CASE REPORT

A 26-year-old asymptomatic man presented for evaluation of an incidentally detected chest radiographic abnormality while undergoing pre-immigration health check-up. There was no history of chest trauma or any symptom suggesting recurrent lung infections in the past. He had never smoked and denied any family member suffering from similar illness. He recalled receiving some medication (possibly anti-tubercular drugs) for about a year when he was 8-year old for an apparent lesion in his chest radiograph. Previous clinical records and radiographs were not available for verification and comparison. He could not provide the details of his birth history. General examination was unremarkable. Chest auscultation revealed diminished breath sound in right upper lobe area. Examination of other systems was normal. Routine hemogram, renal, and liver function tests were normal. The chest radiographs showed a large thin-walled cyst occupying more than two-third of right lung field with flattening of diaphragm and no significant mediastinal shift [Figure 1a and b]. Computed tomography (CT) scan of chest showed a large thin-walled gas containing cyst in right upper lobe with mild hyperinflation of surrounding lung [Figure 2]. Screening sonography of abdomen and heart failed to reveal any abnormality. He refused for a surgical resection to elucidate the exact nature of cyst despite explaining the risk and benefits as he was completely asymptomatic.

QUESTIONS

Q1: What is the most likely diagnosis?
Q2: What are the other differential diagnoses?

Figure 1: (a) Posteroanterior view showing a large thin-walled empty cyst occupying more than two-third of right hemithorax without any gross mediastinal shift. The right hemidiaphragm is flattened (b) Right lateral view demonstrates the cyst in right upper lobe

Figure 2: Computed tomography scan chest showed a large thin-walled gas-filled cyst in right upper lobe with mild hyperinflation of surrounding lung parenchyma without any obvious contralateral mediastinal shift
ANSWERS

Answer 1: Congenital pulmonary airway malformation Type 1.

Answer 2: The other possibilities for such radiographic finding are post-infectious pneumatocele, giant bulla, cystic bronchiectasis, pulmonary sequestration, congenital lobar emphysema and bronchogenic cyst.

DISCUSSION

The term congenital adenomatoid malformation was first used by Chin and Tang in 1949 to describe a left lower lobe lung malformation in a baby born to a mother with polyhydramnios. Subsequently several other cases of congenital bronchopulmonary malformations appearing cystic and composed of bronchial-like and bronchiolar-like structures arranged in a hamartomatous fashion were reported that are collectively known as congenital cystic adenomatoid malformation (CCAM). Nearly 25-30% of all congenital lung malformations are CCAM and about 30% of all patients with CCAM are at risk of respiratory failure at birth. Stocker et al. in 1977 classified the CCAM into three types - Type I, II and III based on clinical and histomorphologic characteristics that is still widely followed. However, over the years it was observed that some bronchopulmonary malformations are neither 'cystic' nor 'adenomatoid' and do not strictly fit into any of the three described categories. In order to accommodate such missing lesions and make CCAM more inclusive, Stocker himself renamed CCAM in 2002 as congenital pulmonary airway malformation (CPAM) - a nomenclature that describes the possible site of origin and components of this bronchopulmonary malformation more meaningfully. CPAM is classified into five types - Types 0, 1, 2, 3, and 4. Types 1, 2, and 3 are same as Stocker's original Types I, II, and III whereas Types 0 and 4 are new additions. Types 0 and 3 are not 'cystic' and Types 0, 1, 2, and 4 are not 'adenomatoid' in nature. Each type of CPAM is discussed briefly.

Type 0 - Tracheal/bronchial origin: Acinar dysgenesis or dysplasia
This is the least common (<2%) of all types and usually affects the entire lung. These cases are born either premature or at term and are severely cyanotic at birth. They do not survive beyond minutes to hours without extracorporeal membrane oxygenation support. Grossly, the lungs are small and firm with diffusely granular surface. Microscopically, the entire lung consists of bronchial-like structures lined by pseudostratified ciliated columnar epithelium separated by loose mesenchymal tissue containing thin-walled vasculature. This type is associated with cardiovascular anomalies and renal hypoplasia. The pulmonary vasculature is poorly developed and situated sufficiently farther from bronchial airspaces that prohibits any gaseous exchange. This form of CPAM is incompatible with life.

Type 1 - Bronchial/bronchiolar origin: The large cyst lesion
This is the most common form of CPAM representing 60-70% of all cases. Most newborns present with respiratory distress within hours to days after birth and the severity of symptoms is determined by the number and size of the cysts. These patients usually present with recurrent respiratory infections. However, some cases may be detected late in adulthood as an incidental chest radiographic finding. Single lung lobe is affected in about 95% of cases with occasional involvement of another ipsilateral lobe. Bilateral lung involvement is extremely rare. The cysts are large (1-10 cm), thin-walled and lined by ciliated pseudostratified columnar epithelium. Polypoidal epithelial projections and clusters of mucigenic cells may be seen. The cysts often intercommunicate and smaller cysts can usually be seen in the tissue adjacent to the larger ones. Bronchoalveolar cell carcinoma has been described either concomitantly or several years after excision of Type 1 CPAM. The prognosis is excellent after surgical resection in this type of lesion.

Type 2 - Bronchiolar origin: The small cyst lesion
Type 2 CPAM is the second most common (15-20%) type. These are mostly unilobar consisting of multiple small (0.5-1.5 cm) cysts lined by a smooth membrane and blends with adjacent normal parenchyma. Microscopic examination reveals ‘back-to-back’ dilated bronchioles lined by cuboidal to low columnar epithelial cells. This type of CPAM is frequently associated with extralobar sequestration and several other anomalies that often dominate the clinical presentation. Anomaly like bilateral renal agenesis is incompatible with life.

Type 3 - Bronchiolar/alveolar duct origin: The adenomatoid lesion
The Type 3 lesion (represents the original congenital adenomatoid malformation described by Chin and Tang in 1949) accounts for 5-10% of cases, occurs almost exclusively in males, and is associated with maternal polyhydramnios in nearly 80% of cases. The lesion is firm, bulky, involves an entire lobe or lung and produces mediastinal shift with compression of adjacent lung. The infants with this lesion may be stillborn or develop severe respiratory distress shortly after birth. Microscopically, randomly scattered bronchiolar/alveolar duct-like structures are lined by low cuboidal epithelium. There is characteristic absence of pulmonary arteries within the lesion. Prognosis is guarded.

Type 4 - Distal acinar origin: The ‘unlined’ cyst lesion
Many Type 4 lesions have been reported earlier as Type I and constitute approximately 10% of all cases. There are no sexual predilections and most are symptomatic between newborn and 4 years. Clinical presentation may vary from mild respiratory distress or pneumonia to tension pneumothorax (a unique feature of the CPAM, Type 4 cases). Occasionally, Type 4 lesions may be an incidental radiographic finding. Grossly, the cysts are large and thin-walled, peripheral and often display prominent
The radiological differentials in this case include post-infectious pneumatocele, giant bulla, cystic bronchiectasis, pulmonary sequestration, congenital lobar emphysema and bronchogenic cyst. The large size of the cyst, absence of preceding lung infection and healthy surrounding parenchyma exclude the possibility of a pneumatocele. The age of the patient, non-smoker status, absence of emphysema or abnormality suggesting a previous tuberculosis insult in adjacent or opposite lung and greater than a hair-line thickness of the cyst wall virtually reject the argument of an emphysematous or post-tubercular bulla. Cystic bronchiectasis is unlikely considering the solitary nature, upper lobe location and lack of recurrent lung infections. Sequestration is more common in posterobasal segments of lower lobes, often on left side, solid or multicystic in appearance and typically demonstrates a systemic arterial supply. There is no hyperinflation of the lung or mediastinal shift to support the diagnosis of congenital lobar emphysema. Most pulmonary bronchogenic cysts are confined to medial one-third of lung, appear as well-circumscribed round or oval nodule or mass and contains air only if communicated with a bronchus from repeated infection.

Surgical resection is the treatment of choice in all patients including those who are asymptomatic considering the risk of recurrent infections and future malignant transformation. Also, the scope for compensatory lung growth after resection is much better in infants compared to adults. Therefore, early surgery should be performed even in asymptomatic patients. Most surgeons prefer to do a lobectomy in order to ensure complete resection and avoid any risk of future malignant process. Lobectomy may be ideal for extensive single lobe involvement and in emergency resection whereas the extent of lung involvement and presence of associated disorders will determine the nature of surgery in those with multilobar disease.

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Conflicts of interest
There are no conflicts of interest.

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