Case Report

Hybrid Pulmonary Sequestration, Cystic Pulmonary Adenomatoid Malformation, and Dextrocardia: A Triple Whammy

Abstract

Pulmonary sequestration and cystic pulmonary adenomatoid malformation are rare congenital cystic disorders of the lungs. The presence of both the diseases in the same individual is therefore very uncommon. Pulmonary sequestration is a nonfunctional pulmonary tissue mass that derives its blood supply from systemic blood supply other than pulmonary circulation. Congenital cystic pulmonary adenomatoid malformation represents a mass consisting of abnormal bronchiolar air spaces and a deficiency of functional alveoli. This is the case report of a 9-year-old girl with intermittent fever, left-sided chest pain, and cough for the past 15 days along with recurrent coughs since childhood suggestive of hybrid pulmonary sequestration, congenital cystic adenomatoid malformation, and dextrocardia.

Keywords: *Congenital anomaly, contrast-enhanced computed tomography thorax, dyspnea, lung mass, pulmonary sequestrations*

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Introduction

Hybrid congenital cystic lesions of the lung comprising both bronchopulmonary sequestration (BPS) and congenital cystic adenomatoid malformation (CCAM) are uncommon congenital malformations of the lung.

An unusual instance of BPS and cystic pulmonary adenomatoid malformation in a 9-year-old girl that mimicked pulmonary cancer is described. The patient complains of recurrent chest infections and a cough and cold. A chest X-ray showed opacity in the left lower lobe. A computed pulmonary tomography-aortogram showed that the lesion had a systemic arterial supply from the descending aorta and multiple cystic lesions. Diagnosis of pulmonary sequestration and cystic pulmonary adenomatoid malformation was made.

Case Report

A 9-year-old girl suffered from intermittent fever, left-sided chest pain, and cough for the past 15 days. She had complaints of recurrent coughs since childhood. On clinical examination, there were no significant findings but on chest examination, vesicular breath sounds were heard but a diminution of sound could be felt on the left infrascapular area. On radiological examination, the left lower zone of the X-ray chest showed a large intrathoracic mass and many tiny air-filled cystic air spaces [Figure 1]. Her past medical history revealed that the physician had started her on antituberculosis therapy despite a negative Mantoux test and there was no acid-fast bacilli (AFB) in her sputum.

A computed tomography (CT) scan thorax revealed a heterogeneous well-defined lesion 7.6 cm × 7.2 cm × 5.3 cm with multiple cystic lesions in the posterior segment of the left lower lobe. The largest cavity measuring 1.4 cm × 1.4 cm with air-fluid level and gross mediastinal shift dextrocardia was also noted [Figure 2]. Due to the persistent lesion and symptoms, a CT aortogram was done. It revealed systemic arterial supply of the lesion was from the descending aorta and venous drainage was to the left inferior pulmonary vein [Figure 3], which inclined our diagnosis toward BPS. We also went for bronchoscopy and lavage that revealed normal endobronchial and did not reveal any AFB, respectively.

Following this, surgery was planned and resection was done. Then, the sample was

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sent for histopathological examination which revealed areas of chronic inflammation with surrounding fibrosis and cystic degeneration that was suggestive of cystic adenomatoid malformation (CCAM) [Figure 4a and b].

Then we made the definitive diagnosis of bronchopulmonary sequestration and cystic adenomatoid malformation (CCAM) along with dextrocardia.

Discussion

Pulmonary sequestration is an infrequent respiratory disorder, consisting of nonfunctional and dysplastic bronchopulmonary tissue, that receives its blood supply from a systemic artery rather than a pulmonary arterial branch and has no relation with the tracheobronchial tree.[1-4]

There are two forms of pulmonary sequestration: intralobar and extralobar. Extralobar sequestration has its own pleura, whereas intralobar sequestration is surrounded by normal lung tissue. The more prevalent kind, intralobar pulmonary sequestration, affects older children and teenagers. Although intralobar pulmonary sequestration usually has only one feeding artery, numerous systemic arterial supplies can be found. The thoracic aorta is the most common source of arterial supply for pulmonary sequestration, followed by the abdominal aorta, the intercostal artery, phrenic artery, subclavian artery, pulmonary artery, left gastric artery, coronary artery, and celiac trunk. The thoracic aorta is the most common source of arterial supply for pulmonary sequestration.[2] Congenital Cystic adenomatoid malformation (CCAM) is the most common type of congenital cystic lung disease. According to Kanavi *et al*. the incidence of CCAM is 1:10,000–1:35,000 live births. It can be detected by prenatal screening at 18–20 weeks of gestation.[5] CCAM is also categorized under many subtypes: Type 0, in which tumor develops from the trachea and this is the rarest type, Type 1 is the most common comprising 50%–70% of the total cases and develops from distal bronchus or proximal bronchioles and Type 2 arises from terminal bronchioles also represent a higher risk of associated anomalies. Type 3 generally represents acinar-like tissue and Type 4 presents as large cysts and have alveolar origin.[5]

It is believed that a hamartomatous change in the tertiary bronchioles causes congenital cystic adenomatoid malformation (CCAM), which affects 1 in 25,000–35,000 live births. A zone of lung parenchyma with aberrant systemic blood flow and no normal contraction to the tracheobronchial tree is known as BPS. Incidence in the general population is estimated to be between 0.15% and 1.7%.[6] Two uncommon congenital lung cystic abnormalities are BPS and congenital cystic adenomatoid

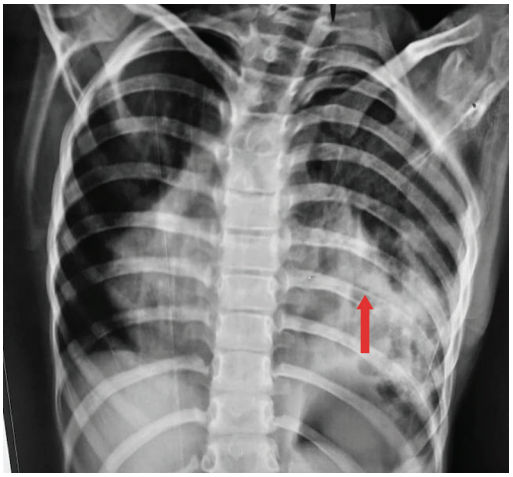

Figure 1: Chest X-ray shows a large intrathoracic mass on the left side with many tiny air-filled cystic spaces (Red Arrow)

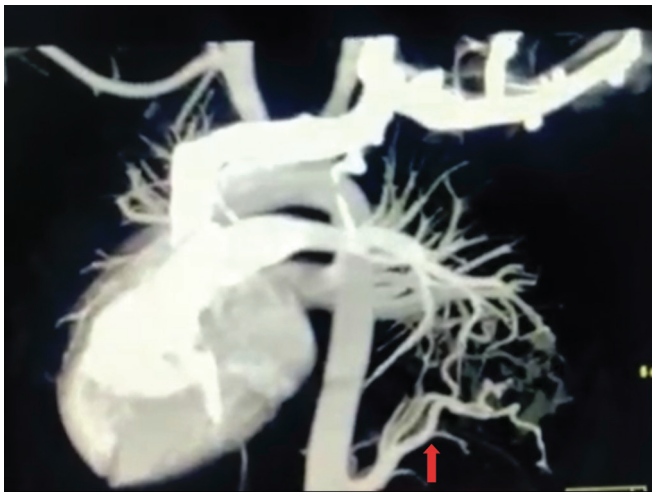

Figure 3: Computed tomography pulmonary angiography shows the systemic arterial supply of the lesion from the descending aorta and venous drainage to the left inferior pulmonary vein (Red Arrow)

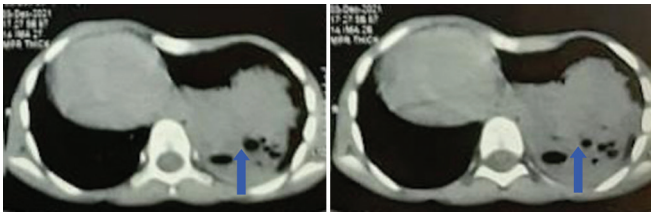

Figure 2: Computed tomography scan thorax revealed a heterogeneous well-defined lesion 7.6 cm × 7.2 cm × 5.3 cm heterogeneous well-defined lesion with multiple cystic lesions in the posterior part of the left lower lobe (Blue Arrow)

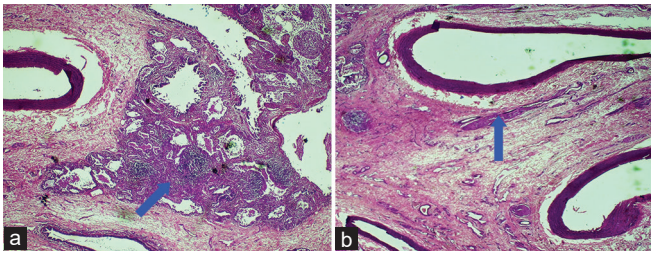

Figure 4: (a) Photomicrograph shows areas of chronic inflammation with surrounding fibrosis and cystic degeneration (H and E, ×40). (b) Photomicrograph shows an aberrant systemic artery with an elastic layer along with areas of fibrosis (H and E, ×40) (Blue Arrow)

malformation (CCAM).[7] Rarely, a small number of congenital cystic lesions have been described as hybrids of the BPS and CCAM. Only 36 hybrid cases of CCAM and BPS have been documented in English literature so far, making them extremely uncommon.[8] From 1995 to 2008, a study carried out in Taiwan revealed that the incidence of mixed-type hybrid lesions was 19%.[9] They emphasized that the growing percentage of these lesions might point to a comparable developmental etiology.[10] The current treatment for symptomatic pulmonary sequestration is mainly surgical resection. Intralobar pulmonary sequestration in asymptomatic patients is also treated by surgery to prevent death at later stages of life because of abundant hemoptysis.[11]

In 2002, Wan *et al*. described for the first time video-assisted thoracic surgery lobectomy for the management of BPS and this technique is widely being used nowadays.[12]

CT scan thorax can misguide the clinicians toward malignancy so, it is important that clinicians should always keep the possibility of cystic disorders of lungs. CT angiography is the noninvasive diagnostic method of preference, which can show both the aberrant lung parenchyma and one or more faulty arterial supplies to the sequestration.[13]

Conclusion

Intrathoracic mass-like lesions with cystic changes give an impression of malignancy. Further investigation leads to the diagnosis of hybrid lesion sequestration with congenital cystic pulmonary adenomatoid malformation. In our case, dextrocardia was present, but two-dimensional echocardiography was suggestive of situs solitus. Hence, dextrocardia is due to the mass effect of the lesion; however, the patient had no symptoms related to this.

This case highlights that congenital pulmonary abnormalities should be considered when generating a differential diagnosis for patients with cystic intrathoracic mass lesions and respiratory symptoms. To prevent infections and lower the risk of cancer, surgical intervention should be performed for a definitive diagnosis and course of therapy, and CT angiography should be the preferred noninvasive diagnostic technique.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Savic B, Birtel FJ, Tholen W, Funke HD, Knoche R. Lung sequestration: Report of seven cases and review of 540 published cases. Thorax 1979;34:96-101.
2. Wei Y, Li F. Pulmonary sequestration: A retrospective analysis of 2625 cases in China. Eur J Cardiothorac Surg 2011;40:e39-42.
3. Sfakianaki AK, Copel JA. Congenital cystic lesions of the lung: Congenital cystic adenomatoid malformation and bronchopulmonary sequestration. Rev Obstet Gynecol 2012;5:85-93.
4. Huang CC, Ko SF, Chung MY, Shieh CS, Tiao MM, Lui CC, *et al.* Infradiaphragmatic pulmonary sequestration combined with cystic adenomatoid malformation: Unusual postnatal computed tomographic features. Abdom Imaging 2004;29:439-42.
5. Kanavi JV, Thangaraj J, Thomas A. Congenital cystic adenomatoid malformation (CCAM): Antenatal and postnatal management'. Int J Reprod Contracept Obstet Gynecol 2017;6:2447. Available from: https://link.gale.com/apps/doc/A534838501/HRCA?u=anon~956256ad&sid=googleScholar&xid=607b0ee0. [Last accessed on 2022 Nov 17].
6. Gornall AS, Budd JL, Draper ES, Konje JC, Kurinczuk JJ. Congenital cystic adenomatoid malformation: Accuracy of prenatal diagnosis, prevalence and outcome in a general population. Prenat Diagn 2003;23:997-1002.
7. Gajewska-Knapik K, Impey L. Congenital lung lesions: Prenatal diagnosis and intervention. Semin Pediatr Surg 2015;24:156-9.
8. Cass DL, Crombleholme TM, Howell LJ, Stafford PW, Ruchelli ED, Adzick NS. Cystic lung lesions with systemic arterial blood supply: A hybrid of congenital cystic adenomatoid malformation and bronchopulmonary sequestration. J Pediatr Surg 1997;32:986-90.
9. Chen HW, Hsu WM, Lu FL, Chen PC, Jeng SF, Peng SS, *et al.* Management of congenital cystic adenomatoid malformation and bronchopulmonary sequestration in newborns. Pediatr Neonatol 2010;51:172-7.
10. Chauhan VH, Taksande AM, Vilhekar KY. Congenital cystic adenomatoid malformation. NIJP 2012;1:123-6.
11. Qian X, Sun Y, Liu D, Wu X, Wang Z, Tang Y. Pulmonary sequestration: A case report and literature review. Int J Clin Exp Med 2015;8:21822-5.
12. Wan IY, Lee TW, Sihoe AD, Ng CS, Yim AP. Video-assisted thoracic surgery lobectomy for pulmonary sequestration. Ann Thorac Surg 2002;73:639-40.
13. Stocker JT. Cystic lung disease in infants and children. Fetal Pediatr Pathol 2009;28:155-84.