

ANSWER TO PHOTO QUIZ (PAGE 377)

IS THAT MASSIVE EFFUSION IN THE RIGHT PLEURAL CAVITY?

DIAGNOSIS

The diagnosis is pulmonary hypoplasia. This is a rare developmental disorder, and right lung agenesis is considered rarer than its left-sided counterpart.¹ Up to 50% of all cases are associated with cardiac, genitourinary, gastrointestinal, and musculoskeletal anomalies, and the outcome of patients with cardiovascular anomalies is reported to be poor. This condition is often diagnosed in childhood but may be delayed until adulthood in the absence of comorbid anomalies.² Presentations vary from clinically asymptomatic to those presenting with respiratory distress, cyanosis, or symptomatology related to the associated anomalies in the genitourinary or musculoskeletal systems. Several syndromes such as Goldenhar syndrome, lung agenesis, congenital heart defects, thumb anomalies syndrome, and Scimitar syndrome are associated with pulmonary agenesis.³ The importance of identifying this is manifold. First, the radiological findings may appear to be similar to pneumonia, with cases of lung agenesis wrongly managed as pneumonia, as often reported in literature. Second, the finding of lung agenesis should prompt a search for other associated anomalies, which may contribute to the burden of comorbidities. Finally, the presence of lung agenesis may predispose the individual to recurrent respiratory infections and its sequelae; hence, appropriate preventive measures could prove to be beneficial to this set of patients. Chest X-ray examination alone may lead to mis-diagnosis of pulmonary hypoplasia. Multi-slice spiral CT examination has the advantages including continuous and

rapid scanning imaging, volume data collection, excellent multi-axial plane, and three-dimensional reconstruction images. It can clearly show the structures of pulmonary blood vessels and bronchi, and is easy to operate with high accuracy. It is therefore considered as the preferred diagnostic method for congenital pulmonary dysplasia. Fiberoptic bronchoscope can also be used as an auxiliary diagnostic method for this disease, which can show the degree of hypoplasia, such as the blind end and stenosis of the diseased bronchus.

Treatment is conservative with very little role for surgery in aplasia or agenesis.⁴ Prognosis largely depends on the functional integrity of the remnant lung as well as the presence of other comorbidities.

DISCLOSURES

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