Case Report

Congenital Cyst Adenoid Malformation Masquerading as Bronchial Asthma

Abstract

Congenital cyst adenoid malformation (CCAM) is a rare congenital malformation occurring in approximately 1–4 in 100,000 births. It is classified into five subtypes with type 1 CCAM is most common subtype. The diagnosis of CCAM is usually made in infancy, and it is rare in adolescents and adults. We report a 15-year-old female, who presented in pediatric outpatient department with a history of recurrent cough since infancy. On the basis of clinical examination, provisional diagnosis of asthma was considered and patient was started on inhaled corticosteroid and long-term β2 agonist. Lung function of the patient revealed low forced expiratory volume-1 s but without bronchodilator reversibility. Therefore, alternative diagnosis was suspected, and the patient was further evaluated with X-ray chest and high resolution computed tomography thorax. Based on radiological findings, a final diagnosis of CCAM was established. The case was highly unusual due to its atypical and late age of presentation. Acquaintance about this condition benefit clinician in making differential diagnosis of recurrent cough.

Keywords: Bronchial asthma, congenital cyst adenoid malformation, lung

Introduction

Congenital cyst adenoid malformation (CCAM) is rare congenital malformation of lung. It is usually due to embryonic insult leading to an arrest in lung development between 4th and 7th week of fetal life.[1] Most cases are diagnosed in neonatal period or in 1st year of life.[2] We describe a 15-year-old female child who presented to us with moderate persistent asthma. The case is unusual due to its late age of onset and its uncommon presentation.

Case Report

A 15-year-old female child presented to us with a history of recurrent cough since infancy. The cough was dry and nocturnal. The patient also had a history of exertional dyspnea and chest tightness. There was no history of rhinorrhea, sneezing, nasal blockage, mouth breathing, and snoring. Family history of child revealed asthma in grandfather who was taking metered dose inhaler. The child received antibiotics, cough syrup, nebulization and steroids on multiple occasions. On examination, her vitals were normal. Her general physical examination was also found to be normal while on respiratory system examination air entry was reduced, and bilateral wheeze was present. Based on history and examination, a clinical diagnosis of asthma was made and child was started on inhaled steroid (Budesonide) and long-acting β2 agonist (Formoterol). On routine spirometry, her forced expiratory volume-1 s (FEV1) was found to be 53% of predicted value and did not show bronchodilator reversibility. As child was not showing bronchodilator reversibility which is typically present in asthma, an alternative diagnosis was suspected, and child was investigated for congenital, inflammatory, autoimmune, and infectious causes. The X-ray chest showed multiple transradiant cystic lucencies of variable size occupying the entire left hemithorax with negligible viable lung tissue visible on the left side with few fibrotic strands. Trachea was pulled toward left side and there was compensatory hyperinflation of the right hemithorax [Figure 1]. High resolution computed tomography thorax confirmed the X-ray finding and showed significant lung herniation from right to left side [Figure 2]. The patient was referred to higher center for surgery where thoracotomy surgery had been done, and biopsy suggested the diagnosis of CCAM type 1.

Jagdish Prasad Goyal, Shishir Jindal, Mayank Mishra1, Bhanu Kiran Bhakhri2

Departments of Paediatrics and 1Pulmonary Medicine, All India Institute of Medical Sciences, Rishikesh, Uttarakhand, 2Department of Paediatrics, SSHPGTI, Noida, India

Received: 16 May, 2016. Accepted: 03 May, 2017.

Address for correspondence:
Dr. Jagdish Prasad Goyal,
Department of Paediatrics,
All India Institute of Medical Sciences,
Jodhpur, Rajasthan,
India.
E-mail: jpgoyal@rediffmail.com

How to cite this article: Goyal JP, Jindal S, Mishra M, Bhakhri BK. Congenital cyst adenoid malformation masquerading as bronchial asthma. Int J App Basic Med Res 2017;7:199-201.
Discussion

Congenital cystic adenomatoid malformation consists of hamartomatous or dysplastic lung tissue mixed with normal tissue which is generally confined to single lobe. This congenital pulmonary disorder occurs in approximately 1–4 in 100,000 births. The lesion probably results from an embryologic injury before the 35th day of gestation, with maldevelopment of terminal bronchiolar structures.[3] Ch’in and Tang in 1949 first described congenital cystic adenomatoid malformation as a distinct entity.[4] In 1977, Stocker et al. first classified CCAM into three subtypes[5] and in 2002 expanded this classification into 5 subtypes.[6] Type 0 (acinar dysplasia) is least common (<3%) and consists of microcystic disease throughout the lungs. It has the worst prognosis, and infant usually die at birth. Type 1 (60%) is macrocystic and consists of a single or several large (>2 cm in diameter) cyst lined with ciliated pseudostratified epithelium; the lesion is localized involving only a part of 1 lobe. One-third of the cases have mucus secreting cells. This type has good prognosis for survival. Type 2 (20%) is microcystic and consist of multiple cyst with histology similar to that of the type 1 lesion. Type 2 is associated with other serious congenital anomalies (renal, cardiac, diaphragmatic hernia) and carries a poor prognosis. Type 3 (<10%) is seen commonly in males, consist of microcyst and solid tissue with bronchiole like structure lined with cuboidal ciliated epithelium and separated by areas of nonciliated cuboidal epithelium. The prognosis for this type is also poor. Type 4 (10%) is commonly macrocystic and lacks mucus cells. It is associated with malignancy (pleuropulmonary blastoma) and can present either in childhood or in asymptomatic adults.[3]

Cystic airway malformation can be diagnosed in utero by ultrasonography.[7] Prenatal ultrasonography findings are classified as macrocystic (single or multiple cyst >5 mm) or microcystic (echogenic cyst <5 mm). Fetal cystic lung abnormalities can include CPAM (40%), pulmonary sequestration (14%), or both (26%), the median age at diagnosis is usually 21 weeks gestation.[8] Chest radiograph and CT chest constitute important diagnostic modality, but a definitive diagnosis can only be made after histopathological examination.

These lesions may be confused with a diaphragmatic hernia. Pulmonary sequestration, pleuropulmonary blastoma, bronchogenic cyst, congenital lobar emphysema, and cystic bronchiectasis also forms close differential diagnosis of CCAM.[9] Sarcomatous and carcinomatous degeneration[9] have been described in patient with congenital pulmonary airway malformation, so early recognition and surgical treatment of CCAM is essential to prevent the consequences of recurrent pulmonary infection and the potential risk of malignant transformation.

The patient can present in newborn period or early infancy with respiratory distress, recurrent respiratory infection, and pneumothorax. Patient with smaller lesions may remain asymptomatic until mid-childhood and can present later as recurrent or persistent pulmonary infection or chest pain. CCAM can also be complicated by secondary bacterial, mycotic or tubercular infection, and can present as lung abscess.[10] CCAM presenting as pneumatocele has also been reported by Anand et al.[11] Our case also presented with a recurrent cough which was treated with antibiotics, cough syrup, and bronchodilator. CCAM presenting as bronchial asthma has been rarely reported in literature and only one such case has been reported to the best of our knowledge.[12] Our case was highly unusual as it clinically presented as asthma and also showed bronchial obstruction on spirometry (FEV1 53% of predicted value), but due to the absence of bronchodilator reversibility, alternative diagnosis was suspected.

Financial support and sponsorship

Nil.
Conflicts of interest

There are no conflicts of interest.

References

1. Morotti RA, Cangiarella J, Gutierrez MC, Jagirdar J, Askin F, Singh G, et al. Congenital cystic adenomatoid malformation of the lung (CCAM): Evaluation of the cellular components. Hum Pathol 1999;30:618-25.
2. Morelli L, Piscioli I, Licci S, Donato S, Catalucci A, Del Nonno F. Pulmonary congenital cystic adenomatoid malformation, type I, presenting as a single cyst of the middle lobe in an adult: Case report. Diagn Pathol 2007;2:17.
3. Blatter JA, Finder JD. Congenital cystic malformation. In: Kliegman R, Stanton B, Geme JS, Schor N, editors. Nelson Textbook of Pediatrics. 20th ed. Elsevier, Philadelphia; 2016. p. 2057-8.
4. Ch’in KY, Tang MY. Congenital adenomatoid malformation of one lobe of a lung with general anasarca. Arch Pathol (Chic) 1949;48:221-9.
5. Stocker JT, Madewell JE, Drake RM. Congenital cystic adenomatoid malformation of the lung. Classification and morphologic spectrum. Hum Pathol 1977;8:155-71.
6. 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.
7. Singh M, Mitra S, Kumar L, Narang A, Rao KL, Kakkar N. Congenital cystadenomatoid malformation of lung. Indian Pediatr 2000;37:1269-74.
8. d’Agostino S, Bonoldi E, Dante S, Meli S, Cappellari F, Musi L. Embryonal rhabdomyosarcoma of the lung arising in cystic adenomatoid malformation: Case report and review of the literature. J Pediatr Surg 1997;32:1381-3.
9. Kaslovsky RA, Purdy S, Dangman BC, McKenna BJ, Brien T, Ilves R. Bronchioloalveolar carcinoma in a child with congenital cystic adenomatoid malformation. Chest 1997;112:548-51.
10. Dahabreh J, Zisis C, Vassiliou M, Arniotannaki N. Congenital cystic adenomatoid malformation in an adult presenting as lung abscess. Eur J Cardiothorac Surg 2000;18:720-3.
11. Anand M, Deshmukh SD, Naik A, Gaopande V. Congenital cystic adenomatoid malformation in an adolescent: An unusual presentation with pleural effusion and pneumatocele. Indian J Chest Dis Allied Sci 2011;53:173-6.
12. Indinnimeo L, De Vittori V, Venuta F, Vitolo D, Anile M, di Coste A, et al. A surprising finding in an adolescent athlete affected by diffuse congenital cystic adenomatoid malformation (CCAM). Clin Respir J 2013;7:420-2.