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Case report

Congenital pulmonary airway malformation in a 36 year-old female

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ABSTRACT

Congenital pulmonary airway malformation (CPAM), previously known as congenital cystic adenomatoid malformation (CCAM), is an inborn abnormality of the lower respiratory system. Most often diagnosed in the perinatal period, these anomalies usually present with tachypnea, cyanosis, and respiratory distress. However, rare cases are asymptomatic and undiagnosed until adulthood.

1. Introduction

Congenital pulmonary airway malformation (CPAM), previously known as congenital cystic adenomatoid malformation (CCAM), is an inborn abnormality of the lower respiratory system. Most often diagnosed in the perinatal period, these anomalies usually present with tachypnea, cyanosis, and respiratory distress. However, rare cases are asymptomatic and undiagnosed until adulthood.

2. Case report

A 36-year-old female with an unremarkable past medical history presented to the Emergency Department complaining of a nonproductive, progressive cough over the past 48 h. She admitted to a 7.5-pack year smoking history; however, she has been tobacco-free for the past 21 months. The patient's vitals were stable, and physical exam revealed mild erythema of the oropharynx but was otherwise unremarkable.

A chest radiograph revealed a nodular opacity in the right upper lobe (Fig. 1), warranting further investigation. A contrast-enhanced Chest CT scan showed a lobulated soft tissue density within the posterior segment of the right upper lobe measuring 2.5 × 1.7 × 2.6 cm (Figs. 2 and 3). Bronchoscopy was negative for any bronchial lesions, masses or hemorrhage; pathology revealed non-malignant cytology. Video-assisted thoracoscopic surgery (VATS) lobectomy with mediastinal lymph node dissection indicated pathology consistent with congenital pulmonary airway malformation (Figs. 4 and 5).

3. Discussion

Prior to vigilant perinatal screening, developmental abnormalities were more likely to go unnoticed throughout adolescence and into adulthood. Historically, asymptomatic malformations were often incidentally found while surveying for other disease processes. First described in 1949, congenital pulmonary airway malformations (CPAMs) are rare, embryologic anomalies originating from the lower respiratory tract. Initially classified by

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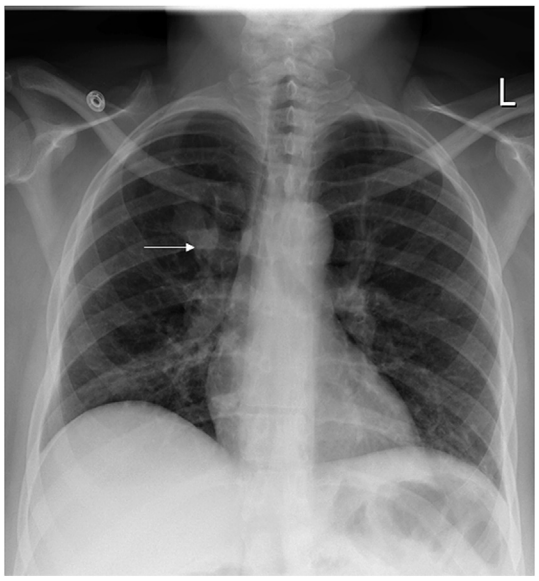

Fig. 1. Chest radiography showed a lobulated medial right upper lobe nodule (arrow).

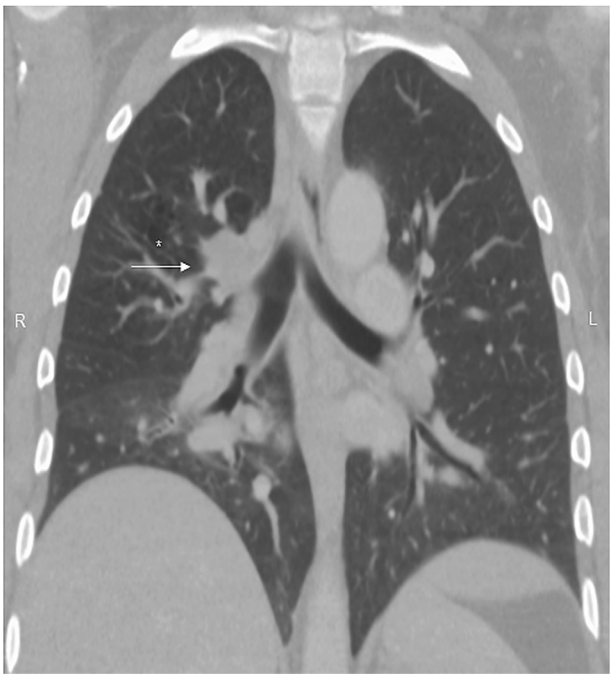

Fig. 3. Thoracic CT on coronal reformation again shows medial right upper lobe nodule (arrow) with surrounding cystic hyperlucency (*).

histology into three groups, more recent taxonomies have increased the spectrum to 5 subcategories (type 0 to type 4) [1]. Type 1 malformations are the most common, making up nearly 70% of all CPAMs. Although the histologic findings can vary, type 1 malformations are often comprised of single or multiple cysts (3–10 cm) usually enclosed within a single lobe [2]. Other categories, such as type 0 and type 3, are almost always incompatible with life. No matter the classification, all forms of CPAM can present with neonatal respiratory distress dependent on the degree of severity [3].

In eleven cases of adult CPAM reported in the literature between 1997 and 2015, including the current case, average age at presentation was 29.5 (range 17–42). Male to female sex ratio, 1.75:1, indicates male predominance. Common presentations include cough, dyspnea and hemoptysis. Incidental findings in the recent era suggest a role for improved imaging over standard radiography. Conservative medical management was only done in two reported cases (see Table 1).

Radiographic presentations vary throughout the spectrum of CPAM; ranging from homogenous masses to multi-cystic lesions. Classically in adults, grossly cystic images in which lucent elements

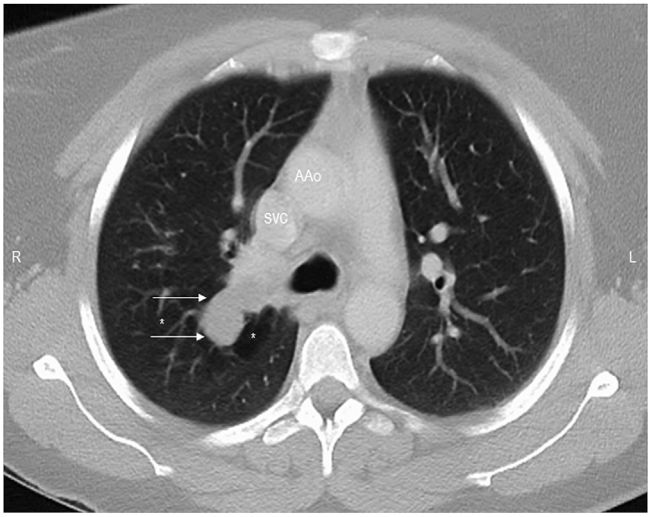

Fig. 2. Axial Thoracic CT showed a soft tissue nodule (arrow) measuring 2.5 × 1.7 × 2.6 cm in medial right with surrounding hyperlucency (*). AAo = ascending aorta, SVC = superior vena cava.

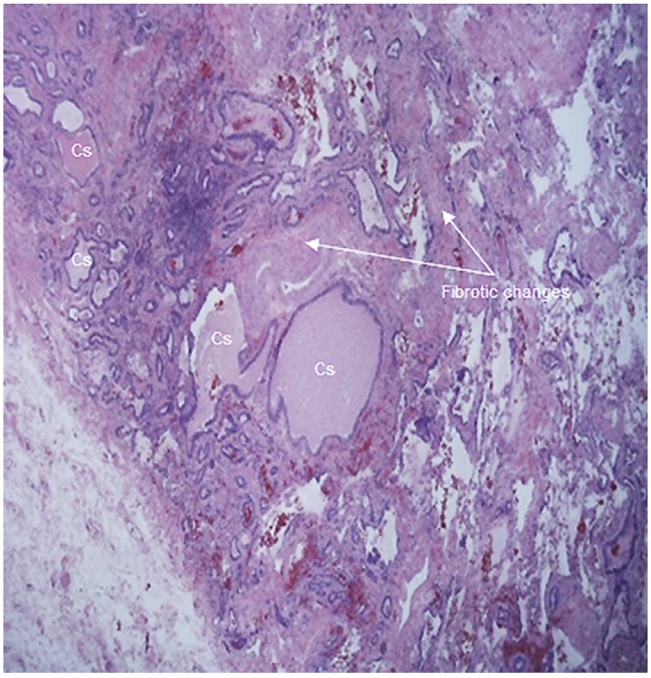

Fig. 4. Video-assisted lung biopsy [H & E stain] showing hemorrhage and cystic adenomatoid malformations(labels).

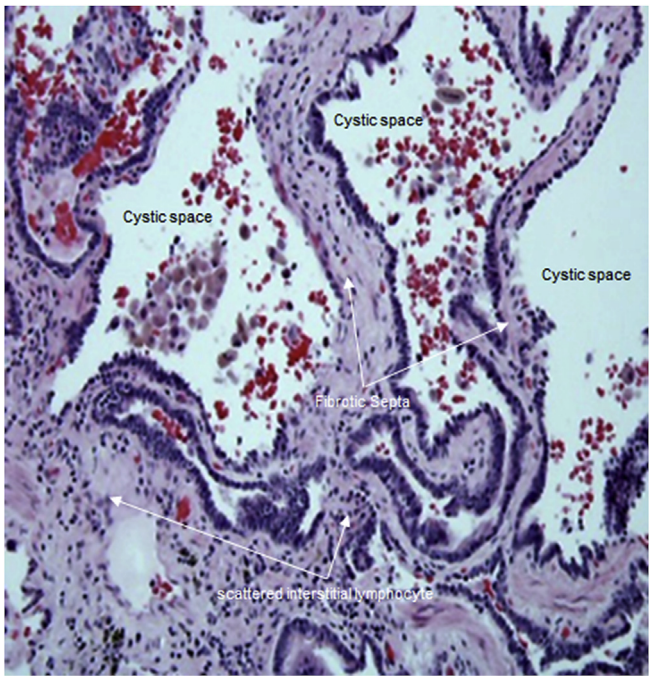

Fig. 5. Video-assisted lung biopsy [H & E stain (high power × 60)] showing cystic structures lined by columnar epithelium along with dense fibrotic septa with predominate lymphocytic infiltration.

Table 1
Case series of 10 Adult Cases Reported in Literature.

Authors	Year reported	Age	Sex	Clinical presentation	Histology	Treatment
Pit et al.	1997	28	Female	Cough and Dyspnea	CCAM Type I	Lobectomy
Mehta et al.	2005	42	Male	Hemoptysis	Type II CCAM + bronchogenic adenocarcinoma	Pneumonectomy
Andrew et al.	2006	24	Male	Pneumothorax	Type II CCAM	Lobectomy
Peegy et al.	2009	40	Female	Incidental finding on chest x-ray	Type I CCAM	Lobectomy
Plit et al.	1997	27	Male	Productive cough	Type II CCAM	Observation
Sarnelli et al.	1997	32	Male	Progressive Dyspnea	Type II CCAM	Observation
Patella et al.	2012	41	Male	Incidental finding on chest x-ray	Type III CCAM	Lobectomy
Harini et al.	2012	19	Male	Hemoptysis	Type I CCAM + mucoepidermoid	Pneumonectomy
Belcher et al.	2007	18	Male	In-flight systemic air embolism	Type I CCAM	Lobectomy
Young et al.	1994	17	Female	Recurrent Pneumonia	Type I CCAM	Lobectomy
Barreiro, Ingnam & Sypert et al.	2015	37	Female	Non-productive cough	Type II CCAM	Lobectomy

predominate are observed. Regardless of the morphology of the tissue, evidence of hemithorax opacification or mediastinal shift is an ominous sign often associated with life-threatening CPAMs [4]. As mentioned previously, asymptomatic lesions often go undiagnosed due to their benign course; however, type 1 and type 4 lesions are noted to carry some malignant potential. Malignancies, particularly those among the adenocarcinoma spectrum previously referred to as bronchioloalveolar carcinoma, may occur in patients with type I CPAM; however, the exact incidence is unknown. Although rare, there have been reports of progression to malignant adenocarcinoma and subsequent death [1]. As in our case, it is important to consider surgical intervention in the management of asymptomatic CPAM to limit the risk of malignant potential and recurrent infection [5,6].

Conflict statement

The author(s) have no personal or financial support or involvement with any organization(s) with a financial interest in the subject matter. They also deny any conflicts of interest or sponsorship. The author verifies that this manuscript is not under review by any other journals. The author(s) have solely contributed to the making of this manuscript.

References

[1] P.S. Lai, D.W. Cohen, M.M. DeCamp, S. Fazio, D.H. Roberts, A 40-year old woman with an asymptomatic cystic lesion in her right lung, Chest 136 (2009) 622–627.
[2] M.M. Cloutier, D.A. Schareffer, D. Hight, Congenital cystic adenomatoid malformation, Chest 103 (1993) 761–764.
[3] S. Sahu, S. Muthuvel, S.S. Naware, S.S. Dhavala, Congenital cystic adenomatoid malformation of lung, Med. J. Armed Forces India 64 (2008) 268–269.
[4] J.T. Stocker, J.E. Madewell, R.M. Drake, Congenital cystic adenomatoid malformation of the lung. Classification and morphologic spectrum, Hum. Pathol. 8 (1977) 155–171.
[5] Y. Herrero, et al., Adult CCAM, Am. Thor. Surg. 79 (2005) 326–329.
[6] A. Nasr, S. Himidan, A.C. Pastor, G. Taylor, P.C. Kim, Is congenital adenomatoid malformation a premalignant lesion for pleuropulmonary blastoma? J. Pediatr. Surg. 45 (6) (2010 June) 1086–1089 . PMID: 20620300.