A 38-year-old man with lung cysts

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A 38-year-old man gave history of cough with expectoration and dyspnea since childhood. He had 2-3 exacerbations a year. He denied history of wheezing or hemoptysis. He had no history of exanthematous fever or pneumonia in childhood. There was no relevant occupational history or systemic complaint. He was an ex-smoker, having quit smoking 10 years ago; he had a smoking index of 7.5 pack years. He was married, with three children. His maternal aunt was diagnosed to have bronchial asthma. The physical examination was normal.

The chest radiograph and high-resolution computed tomography (HRCT) of thorax are shown in Figures 1 and Figures 2a and b, respectively. Hemogram and biochemical investigations were normal. Spirometry showed forced vital capacity (FVC): 2.23 L (55% of predicted), forced expiratory volume in one second (FEV_1): 1.94 L (56% of predicted), and FEV_1/FVC: 87%; there was improvement in FEV_1 of 200 mL following inhalation of 200 µg of salbutamol.

Questions

1. What do the chest radiograph and HRCT show?
2. What are the possible differential diagnoses on HRCT thorax? What is the most likely diagnosis if the history and HRCT thorax are correlated?
3. Which one of the two is the correct terminology: 'congenital pulmonary airway malformation' or 'congenital cystic adenoid malformation'? What is the new classification of this condition?

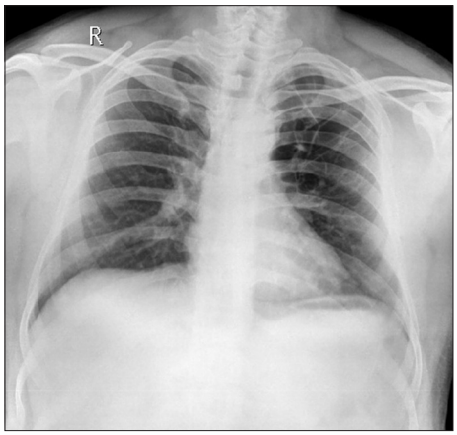

Figure 1: Chest radiograph (posteroanterior view)

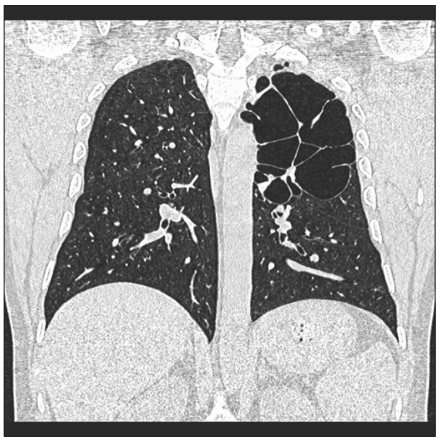

Figure 2a: High-resolution computed tomography of thorax (transverse section)

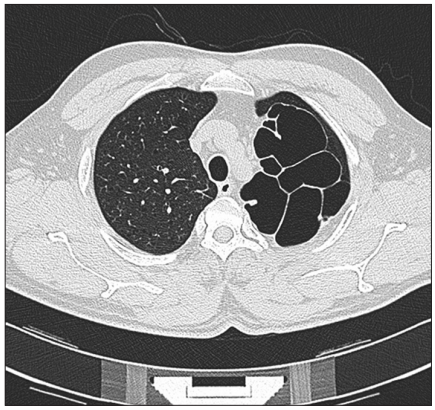

Figure 2b: High-resolution computed tomography of thorax (sagittal section)

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Answers

1. The chest radiograph shows multiple ill-defined air lucent opacities in the left upper lobe. On HRCT of the thorax, however, the air lucent opacities appear to be due to multiple confluent cysts of 2-5 cm size each.
2. The various causes of cystic lesions on imaging are given in Table 1.[1-3] Amongst the congenital malformations, congenital lobar emphysema (CLE) is unlikely because it usually presents before 6 months of age. Also, in CLE, HRCT thorax will show hyperinflated lungs. Bronchogenic cysts are usually single and are seen in the middle third of the lung. The possibility of intralobar sequestration can be considered as the lesions are cystic; however, sequestration is usually seen in the medial aspect of a posterior lung base and hence is unlikely in this case. Congenital pulmonary airway malformation (CPAM) appears to be the most plausible amongst the congenital etiologies. Acquired causes like infection and cysts with interstitial lung disease are unlikely because the cysts are thin walled and the remaining lung parenchyma is normal. The cysts are too large and are located in the upper lobe and hence bronchiectasis is also improbable. A cystic neoplasm is the other possible differential diagnosis. Thus, amongst the congenital malformations and acquired causes of cysts, CPAM and cystic neoplasm are the most likely diagnoses. The patient was subjected to left upper lobectomy. The resected specimen confirmed the presence of multiple large intercommunicating cysts filled with gelatinous mucoid material [Figure 3]. The histopathology showed the cysts to be lined by pseudostratified ciliated columnar epithelium [Figure 4], with underlying fibromuscular bands; the appearance was suggestive of type 1 CPAM.
3. CPAM was earlier known as congenital cystic adenomatoid malformation.[4] Recently, it has been renamed as CPAM because not all the lesions are cystic or adenomatoid.[5] On the basis of the apparent site of origin, five types [Figure 5] have been described in the new classification[6] [Table 2]: Type 0 - alveolar dysplasia or dysgenesis involving the proximal tracheobronchial tree; type 1 - proximal acinus (bronchial/bronchiolar); type 2 -midacinus (bronchiolar); type 3 - bronchiolar/alveolar duct; and type 4 - alveolar saccular/distal acinar.

Table 1: Causes of cystic lung

Congenital cystic lung diseases
Congenital lobar emphysema
Intrapulmonary bronchogenic cyst
Sequestration
Congenital pulmonary airway malformation (CPAM)
Cysts in hypoplastic lung associated with absent pulmonary artery or vein
Infectious causes for cavity/abscess/cysts
Abscess
Necrotizing pneumonia
Endobronchial tuberculosis
Fungal infection
Post-infectious pneumatocele
Pneumocystis jiroveci
Hydatid
Chronic stage of interstitial lung diseases
Acute interstitial pneumonia
Hypersensitivity pneumonitis
Heiner syndrome (cow's milk hypersensitivity)
Nonspecific interstitial pneumonia
Chronic pneumonitis of infant
Pulmonary Langerhans cell *histiocytosis*
Cystic neoplasms:
Pulmonary blastoma arising from CPAM
Congenital pulmonary myofibroblastic tumor
Multicystic mesothelioma
Mesenchymal hamartoma
Congenital pulmonary myofibroblastic tumor
Cystic metastasis
Others
Post-traumatic
Pulmonary interstitial emphysema
Bronchiectasis

Discussion

CPAM is a rare hamartomatous developmental anomaly that results from unsystematic spread of tubular bronchioles and enlarged alveolar tissue.[5] It was first described by Chin

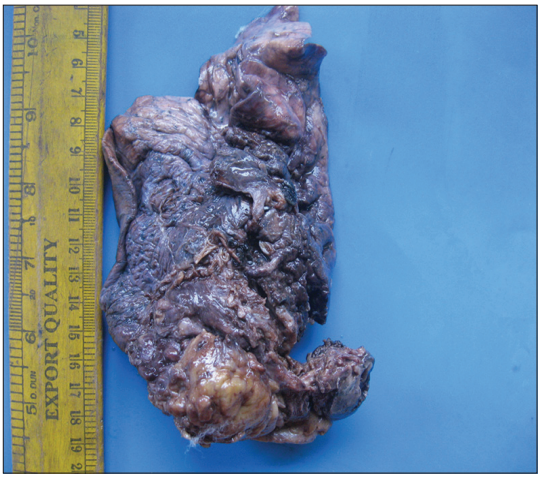

Figure 3: Specimen of resected lung showing multiple intercommunicating cysts

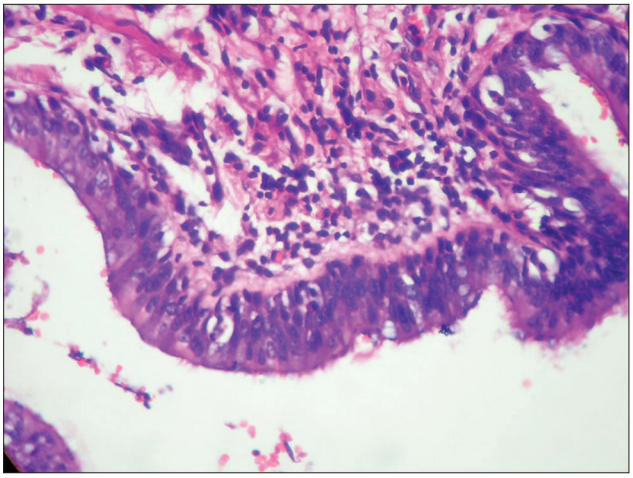

Figure 4: High-resolution photomicrograph of resected lung

Table 2: Types of congenital pulmonary airway malformation

Types	Frequency	Age of presentation	Lobe/lung involved	Associated abnormality	Radiology	Pathology	References
Type 0 tracheobronchial subtype	Rarest, seen in <2% of cases	At birth	Both the lungs are involved. Death likely immediately after delivery unless the fetus is put on extracorporal membrane oxygenation	It is commonly associated with congenital cardiovascular abnormalities or renal hypoplasia	Since the proximal tracheobronchial tree is involved the lungs appear nonaerated	Multiple irregular bronchiole-like structures, many of which are surrounded by thick cartilaginous plates and bundles of smooth muscle. Vascular structures are often >100 mm away from these bronchiole-like structures, making gas exchange impossible	6
Type 1 bronchial/ bronchiole subtype	Most common, 60-70% of cases	Neonatal period, childhood, adulthood; commonest amongst all CPAM to present in adult life	Usually restricted to a single lobe	Primary pulmonary rhabdomyosarcoma, mucinous bronchoalveolar carcinoma, focal mucous cell hypoplasia	Large cysts up to 10 cm	Epithelial lining consisting of a ciliated pseudostratified columnar epithelium overlying fibromuscular tissue	7
Type 2 bronchiole subtype	10-15% of cases	Diagnosed usually in the neonatal period due to associated anomalies	Usually single lobe	Other congenital abnormalities such as renal agenesis, extralobar sequastration	Small cysts <2 cm in diameter	Back-to-back bronchiolar structures lined by low columnar cells blend in with normal alveolar structures	8
Type 3 bronchiole/ alveolar duct	5% of cases	Usually neonatal or early childhood	Entire lung or lobe unilaterally	Pulmonary hypoplasia, heart failure, polyhydramnios, extralobar sequestration	Very small cysts	Scattered bronchiolar or alveolar duct-like structures lined by a low cuboidal epithelium are seen; there is a characteristic absence of pulmonary vasculature	5
Type 4 distal acinar subtype	10% of cases	Neonatal, childhood, rarely adult	Usually single lobe	Pleuropulmonary blastoma; it is important to distinguish type 4 CPAM from pleuropulmonary blastomas, as the cystic form of the latter can very closely resemble CPAM type 4	It grossly resembles type 1 CPAM, with large peripheral cystic lesions	Cysts are lined by both type 1 and type 2 pneumocytes	9

and Tang in 1949. It is seen in about 1 in 25000-35000 live births.[4] CPAM type 0 is incompatible with life and hence always presents before or immediately after birth. The other types are diagnosed during the neonatal period, when respiratory distress can be caused by inflation of the cysts with spontaneous respiration, or during childhood or

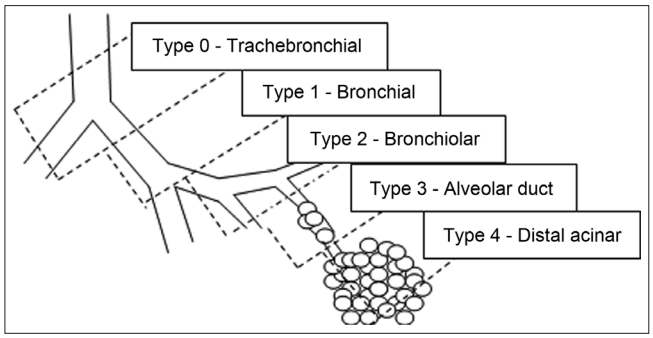

Figure 5: Types of congenital pulmonary airway malformation

adulthood, when the condition causes repeated respiratory tract infection, pneumothoraces, or hemoptysis.[7] CPAM type 1 may sometimes remain asymptomatic.[7] There is no sex predilection or chromosomal anomaly observed with CPAM. Radiologically, type 1 and type 4 have large cysts that may be up to 10 cm in size and type 2 and type 3 have small cysts that are <2 cm in size.[8,9] Histopathology is essential to confirm the diagnosis and ascertain the type of CPAM.

CPAM may be associated with congenital anomalies such as congenital heart disease (e.g., left heart hypoplasia); pulmonary malformations (e.g., sequestration, pulmonary hypoplasia); skeletal anomalies (e.g., pectus excavatum, genu varum, scoliosis); and renal anomalies (e.g., renal agenesis).[10] Some of these anomalies are common with specific types of CPAM [Table 2]. If CPAM is diagnosed during the early neonatal period, conservative management is recommended, as postnatal regression of the lesion may occur. Surgical intervention remains the gold standard for CPAM diagnosed at later ages because remissions are infrequent and because of the risk of repeated infections and malignant transformation.[11]

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