

Case Report

Congenital Pulmonary Airway Malformation with Sequestration - Hybrid Lesion - Presenting As a Non-Resolving Pneumonia: A Case Report with Radiological Review

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Received: 16/12/2015

Revised: 28/12/2015

Accepted: 01/01/2016

ABSTRACT

Congenital lesions of lung are rare, with overall incidence of 1/10000 to 1/25000 births (2.2 % as compared to acquired lesions). CPAM and BPS are the commoner lesions. Hybrid lesion (coexistent CPAM and BPS) are extremely rare. Diagnosis can be made antenatally in-utero or postnatally due to complications of the lesion, such as recurrent pneumonias, lung abscesses and pneumothorax. We report a case of a 4 year old male child presenting with recurrent pneumonia. Chest radiograph showed dense consolidation in the left lower zone and multiple small air filled cystic spaces in the left mid and lower zones, few showing air-fluid levels. HRCT showed dense consolidation in the left lower lobe with air-bronchogram and small air-filled cavities. Contrast study showed two arterial feeders arising from descending thoracic aorta with venous drainage into left inferior lobe pulmonary vein. A diagnosis of lung sequestration with associated CCAM was made.

Keywords: CCAM, CPAM, Sequestration, Hybrid lesion.

INTRODUCTION

Congenital lesions of lung are rare, with overall incidence of 1/10000 to 1/25000 births (2.2 % as compared to acquired lesions). CPAM and BPS are the commoner lesions. Hybrid lesion (coexistent CPAM and BPS) are extremely rare. Diagnosis can be made antenatally in-utero or post-natally due to complications of the lesion, such as recurrent pneumonias, lung abscesses and pneumothorax.

Congenital pulmonary airway malformations (CPAM) are a heterogeneous group of cystic and non-cystic lesions of the lung which result from maldevelopment of the early airways. [1] It is a hamartomatous lesion which contains tissues from different pulmonary origins. It

is characterised by excessive proliferation of the conductive airways with varying types of epithelial lining. The lesions are mainly cystic and intrapulmonary, usually unilobar with a slight predilection for the lower lobes of the lung. [2] Incidence ranges from 1-11000 to 1-30000 live births, with increasing incidence in mid trimester with spontaneous resolution [3] previously; this entity was known as congenital cystic adenomatoid malformation (CCAM).

CASE REPORT

A 4 year old male patient presented with on and off fever since 15 days, left sided chest pain and cough. X ray chest showed dense consolidation in the left lower zone and multiple small air filled

cystic spaces in the left mid and lower zones, few of which were showing air-fluid levels (figure 1). Previous history of admission for recurrent pneumonitis was noted 6 months ago. HRCT was done taking 1 mm thick axial sections in helical mode followed by contrast study. It showed dense consolidation in the left lower lobe with air-bronchogram and small air-filled cavities. Multiple small fluid-filled cavities/abscesses of size approx. 2-10 mm were seen (figure 2). Multiple air-filled, thin walled cysts of size ranging from 0.5 cm to 2 cm were noted in apical, postero-basal and lateral-basal segments of left lower lobe (figure 3). Two arterial feeders were noted arising from lateral and postero-lateral aspect of adjoining portion of lower descending aorta and entering the region of dense consolidation (figure 4A). Venous drainage from the dense consolidation was into the left inferior lobe pulmonary vein (figure 4B). Hence a diagnosis of lung sequestration of intralobar type with associated CCAM (Stocker type II) was made. Haematological work up revealed leucocytosis with TLC 18,500 with neutrophilia (83 %). Patient was given a course of antibiotics and referred to the paediatric surgeon. The patient underwent left lower lobe lobectomy. Biopsy was confirmatory of CCAM with sequestration (figures 5A and B).



Figure 1: Radiograph chest showing dense consolidation in left lower zone with multiple air filled cystic spaces in left mid and lower zones.



Figure 2: NCCT thorax axial view (mediastinal window) showing multiple air filled cystic spaces in left lower lobe with surrounding dense consolidation.

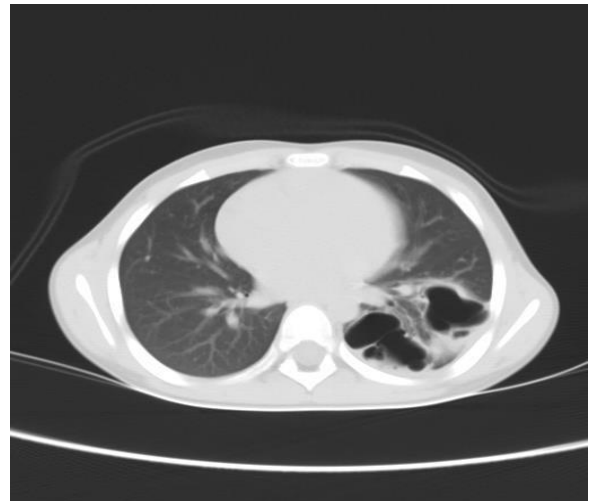


Figure 3: CT thorax axial view (lung window) showing multiple thin walled air filled cysts in left lower lobe with associated dense consolidation.



Figure 4A: CECT thorax axial view (mediastinal window) showing two arterial feeders arising from the descending thoracic aorta.



Figure 4B: CECT thorax MIP oblique-coronal view (mediastinal window) showing venous drainage from the BPS to left pulmonary vein.

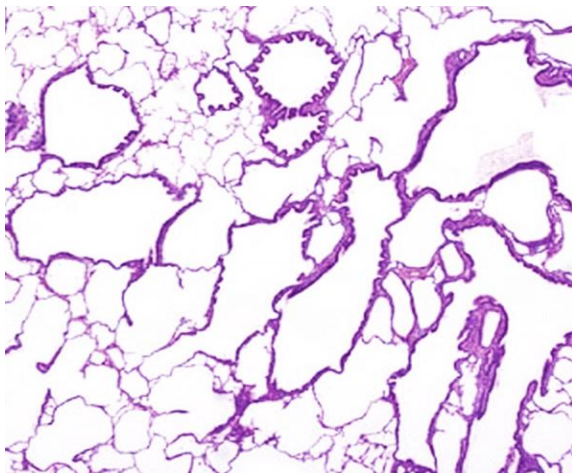


Figure 5A: Tissue sample with Hematoxylin Eosin stain (x 100) shows distorted lung architecture due to multiple irregularly shaped bronchiole-like structures which are lined by ciliated, columnar to cuboid epithelium forming cystic spaces suggestive of CPAM. Larger cysts are lined by smooth muscle layer beneath epithelium. Distal air spaces are lined by flat epithelium.

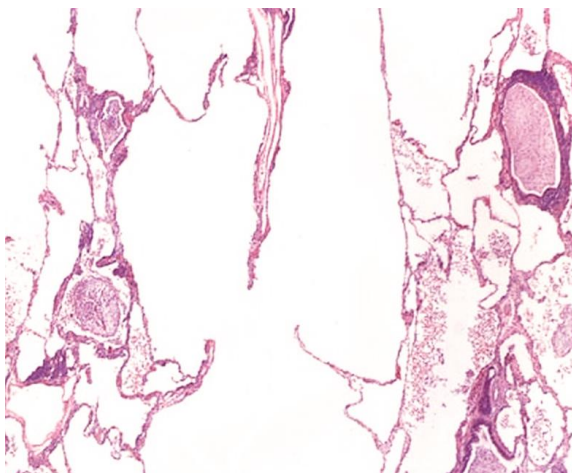


Figure 5B: Tissue sample with Hematoxylin Eosin stain (x 100) showing distended respiratory bronchiole and dilated alveolar spaces with preservation of normal lung architecture with superimposed patchy inflammatory exudate and interstitial inflammation, suggestive of sequestration.

DISCUSSION

CPAM may communicate with proximal airways, through this communication is abnormal. [1] Most of the CPAMs derive their blood supply from the pulmonary artery, and drain via pulmonary veins. Exception is of hybrid lesions, which have systemic blood supply. CPAM is now preferred over CCAM since the lesions are cystic in only 3 of the 5 types of these lesions and adenomatoid in only one type.

Classifications for CPAM are based on embryological level of origin and histological features and prenatal ultrasound features.

Classification system for CPAM

I – Based on embryological level of origin and histological features by Stocker et al. [4]

0. It is the rarest form and arises from trachea/bronchus. It involves the entire lungs. Cysts are small. Presentation is severe and lethal.

1. Commonest, representing 50-70 % of all cases, arises from distal bronchus or proximal bronchiole. It shows multiple small number of large cysts, of size 2-10 cm. A single dominant cyst may be seen. Cyst walls are thin, lined by ciliated pseudo-stratified epithelium. Other cell types like cartilage may be found between the cysts. Due to the large size, these CCAMs may cause significant mass effect, which may cause hydrops fetalis

2. Constitute 15-30 % of all cases, arises from the terminal bronchioles. They are composed of smaller cysts, of size 0.5 – 2 cm, with solid areas which are difficult to differentiate from surrounding tissue. They are lined by ciliated columnar or cuboidal epithelium with elements of bronchioles or alveoli. Cysts are more evenly spaced than in type I. Associated abnormalities are seen in up to 60% cases including most organ systems.

3. They account for 5-10 % of cases, arising from acinar type of tissue. They are composed of small cysts; hence the mass appears echogenic and solid on USG. The

tissue is acinar with adenomatoid elements, arising from distal airways.

4. They account for 5-15 % of cases and are alveolar in origin with unlined cysts. They contain large cysts that may be as large as 10 cm and may be associated with malignancy, especially pleuro-pulmonary blastoma.

Types 1, 2 and 4 are classified as macrocystic or both macrocystic and microcystic. Type 3 is microcystic.

Only 3 types of CPAM are distinguished on imaging –

Type 1 – large cyst CPAM

Type 2 – small cyst CPAM

Both of which are macrocystic CPAMs.

Type 3 – solid type CPAM which is microcystic

Type 4 CPAMs are indistinguishable from cystic pleuropulmonary blastoma on imaging and require HPE for diagnosis.

II – Prenatal USG classification of Adzick [5]

A. Macrocystic – cysts > 5 mm

B. Microcystic – cysts < 5 mm.

Lung dysplasias are classified based on pathological features and in utero appearances.

I – Pathological classification proposed by Langston. [6]

1) Bronchopulmonary malformation

a) Bronchogenic cyst (noncommunicating bronchopulmonary foregut malformation)

b) Bronchial atresia

i. Isolated

ii. With systemic arterial/venous connection (intralobar sequestration)

iii. With connection to gastrointestinal tract (intralobar sequestration/complex or communicating bronchopulmonary foregut malformation)

iv. Systemic arterial connection to normal lung

c) CCAM: large cyst type (Stocker type 1)

i. Isolated

ii. With systemic arterial/venous connection (hybrid lesion/intralobar sequestration)

d) CCAM: small cyst type (Stocker type 2)

i. Isolated

ii. With systemic arterial/venous connection (hybrid lesion/intralobar sequestration)

e) Extralobar sequestration

i. Without connection to gastrointestinal tract (with/without CAM, small cyst type)

ii. With connection to gastrointestinal tract (complex/communicating bronchopulmonary foregut malformation)

2) Pulmonary hyperplasia and related lesions

a) Laryngeal atresia

b) Solid or adenomatoid form of CCAM (Stocker type 3)

c) Polyalveolar lobe

3) Congenital lobar overinflation

4) Other cystic lesions

a) Lymphatic/lymphangiomatous cysts

b) Enteric cysts

c) Mesothelial cysts

d) Simple parenchymal cysts

e) Low-grade cystic pleuropulmonary blastoma

II – In utero classification by Achiron et al [7]

Each unit of embryonic lung has a bronchial bud supplied by systemic capillary plexus derived from primitive aorta and containing a small branch of pulmonary artery. As the growing lung advances and pulmonary artery develops, the systemic plexus regresses. Hence, lung dysplasias are divided into 5 categories, considering each lung component involved in pathology.

1. Agenesis of lung

Most severe dysplasia.

Insult is early and proximal to bronchial bud, to arrest lung / lobe / lobule development.

2. Normal lung with abnormal vascular supply

Bronchial tree continues to develop normally, pulmonary artery growth is arrested. Hence, normal lung with isolated segment is supplied by a persistent systemic vessel instead of pulmonary artery.

3. Abnormal lung with abnormal vascular supply

Includes BPS with disruption of both airway tract and pulmonary artery development with resultant systemic arterial supply to abnormal lung parenchyma

4. Abnormal lung with normal vascular supply

Includes bronchogenic cyst and classic CCAM

5. Miscellaneous

Includes laryngeal atresia with bilateral echogenic lungs (CHAOS syndrome), split notochord syndrome, bronchopulmonary dysplasia with neuro-enteric association

Advantage of this new classification system is that it includes each foetal lung component involved in the anomaly – 1, parenchyma 2, arterial circulation, 3, venous drainage. It is based on prenatal sonographic identification and Doppler flow studies – antenatal supply and venous drainage of lesions as opposed to tissue consistency.

Malignant transformation of CCAM [2]:

Bronchiolo-alveolar carcinoma (BAC) and rhabdomyosarcoma in association with CCAM have been reported in children and adults. Hence, along with possibility of lung infection, surgical resection in all cases of CCAM is recommended.

BPS [2]: It constitutes 10-30 % of congenital cystic lung lesions. These are solid non-functioning congenital lung lesions which derive blood supply from aorta rather than pulmonary artery, with absence of communication with the bronchial tree. In comparison to CCAM,

BPS is rarer, with no published population incidence. CCAM and BPS abnormalities occur during branching and proliferation of the bronchial structures. This insult occurs during pseudo-glandular phase of lung development from 7-17 weeks. Both the lesions have malignant potential. [8] BPS is made up of extraneous and non-functioning lung parenchyma, which has separated from normal pulmonary structure.

It is subdivided into intra-lobular and extra-lobular types.

a) Intra-lobular

It is the more common type, accounting for 75 % of all BPS. They are located within the substance of lung parenchyma with the same pleural investment. They are limited to the lung. They usually affect the left lower lobe, typically the posterolateral segment of left lower lobe. Arterial supply most commonly arises from the thoracic aorta and sometimes from the abdominal aorta, intercostal vessels and brachio-cephalic vessels. Venous drainage is to the pulmonary veins or rarely to the hemiazygous vein. It can present as a non-resolving pneumonia, lung abscess or rarely as haemoptysis. On X ray chest, they appear as non-aerated atelectatic mass. CECT shows blood supply from a systemic artery. Treatment is surgical.

b) Extra-lobular

They account for 25 % of all BPS. [9] They have their own pleural investment. They are not limited to the lung, and can occur in the chest, pericardial sac or in the abdomen. 90% of these lesions are found in left lower chest, of which 80-90 % are above the diaphragm and the remaining below. Blood supply is mainly from the thoracic aorta with venous drainage mostly systemic – either azygous or hemiazygos or portal, as opposed to intralobular. Associated abnormalities are seen in up to 50% cases and include diaphragmatic hernia, cardiac defects, AV malformations and other abnormalities. Prenatal scan can demonstrate the aberrant blood supply.

Chest X-ray shows left posterior mediastinal mass. CECT is confirmatory.

Differences between CCAM and BPS can be summed up in table 1 [10]

Table 1: Differences between CCAM and BPS.

	CCAM	BPS	
		INTRALOBAR	EXTRALOBAR
Incidence	1/11,000-1/35,000	Rare	
Vascular Supply	Pulmonary	Systemic: lower thoracic or upper abdominal aorta	Systemic: lower thoracic
Laterality	80%-95% unilateral, either lobe	60% left sided	90% left sided
Sex	Male > Female	Male = Female	Male > Female
Tracheobronchial communication	Present, constricted/anomalous	None	None
Associated anomalies	Rare except for Type 2 (60%)	17%	40%

Hybrid lesions

Hybrid lesions are extremely rare and have features of both CPAM and BPS. This suggests a common developmental origin for both and perhaps each represent two ends of a broad spectrum of pathologies. [2,11] Theories of their pathogenesis include abnormal proliferation of tissues, dysplasia and metaplasia of normal tissues and airway obstruction.

Prognostication

a) CPAM volume ratio (CVR)

Ratio of volume of mass / Head circumference (HC) is used to normalise the size of mass to foetal size. A CVR of > 1.6 is associated with hydrops with poor outcome and is an indication for foetal surgery in many centres. Foetuses with CVR < 1.6 are treated conservatively with close foetal monitoring for complications. Antenatal steroids are administered to reduce the size of lesions and delay progression, thus reducing the number of cases needing foetal surgery. Foetal surgery is associated with increased maternal and foetal mortality. [1]

Hydrops develops more commonly in CCAM than in BPS, with reported rates up to 40 %. Hydrops is more commonly seen in microcystic CCAM, CCAM with dominant cyst and CCAM with higher volume as measures in CVR. Hydrops is unlikely to develop before 28 weeks as natural growth of CCAM plateaus by 25 weeks. Open surgery is reserved for cases with poorest prognosis and those prior to 32-34 weeks gestation. [10]

b) Quantitative measurement of mass size can be done by mass-thorax ratio. If it is > 0.56, it indicates poorer outcome.

A fast growing CPAM causes mediastinal shift with resultant development of polyhydramnios and hydrops. Established indicators of poor prognosis are large lesions, bilateral lung involvement and hydrops. [1]

CONCLUSION

CPAM and BPS can co-exist together (hybrid lesion). They share same developmental ancestry and most likely represent a similar pathogenesis. This case report with radiological review illustrates various classification systems for congenital pulmonary diseases, diagnostic evaluation of hybrid lesion and its prognosis.

Conflict of Interest: The authors declare that there is no conflict of interest regarding the publication of this paper.

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How to cite this article: Khaladkar SM, Kalra R, Garg SV et al. Congenital pulmonary airway malformation with sequestration - hybrid lesion - presenting as a non-resolving pneumonia: a case report with radiological review. *Int J Health Sci Res.* 2016; 6(1):609-615.
