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.

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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
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

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

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

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

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