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CONGENITAL CYSTIC ADENOMATOID MALFORMATION OF LUNGS- Study Of 20 Cases

Pathology

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ABSTRACT

BACKGROUND: Congenital cystic adenomatoid malformations (CCAM) of lung are rare congenital cystic lung lesions. It is a developmental non hereditary hamartomata's abnormality of lung with unknown aetiology. It is a rare lesion with incidence of 1 in 25,000 to 1 in 35,000 pregnancies . **MATERIALS AND METHODS:** Present study comprises of 600 cases of perinatal autopsies from 2017 to 2021, out of which CCAM was seen in 20 cases. **RESULTS:** Total number of cases studied were 20 in which type I -5cases, Type II - 2cases, Type III-11 cases and Type IV -2 Cases. **CONCLUSION :** Although CCAM can be diagnosed by ultrasonographic evaluation, foetal lung mass size and foetal echocardiography, fetal autopsy helps to identify and study various types of CCAM in helps to identify and study various types of CCAM in detail and helps in preconceptional genetic counselling, therefore fetal autopsy plays vital role in conforming the congenital malformations and future recurrences.

KEYWORDS

Congenital cystic Adenomatoid malformation, fetal autopsy, cystic lesion of lung.

INTRODUCTION

Congenital cystic adenomatoid malformations (CCAM) is a rare congenital cystic lung lesions which occur due to excessive proliferation of tubular bronchial structures & Its incidence is 1 in 25,000 to 1 in 35,000 pregnancies[1]. It is characterized as benign hamartomata's or dysplastic lung tumors due to overgrowth of terminal bronchioles & reduction in the number of alveoli It affects mainly airways, vasculature and, parenchyma lead to aberrant embryological lung development which occur at different stages of intrauterine life. CCAM was first described in 1949 & In 1977 It was classified into 3 subtypes and In 2002 expanded into five types and renamed as congenital pulmonary airway malformation (CPAM) by Stocker. Eighty percent of the lesions are recognized in neonatal period[2]; Though USG is a valuable examination tool, our study access value of fetal autopsy after the termination of a pregnancy which is essential to confirm the finding and to arrive at a definite diagnosis and further management[3].

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MATERIALS AND METHODS

The present study comprises perinatal autopsies conducted in pathology department of tertiary care centre during nov2017 to oct 2021.Ethical clearance was obtained from the Institutional Ethical Committee. Foetal autopsies were performed after taking informed consent of parents. Standard protocol was followed. Anthropometric data and photographs were recorded for each fetus. External examination for deformities was done. Internal examination for gross organ anomaly was done by sectioning of each organ for histopathological examination.

RESULTS:

A total of 600 fetal autopsies were conducted from November 2017 to october 2021, out of which 20 cases were of CCAM, which accounts for 3% of cases. All these cases were of intra-uterine death (induced or spontaneous). The age of foetuses ranged from 17-30 weeks of gestation. Most of cases where in the age group of 20-30 weeks [Table 1]. The weight of the fetus was in the range of 500gms to 3200gms with majority weighing from 1500 to 2000gms[Table 2]. Twenty cases with all four types of CCAM were noted in this study and findings of which has been summarized [Table 3 -6 and Figures 1-7]

Table 1: Gestational age and number of cases.

Gestational age(weeks)	No. of cases
10-20	4
20-30	11
30-40	5
Total	20

Table 2: Foetal weight and number of cases.

Weight (gms)	No. of cases
500-1000	1
1000-1500	2
1500-2000	8
2000-2500	6
2500-3000	2
3000-3500	1
Total	20

Table 3: Cases of type 1 CCAM

SL.No	Age of mother	Parity	Gestational age	Usg findings	Weight & Sex	Gross	Microscopy	Associated anomaly	CCAM Type
Case1	22yr	G3A2	30wk	oligohydramnios , still born, B/L Multiple air filled cysts	600 gm, male	Left lung multiple cysts	Cyst lined by flattened epithelium to stratified epithelium	-	I
Case2	29yr	G3P2L2	32wk	B/L Multiple air filled cysts	600gm, male	B/L lung cysts	Cyst lined Columnar epithelium to stratified epithelium	-	I
Case3	26yr	Primigravid a	36wk	B/L Multiple air filled cysts	3.2kg , male	B/L lung cysts of different cyst.	Cyst lined Columnar epithelium to stratified epithelium	-	I
Case4	33yr	G3P2L2	23wk	Oligohydramnios , B/L Multiple air filled cysts	400 gm, male	B/L lung cysts	Cyst lined Columnar epithelium to stratified epithelium	Single umbilical artery	I
Case5	25yr	Primigravida	36wk	Oligohydramnios, B/L Multiple air filled cysts	3.4kg, male	B/L lung cysts	Cyst lined Columnar epithelium to stratified epithelium Partially aerated alveoli	-	I

Table 4:Cases of type II CCAM

SL.No	Age of mother	Parity	Gestational age	Usg findings	Weight & Sex	Gross	Microscopy	Associated anomaly	CCAM Type
Case1	21yr	Primi with twin pregnancy	25wk with IUD	Hypoechoic lesion in right lung.	250gm ,Female	Right lung enlarged with multiple small cyst	Cyst lined by cuboidal epithelium	-	II
Case2	26yr	-	-	Multiple air filled cysts in right lung	1.7kg,male	B/L lungs with multiple cysts	Cyst lined by cuboidal epithelium	Both kidney multiple cyst with dysplasia	II

Table 5:Cases of type III CCAM

	Age of mother	Parity	Gestational age	USG finding	Weight and sex	Gross	Microscopy	Associated anomaly	CCAM Type
Case1	25yr	G6P3L3A 2	25wk with IUD	Hypoechoic lesion in right lung	250gm ,Female	Right lung enlarged with multiple small cyst	Cyst lined by cuboidal epithelium	-	III
Case2	27yr	primigravida	28wk	B/L Hypoechoic lesion	1kg, Male	B/L lung cysts	Cyst lined by cuboidal epithelium	-	III
Case3	23yr	G2A1	39wk	Oligohydramnios, B/L Multiple air filled cysts	2.4kg, Male	B/L lung cysts	Cyst lined by cuboidal epithelium	-	III
Case4	22Yr	Primigravida	22wk	Multiple air filled cysts in right lung	400gm, Male	B/L lung cysts	Cyst lined by cuboidal epithelium	-	III
Case5	23Yr	G2P1L1	23wk	Hypoechoic lesion in right lung	250, Female	B/L lung cysts	Cyst lined by cuboidal epithelium	-	III
Case6	28yr	G3P2L2	25wk	Multiple air filled cysts in right lung	500gm, Male	B/L lung cysts	Cyst lined by cuboidal epithelium	-	III
Case7	30yr	G2A1	21wk	B/L Hypoechoic lesion	400gm, Female	B/L lung cysts	Cyst lined by cuboidal epithelium	-	III
Case8	27yr	Primigravida	22wk	Oligohydramnios, B/L Multiple air filled cysts	350gm, Female	B/L lung cysts	Cyst lined by cuboidal epithelium	-	III
Case9	28yr	Primigravida	30wk	B/L Hypoechoic lesion	700gm, Male	B/L lung cysts	Cyst lined by cuboidal epithelium	-	III
Case10	31yr	G3P1L1A1	24wk	Multiple air filled cysts in right lung	700gm, Female	B/L lung cysts	Cyst lined by cuboidal epithelium	-	III
Case11	23yr	G2P0A1	19wk	Oligohydramnios, B/L Multiple air filled cysts	450gm, Male	B/L lung cysts	Cyst lined by cuboidal epithelium	-	III

Table 6:Cases of type IV CCAM

SL.No	Age of mother	Parity	Gestational age	Usg findings	Weight & Sex	Gross	Microscopy	Associated anomaly	CCAM Type
Case1	23yr	G3P2L2	22wk+3day	oligohydramnios with hypoechoic right lung	350 gm, male	Right lung multiple cysts	Cyst lined by flattened epithelium	-	IV
Case2	21yr	G2P1L1	22wk	B/L Hypoechoic lesion	500gm, male	B/L lung cysts	Cyst lined by flattened epithelium	-	IV

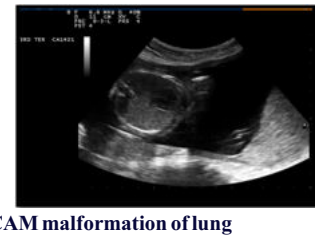

Fig1:USG: CCAM malformation of lung

Fig2: GROSS: Nodular surface noted

Fig3:Cut section-Multiple cyst of varying size seen in both lungs

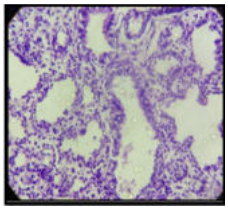

Fig4: CCAM Type I:H &E(400X) Cyst lined by flattened epithelium to stratified epithelium

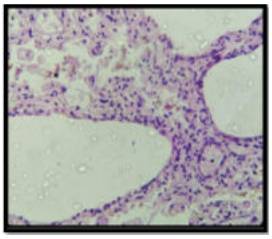

Fig 5: CCAM TYPE II:H &E(400X) Cyst lined by cuboidal to flattened epithelium.

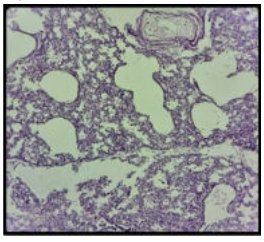

Fig 6: CCAM TYPE III:H&E(100X) Cyst lined by flattened epithelium

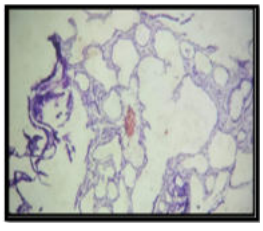

Fig 7: CCAM TYPE IV(100X) Cyst lined by flattened epithelium

DISCUSSION:

In the present study, total of 600 cases of perinatal autopsies, conducted during November 2017to October 2021 were studied. Out of which 20 were having rare CCAM. In that most common was type III. It was first described by Chi'n Tang in 1949. It is usually unilateral or maybe bilateral occurs mainly due to excessive proliferation of lobular bronchial structures during foetal development. It is usually a unilateral condition, but can involve both the lungs. It is divided into 5 types based on the anatomical & development changes of human lung 1)embryonal (3-7 weeks), 2)pseudo glandular (7-17 weeks), 3)canalicular (17-29 weeks), 4)saccular (24-36 weeks), and 5) alveolar (36 weeks to maturity)[4].Most common CCAM develops during the pseudo glandular and saccular period (7-35 weeks). In 1977 Stocker et al subdivided CCAM into three subtypes. Type I lesion constitute 50-70% and is composed of single or multiple large cysts (> 2 cm) lined by flattened(FIG4), cuboidal cells frequently producing mediastinal herniation. The walls of the cysts contain prominent smooth muscle and elastic tissue. Occasionally mucus producing cells are seen and presence of cartilage is extremely rare. Mucin production is unique to type I lesion. Type II lesion constitute 15-30% and are composed of multiple small cysts < 2 cm), lined by ciliated cuboidal to columnar epithelium(FIG5), structure resembling that of respiratory bronchioles, and distended alveoli are present between the epithelium lined cyst. Mucus cells and cartilage are not seen. This type is usually associated with other systemic anomalies. Type III lesion constitutes 5-10% and are usually large bulky non cystic lesions producing mediastinal shift[5]. Bronchial-like structures are lined by ciliated cuboidal epithelium and separated by masses of alveolus-sized structures by non ciliated cuboidal epithelium[1](FIG6), In 2002 Stocker,modified this classification by adding two more types (type 0 and IV) and renamed the lesion as CPAM, type 0 of tracheobronchial origin has solid appearance with small and firm lungs; and microscopically shows bronchiolar type airway with cartilage, smooth muscle, and glands separated by abundant mesenchymal tissue[6]. Type IV of distal acinar origin, has peripheral cystic type, large cysts (> 10 cm) lined by flattened epithelium and resting on loose mesenchymal tissue(FIG7)[2].

CONCLUSION:

CCAM is a rare developmental malformation of lung. It is usually diagnosed in fetal or neonatal life. Even though the prenatal ultrasonogram reasonably predicts the malformations, fetal autopsy is essential to look for additional malformations and to confirm the diagnosis. Proper antenatal check ups, maternal nutrition ,and pre conceptional genetic counselling plays a vital role in reducing the congenital malformations and future recurrences. We report these cases for their rarity.

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