CASE OF THE MONTH

Can you diagnose this condition on plain radiography?

K GANGADHAR, MD, RD

Department of Radiodiagnosis and Imaging, IMS, Banaras Hindu University, Varanasi, India

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Case report

A 40-year-old male presented with gradual progressive shortness of breath, mild cough without expectoration and right-sided chest discomfort of 6 months' duration. The pain was diffuse, located anteriorly and posteriorly, and did not increase on deep inspiration or coughing. Patient history did not reveal orthopnoea, palpitations, expectoration, fever, anorexia or weight loss. The patient also had no history of pulmonary tuberculosis or rheumatic heart disease. The patient was of average build and afebrile. There was no pallor, icterus, cyanosis, clubbing, lymphadenopathy, oedema of feet or raised jugular venous pulse. There was marked reduced movement of the right hemithorax, right chest deformity, and the trachea and heart were shifted to the right. The entire right hemithorax, except the

infraclavicular area, were dull on percussion. Breath sounds were absent on the right side (Figures 1–5). Could the diagnosis have been made with plain radiography alone?

Discussion

A diagnosis of unilateral pulmonary agenesis was made. Pulmonary agenesis is a rare malformation that can be seen in isolation or associated with other anomalies, such as oesophageal atresia, tracheo-oesophageal fistula, cardiac malformation, horse-shoe kidney and anal atresia. Abnormal development of the aortic arches during embryogenesis, especially involving the first and the second branchial arches, is the most common overt malformation and may be the cause of these associated anomalies [2]. Parental consanguinity through an autosomal recessive mode of inheritance may be a predisposing factor [3]. The incidence of this condition, although not known precisely, is between 0.0034% and 0.0097% [3].

Figure 1. Chest radiograph showing homogeneous opacification of the right lung zones with areas of radiolucency in the mid- and lower lung zones. A gross mediastinal shift towards the right side is noted with tracheal deviation. Crowding of the ribs is noted on the right side, suggestive of volume loss features.

Figure 2. Thorax coronal view contrast-enhanced CT with lung window settings showing absent right lung tissue, absent right main bronchus and gross mediastinal shift towards the right side.

Address correspondence to: Dr Kiran Gangadhar, Radiodiagnosis, Banaras Hindu University, BHU Varanasi, Uttar Pradesh 221005, India. E-mail: kirang.585@gmail.com

Figure 3. Thorax axial view contrast-enhanced CT with lung window settings showing absent right lung tissue, absent right main bronchus and gross mediastinal shift towards the right side. Anterior transmediastinal herniations in the left lung towards the right side are noted (causing right-sided radiolucencies on the chest radiograph).

Pulmonary agenesis is morphologically classified by the extent by which bronchopulmonary tissue is absent. Gould and Hasleton [4] divide pulmonary agenesis into (1) bilateral complete agenesis; (2) unilateral agenesis with (a) complete absence of bronchi, (b) rudimentary bronchus present but no pulmonary tissue, or (c) poorly developed main bronchus with poorly organised parenchyma; and (3) lobar agenesis. However, a more useful clinical classification is to use angiographic data to divide the patients into two groups: the first with complete absence of lung parenchyma and the ipsilateral pulmonary artery and the second with hypoplastic pulmonary tissue present. The

Figure 4. Thorax axial view contrast-enhanced CT with abdominal window settings showing absent right lung tissue, absent right pulmonary artery main branch, absent right main bronchus and gross mediastinal shift towards the right side. Anterior transmediastinal herniations in the left lung towards the right side are noted with crowding of ribs at the right side.

Figure 5. Thorax coronal view contrast-enhanced CT with abdominal window settings showing absent right lung tissue, absent right pulmonary artery main branch and gross mediastinal shift towards the right side with crowding of the ribs on the right side.

latter group may be further subdivided into two groups: the first where the affected lung receives its blood supply from the ipsilateral pulmonary artery and the second where the lung is supplied by systemic circulation [3]. This may have important therapeutic implications because, in the absence of a systemic blood supply, if the ipsilateral pulmonary artery is also absent, there will be no lung tissue on that side, hence surgery should not be considered. By contrast, pneumonectomy or lobectomy may be required in the second subgroup when the systemic blood flow causes congestive heart failure or recurrent respiratory infections.

Almost 70% of cases affect the left side and there is a male predominance over females [1]. Associated malformations are present in 50% of the cases and are a major cause of morbidity and mortality. Most patients suffer from recurrent chest infections in childhood, and about half of the affected individuals die within the first 5 years of life itself. However, as in the present case, many affected individuals remain asymptomatic until adulthood and may even survive to old age. By contrast, bilateral pulmonary agenesis is obviously incompatible with life.

Asymptomatic cases need no intervention. However, chest infections should be treated promptly and aggressively. Patients with a rudimentary bud may require surgical resection if medical management fails to control the infection. Associated congenital anomalies should be corrected wherever feasible. The present case report highlights the fact that in adults, clinical features suggestive of collapse may also be due to uncommon conditions like pulmonary agenesis.

References

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