Indian Journal of Thoracic and Cardiovascular Surgery (October–December 2018) 34(4):488–490
https://doi.org/10.1007/s12055-018-0650-2

CASE REPORT

Congenital cystic adenomatoid malformation of the lung in adults: report of two cases and review of the literature

Sidharth Garg[1] · Rana Sandeep Singh[1] · Harkant Singh[1]

Received: 26 October 2017 / Revised: 9 January 2018 / Accepted: 11 January 2018 / Published online: 27 February 2018

Abstract

Congenital cystic adenomatoid malformation (CCAM) is characterized by adenomatoid proliferation of bronchiole-like structures and cysts formation. It isan uncommon cause of respiratory distress in infants. Most common presentation is in first two years of life with complaints of respiratory distress.Presentation in adulthood is rare. Surgical intervention is the mainstay treatment.We describe two cases of atypical CCAM presenting in adulthood with past history of tuberculosis. To our knowledge only one case of CCAM with tuberculosis has been reported in literature.

Keywords CCAM · Adults · Tuberculosis

Introduction

Most congenital lower respiratory tract malformations are detected during pregnancy or at birth, thanks to antenatal imaging. Congenital cystic adenomatoid malformation (CCAM), currently referred as congenital pulmonary airway malformation (CPAM), is characterized by an adenomatoid proliferation of bronchiole-like structures and cyst formation. The condition is most commonly found in newborns and children and it may be associated with other malformations. Rarely the presentation is delayed until adulthood. Patients with this condition who survive to adulthood usually suffer from recurrent respiratory bacterial infections.

Here we describe two cases of CCAM presenting in adulthood with unusual clinical findings. Both the patients had a past history of pulmonary tuberculosis. To our knowledge, only one case of CCAM with tuberculosis has been reported [1].

✉ Sidharth Garg
sidharthgarg.pgi@gmail.com

Rana Sandeep Singh
ranassk@rediffmail.com

Harkant Singh
baryah@hotmail.com

[1] Department of Cardiothoracic and Vascular Surgery, Postgraduate Institute of Medical Education and Research, Chandigarh 160012, India

Case reports

Case 1

A 30-year-old female, with history of pulmonary tuberculosis 5 years ago for which she took anti-tuberculosis treatment (ATT), presented with complaint of recurrent episodes of hemoptysis for the past 6 months. On clinical examination, breath sounds were decreased on the right side. On investigations, sputum for acid-fast bacilli was negative. Bronchoscopy revealed blood clots in the right middle bronchus with extension into the right upper and lower lobe bronchi. Contrast-enhanced computed tomography (CECT) chest with CT (computed tomography) angiography showed a destroyed right lung with volume loss predominantly in the right upper lobes. Cystic bronchiectatic changes were seen in the right middle and lower lobe. Contrast outpouching suggestive of Rasmussen's aneurysm was seen in relation to the posterior segment of the right lower lobe.

The patient underwent right pneumonectomy. Gross specimen revealed a large 9 × 4 × 3.5 cm cavity with smooth inner surface close to the apex. On examining sections, multiple cystic spaces lined by pseudostratified ciliated columnar to low cuboidal epithelium were seen. Wall of these cystic structures contained smooth muscle bundles, cartilage fragments, and mucinous glands (Fig. 1). The histology and CT findings were consistent with diagnosis of CCAM type 1.

Case 2

A 20-year-old male with a history of open drainage of right-sided empyema 10 years ago in another institute, which had developed following pulmonary tuberculosis, presented with complaint of discharging sinus from the right lateral chest wall. Clinical examination revealed deformed right side chest wall with pus discharge from previous scar site. Air entry was reduced on the right lower lung field. Pulmonary function testing showed severe restriction. On CECT chest, a large cavity with small amount of fluid in dependent position in relation to the right lower lobe was seen (Fig. 2). A subsegmental bronchus from the right lower lobe was seen communicating with the cavity suggestive of broncho-pleural fistula. The cavity was also seen to be communicating externally suggestive of pleuro-cutaneous fistula.

The patient underwent right postero-lateral thoracotomy, which revealed unhealthy and sequestered right lower lobe with multiple broncho-pleural fistulous openings into a small abscess cavity. Right lower lobectomy along with thoracoplasty was done. Gross specimen showed a large cystic lesion 12 × 9 cm with collapsed adjacent lung parenchyma. On sectioning, multiple large cysts with pseudostratified ciliated columnar epithelium surrounded by smaller cysts with cuboidal to columnar lining were seen. Many smooth muscle bundles and cartilage plates were seen in the cyst wall (Fig. 3). The histology findings were consistent with diagnosis of CCAM type 1.

Discussion

CCAM is a rare developmental, non-hereditary, hamartomatous abnormality of the lower respiratory tract, first described as a distinct disease entity by Ch'in and Tang in 1949 [2]. It occurs sporadically and is not related to genetic predisposition, gender predilection, or maternal factors such as race, age, or environmental exposures. The pathogenesis of CCAM is uncertain but may include an imbalance between cell proliferation and apoptosis during organogenesis. Eighty percent to 85% of cases are recognized in the first 2 years of life and adult presentation is uncommon [3].

It was classified into three subtypes in 1977 and expanded into five types with a new name as CPAM by Stocker in 2002 [4]. Typical histologic feature of CCAM are adenomatoid proliferation of bronchiole-like structures and macro- or microcysts lined by columnar or cuboidal epithelium and absence of cartilage and bronchial glands.

Type 1 CCAM accounts for 70% of cases and is characterized by single or multiple cysts more than 3 cm in diameter lined by pseudostratified ciliated columnar epithelium, along with mucous cells which are considered to be potentially mutant to adenocarcinoma [5]. Type 2 lesion consists of multiple terminal bronchiolar-like uniform cysts smaller than 2 cm in diameter, lined by cuboidal to columnar epithelium. Stocker reported an association of other congenital anomalies specifically renal agenesis in up to 60% cases of type 2 lesions. Type 3 CCAM usually involves an entire lobe of lung and has a sponge-like appearance, constructed by bulk gland-like structures. Type 0 refers to acinar atresia (a Tracheo-bronchial defect) and type 4 has multiple cysts lined by flattened epithelium (an alveolar defect) [4].

Prenatal ultrasonography is able to diagnose most cases of congenital lung anomalies. However, it is difficult to distinguish CCAM from other congenital lung lesions [6]. Chest radiography is essential for CCAM workup, which shows a mass containing air-filled cysts. Other signs include mediastinal shift, pleural and pericardial effusions, and pneumothorax [7]. CT scan is a rapid modality of defining the extent of CCAM in all ages. The typical appearance is multilocular cystic lesions with thin walls surrounded by normal lung parenchyma. Superadded

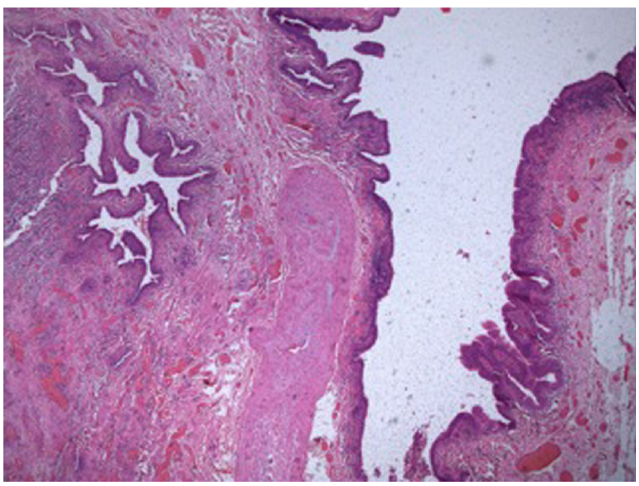

Fig. 1 Cystically dilated airways lined by pseudostratified ciliated columnar to cuboidal epithelium and surrounded by bronchial smooth muscle and cartilage

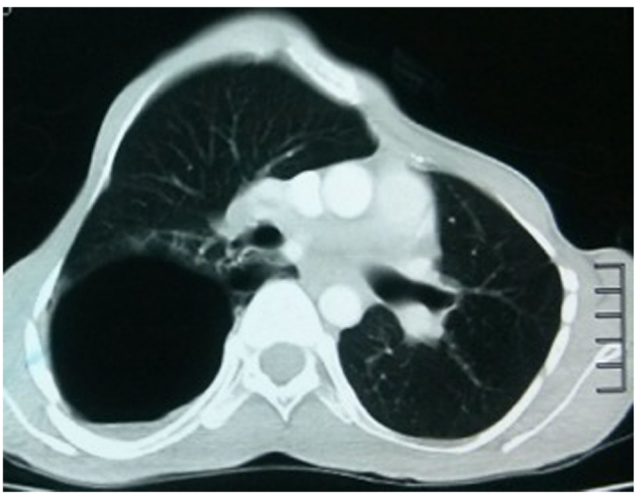

Fig. 2 Contrast-enhanced computerized tomography of the chest. Large cavity with small amount of fluid in dependent position in relation to the right lower lobe

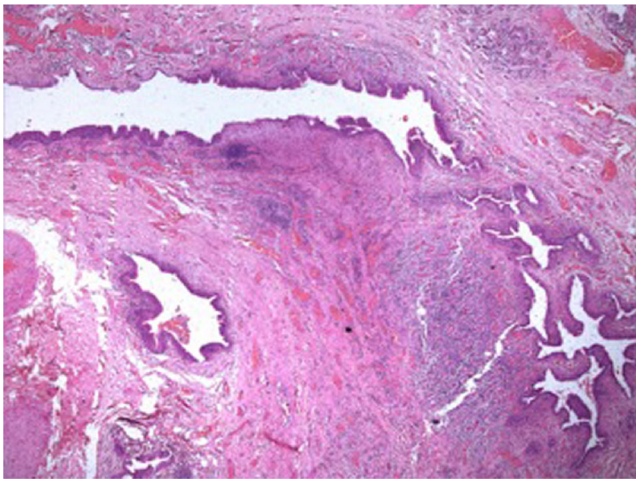

Fig. 3 Cystically dilated airways lined by pseudostratified ciliated columnar to cuboidal epithelium. These airways are surrounded by bronchial smooth muscle and show dense lymphoplasmacytic inflammation in subepithelium

infections may be visualized as well. HRCT (high-resolution computed tomography) is sufficient to differentiate between microcystic and macrocystic lesions [8].

CCAM may have prenatal and postnatal presentation. Prenatally large lesions may be associated with the development of hydrops fetalis, which is a poor prognostic sign. It may also cause compromised pulmonary growth resulting in pulmonary hypoplasia and postnatal respiratory distress [6].

The typical manifestations in the newborns include progressive respiratory distress. Older patients present with history of recurrent pulmonary infections, due to bronchial compression, air trapping, and inability to clear out secretions. Most CCAM in adults involve unilateral lobes of the lung and may be complicated with pulmonary bacterial infections and abscesses [9]. Other presentations may be cough, fever, chest pain, and hemoptysis. CCAM has been associated with the development of malignancies, such as bronchiolo-alveolar carcinoma and adenocarcinoma in the aged [5].

Medical therapy is limited to antibiotics for recurrent pneumonias and supportive therapy in the form of oxygen supplementation and mechanical ventilator support in patients with respiratory distress.

Surgical intervention including both fetal and postnatal surgery is the mainstay therapy. Fetal surgery can be considered in patients with large CCAMs and those complicated by hydrops. Options include thoracocentesis with drainage of a large cyst, thoraco-amniotic shunt that continually drains fluid from the CCAM to the amniotic space, and resection of the affected lobe in cases where no dominant cyst is available [6].

Postnatal resection of the affected part of the lung is the treatment of choice, and this prevents recurrent infections, chances of pneumothorax, and malignant transformation. Surgery should preferably be done before 12 years of age to enhance compensatory lung growth. In older patients, more extensive resections may be required in presence of pneumonia [10].

Conclusion

CCAM is a rare condition and should be kept in mind during workup of adult patients presenting with recurrent respiratory tract infections, not being managed successfully with medical therapy alone. CT scan is the investigation of choice and surgical resection remains the mainstay treatment.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Ethical approval All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. This article does not contain any studies with animals performed by any of the authors.

Informed consent Informed consent was obtained from all individual participants included in the study.

References

1. Cho BS, Kim SJ, Jeon MH, et al. Congenital cystic adenomatoid malformation (Type II) with active tuberculosis in an adult. Br J Radiol. 2008;81:e197–200.
2. Ch'in KY, Tang MY. Congenital adenomatoid malformation of one lobe of a lung with general anasarca. Arch Pathol (Chic). 1949;48:221–229.
3. Rosado-de-Christenson ML, Stocker JT. Congenital cystic adenomatoid malformation of the lung. Radiographics. 1991;11:865–86.
4. Stocker JT. Congenital pulmonary airway malformation: a new name and an expanded classification of congenital cystic adenomatoid malformation of the lung. Histopathology. 2002;41:424–31.
5. Benouaich V, Marcheix B, Begueret H, Brouchet L, Velly JF, Jougon J. Malignancy of congenital cystic adenomatoid malformation of lung in aged. Asian Cardiovasc Thorac Ann. 2009;17:634–6.
6. Adzick NS, Harrison MR, Crombleholme TM, Flake AW, Howell LJ. Fetal lung lesions: management and outcome. Am J Obstet Gynecol. 1998;179:884–9.
7. Schwartz DS, Reyes-Mugica M, Keller MS. Imaging of surgical diseases of the newborn chest. Intrapleural mass lesions. Radiol Clin N Am. 1999;37:1067–78.
8. Kim WS, Lee KS, Kim IO, et al. Congenital cystic adenomatoid malformation of the lung: CT-pathologic correlation. AJR Am J Roentgenol. 1997;168:47–53.
9. Hulnick DH, Naidich DP, McCauley DI, et al. Late presentation of congenital cystic adenomatoid malformation of the lung. Radiology. 1984;151:69.
10. Makhija Z, Moir CR, Allen MS, et al. Surgical management of congenital cystic lung malformations in older patients. Ann Thorac Surg. 2011;91:1568–73.