cannula were pH 7.42, $PaCO_2$ 67, PaO_2 64, HCO_3^- 42.7, and BE 17.

The patient was discharged after 9 days with reduced shortness of breath, decreased cough, and resolution of pneumonia. Upon discharge, the patient was given the pneumococcal vaccine, and his blood cultures were negative. He was discharged with his home medications and was to follow up with pulmonology as an outpatient. (See Figure 2)

Figure 3. Follow-up Chest X-Ray



During his follow-up, a chest radiograph was taken and revealed improved but not completely resolved patchy areas of opacities, consistent with COPD in the right lung, with an unchanged blunted right costophrenic angle possibly due to pleural thickening. (See Figure 3)

DISCUSSION

This patient had a history of congenital left lung abnormality with non-development. Of the three underdevelopment classifications proposed by Schneider, pulmonary aplasia is the most similar to this patient in comparison to chest radiographs and CT scan images from previous case studies. Pulmonary aplasia involves a rudimentary bronchus with the absence of lung parenchyma, as with the patient in this case.[1,2] Because this is a congenital lung abnormality, the patient was born with only one developed lung: the right lung. Compensation for the non-development of his left lung was evident in the CT scan with hyperexpansion of the right lung and a leftward shift of the mediastinal structures. Although a mediastinal shift was present, the patient's heart was of normal size. Further tests and studies on this patient, and patients like him, would be beneficial in learning more about the effects of pulmonary underdevelopment in the elderly.

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