In-Flight Spontaneous Pneumothorax: Congenital Cystic Adenomatoid Malformation of the Lung

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Established Facts

- Congenital cystic adenomatoid malformation is a rare congenital lung abnormality which usually presents with symptoms in newborns.
- Symptomatic presentation of congenital cystic adenomatoid malformation in adult life is extremely rare.

Novel Insights

- Patients with congenital cystic adenomatoid malformation and patients with other forms of cystic lung disease can develop spontaneous pneumothorax during air travel.

Key Words

Air travel • Spontaneous pneumothorax • Congenital cystic adenomatoid malformation

Abstract

Congenital cystic adenomatoid malformation (CCAM) is a rare congenital abnormality. Symptomatic presentation in adult life is extremely uncommon. The usual radiological appearance of CCAM is a cystic space-occupying lesion. Patients with underlying cystic lung disease can develop in-flight complications because of pressure-volume changes during ascent. We report the first ever case in which sponta-

neous pneumothorax during flight was the presenting manifestation of CCAM of the lung in a previously healthy and asymptomatic young adult. We also discuss the physiological changes during air travel which contribute to the pathogenesis of respiratory complications during air travel.

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Introduction

Congenital cystic adenomatoid malformation (CCAM), also known as congenital pulmonary airway malformation (CPAM), is an uncommon developmental

Fig. 1. Posteroanterior chest radiograph showing a right-sided pneumothorax, cystic lucencies in the collapsed right lung, and contralateral shift of the mediastinum.

anomaly of the lower respiratory tract [1]. The condition is presumed to arise due to defective lung development between the 7th and 10th weeks of embryonic life. The condition can have a number of presenting manifestations in neonates and young children, which are the commonest age group affected, ranging from incidental diagnosis on prenatal ultrasonography to acute respiratory failure due to mass effect. A review of this topic in 2005 reported only 29 cases of CCAM identified in adults [2]. The present case, to the best of the authors' knowledge, is the first ever reported case of CCAM presenting as in-flight spontaneous pneumothorax.

Case Presentation

A 26-year-old never-smoker female developed right-sided pleuritic chest pain and dry cough 20 min after takeoff during a short haul (2-hour) flight. The pain improved partially with paracetamol. As the symptoms partially improved after landing, the patient did not seek any further medical attention. During the return flight a day later, the chest pain and cough worsened in-flight and she presented to the emergency outpatient room immediately after landing. On enquiry, she reported that since landing the symptoms had marginally improved. There were no constitutional symptoms or hemoptysis. The patient's past and personal history was unremarkable.

On examination, the patient was afebrile with a pulse rate of 100/min and blood pressure 120/82 mm Hg. The respiratory rate was 28/min and there was no use of accessory muscles of respiration. The general physical examination was normal. Oxygen saturation by pulse oximetry while breathing room air was 93%. On respiratory system examination, the trachea was deviated to the left side. The right hemithorax was hyper resonant to percussion, and breath sounds were markedly diminished in intensity over the right hemithorax. The rest of the systemic examination was normal.

Chest radiograph revealed a large right-sided pneumothorax with contralateral shift of the mediastinum (fig. 1). As there was suggestion of cystic lucencies in the collapsed right lung on the chest radiograph, a high-resolution computed tomographic (HRCT) examination of the thorax was subsequently done. HRCT revealed a right-sided pneumothorax, and on lung window sections (fig. 2a) the right lung appeared to be nearly totally replaced by multiple variably sized cysts. Tube thoracostomy was done and high flow oxygen was administered through a venturi mask. The patient improved and pneumonectomy was planned in view of extensive cystic disease of the right lung. Flexible bronchoscopy performed for preoperative evaluation revealed a normal bronchial anatomy. Preoperative spirometry was suggestive of a moderate restrictive defect. A right posterolateral thoracotomy was performed. Intraoperative findings included cystic disease limited to the right upper lobe which was filled with clear to purulent fluid. The middle and right lower lobes appeared normal intraoperatively; therefore, only an upper lobectomy was performed with intercostal muscle reinforcement of the bronchial stump.

On gross examination of the resected upper lobe (fig. 3a), the external surface showed multiple small thin walled air- and fluid-filled cysts. The cysts were not communicating with the bronchi. Microscopic examination of the sections from the resected lobe (fig. 3b) demonstrated multiple cystic spaces lined by cuboidal to columnar lining epithelium. The cysts were unencapsulated and admixed with adjoining alveoli of the lung. At places hypertrophied muscle fibers were seen separating the cysts. The diagnosis of CCAM was established.

The postoperative period was uneventful. The follow-up CT scan showed normal and expanded right middle and lower lobes along with normal appearance of the left lung (fig. 2b). The patient was discharged home 12 days after surgery and continues to be completely asymptomatic.

Discussion

The first description of CCAM was made by Chin and Tang in 1949 [3]. The condition is characterized by a failure of normal pulmonary development but the underlying pathogenesis remains unclear. CCAM occurs sporadically and there is no known genetic predisposition or gender predilection. The reported incidence is 1.2/10,000 births [4]. The majority (90%) of cases present within the first 2 years of life, usually in newborns that manifest respiratory distress due to the mass effect of expanding

Fig. 2. HRCT scan of the thorax at presentation showing right-sided pneumothorax with multiple variably sized cysts in the right lung (a). Repeat CT scan of the thorax after surgical resection of the right upper lobe showing expansion of the middle and the lower lobes of the right lung which appear normal along with the normal left lung (b).

Fig. 3. Photograph of the resected right upper lobe showing multiple small thin-walled air- and fluid-filled cysts on the external surface (a). Microscopic examination (hematoxylin-eosin stain; $\times 40$) of the sections from the resected lobe showing unencapsulated multiple cystic spaces lined by cuboidal to columnar lining epithelium (b).

cysts. Most of the remaining cases present with recurrent pulmonary infections in young children. Uncommonly, the condition may remain asymptomatic until late in life. The radiographic appearance of CCAM is that of a cystic space-occupying lesion. On chest radiograph and CT scan, radiological features may resemble a unilobar, multicystic, fluid-filled, mass-like, space-occupying opacity

similar to a lung abscess. There is no unilateral predilection and usually the basal segments (most commonly the posterior basal segments) of either lung are involved. Multilobar and bilateral involvement is uncommon. HRCT scan is a very useful imaging modality which can suggest the site and extent of involvement preoperatively [4].

Morphologically, CCAM encompasses a spectrum of abnormalities ranging from disordered development of airways (common) to alveoli (rare). Accordingly, the condition is pathologically divided into five types (types 0–4) by Stocker's classification [5]. These are: type 0 – tracheo-bronchial; type 1 – bronchial/bronchiolar; type 2 – bronchiolar; type 3 – bronchiolar/alveolar duct, and type 4 – distal acinar. Type 0 is the rarest (2% of all cases) whereas type 1 is the most common type (60–70%). Other congenital abnormalities can also occur in association with CCAM, especially type 2. These include renal agenesis, pectus excavatum, Potter's syndrome, bile duct hypoplasia, esophageal atresia with trachea-esophageal fistula, intestinal atresia, and bony abnormalities [5]. The type 4 variety may be associated with childhood malignancies like pleuropulmonary blastoma. The renaming of CCAM to CPAM is based upon two facts; first, the morphological/pathological spectrum follows the anatomic and microscopic features of the 'pulmonary airways', and second, types 0 and 3 are not 'cystic' while types 0, 1, 2, and 4 are not 'adenomatoid' [5].

Air travel is a recognized risk factor among patients with underlying respiratory diseases. Also, respiratory disorders are among the most commonly reported 'in-flight' medical problems after neurological disorders. According to international laws, the aircraft cabin pressure must not surpass that measured at 8,000 ft above sea level (osl) and higher levels of pressurization are associated with decreased aircraft performance and reduced aircraft life. The normal cruising altitude of ordinary flights is between 25,000 and 45,000 ft osl. In cabins pressurized at 8,000 ft osl, the barometric pressure falls from 760 mm Hg (sea level) to 565 mm Hg. Because of this, the partial pressure of oxygen in inspired gas (PiO_2) decreases by 40 mm Hg as compared to sea level. These pressure changes are attained approximately 15 min following takeoff [6].

According to Boyle's law there is an inverse relationship between gas volume and pressure, i.e. volume increases with a reduction in pressure. The average increase in gas volume upon moving from the ground to 8,000 ft osl is approximately 30%. The reduction in atmospheric pressure that occurs during ascent can cause a dramatic increase in noncommunicating gas volume and consequently rupture of an underlying pulmonary cystic lesion leading to pneumothorax, and this is the likely mechanism in the index case also [6].

Symptomatic presentation of CCAM in adulthood is uncommon and therefore it is a diagnosis that is rarely entertained by adult pulmonologists and physicians. To

the best of the authors' knowledge, this is the first reported case of CCAM presenting with spontaneous pneumothorax during air travel and as the initial manifestation in a previously asymptomatic 'healthy' young adult. Despite the rarity of its manifestation in adult life, a spectrum of presentations have been reported including recurrent respiratory infections (commonest of the all), spontaneous pneumothorax, hemothorax, and hemoptysis. A single case of cerebral air embolism during a long haul flight has been previously reported in a patient with CCAM [7].

The index case fitted into the type 2 (bronchiolar) variety. However, no other congenital anomalies were found upon evaluation in the index patient. The patient was fortunate enough not to have developed tension pneumothorax, life-threatening hypoxemia, or fatal systemic gas embolization during the return flight. As suggested by the timing and severity of the symptom profile, our patient most likely developed pneumothorax on the first (forward journey) flight and an increase in pneumothorax volume occurred during the second (return) flight.

In addition to a description of this rare manifestation of a congenital abnormality in adult life, the present case also highlights an important and rare complication, i.e. the occurrence of spontaneous pneumothorax during air travel. Physicians need to be aware that, among patients with underlying cystic lung disease, life-threatening pneumothorax or fatal systemic air embolization can manifest during air travel. Also, presence of an underlying pneumothorax is an absolute contraindication for air travel as a critical increase in pressure and volume may occur. The other two absolute contraindications for air travel are bronchogenic cysts and severe pulmonary arterial hypertension [6]. Cases of patients with bronchogenic cysts developing in-flight complications like systemic gas embolism (mostly fatal) have been previously reported [6]. In patients with suggestive symptoms, spontaneous pneumothorax or pneumomediastinum must be excluded prior to air travel. The authors would also like to highlight here that CCAM should be considered in the list of differential diagnoses when encountering an adult patient with pneumothorax or cystic lung disease.

Financial Disclosure and Conflicts of Interest

None of the authors have any conflict of interest.

References

- 1 Lee SC, Cheng YL, Yu CP: Haemopneumothorax from congenital cystic adenomatoid malformation in a cryptorchidism patient. *Eur Respir J* 2000;15:430–432.
- 2 Herrero Y, Pinilla I, Torres I, Nistal M, Pardo M, Gomez N: Cystic adenomatoid malformation of the lung presenting in adulthood. *Ann Thorac Surg* 2005;79:326–329.
- 3 Ch'In KY, Tang MY: Congenital adenomatoid malformation of one lobe of a lung with general anasarca. *Arch Pathol (Chic)* 1949;48:221–229.
- 4 Kim SR, Lee MH, Lee KA, Park SJ, Min KH, Han HJ, Chung CR, Choi KH, Lee HB, Jin GY, Lee YC: A 38-year-old woman with bilateral cystic lesions in both lower lung lobes. *Chest* 2011;140:544–548.
- 5 Stocker JT: Congenital pulmonary airway malformation – a new name for and an expanded classification of congenital cystic adenomatoid malformation of the lung. *Histopathology* 2002;41:424–431.
- 6 Tzani P, Pisi G, Aiello M, Olivieri D, Chetta A: Flying with respiratory disease. *Respiration* 2010;80:161–170.
- 7 Belcher E, Lawson MH, Nicholson AG, Davison A, Goldstraw P: Congenital cystic adenomatoid malformation presenting as in-flight systemic air embolisation. *Eur Respir J* 2007;30:801–804.