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Original Research Paper

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NEWBORN WITH RESPIRATORY DISTRESS DUE TO CONGENITAL CYSTIC ADENOMATOID MALFORMATION.

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ABSTRACT Congenital cystic adenomatoid malformation is a rare, usually unilateral entity of the lung. It was first reported in the year 1787. It is marked by presence of multiple cysts with normal lung tissue in between. It is mostly diagnosed antenatally. This condition may present as respiratory distress at birth, or recurrent chest infections during early years of life. Associations with other congenital anomalies may be seen. Fetal hydrops, cardiac and skeletal anomalies, potters syndrome, gastrointestinal atresia are common co-findings.

KEYWORDS :

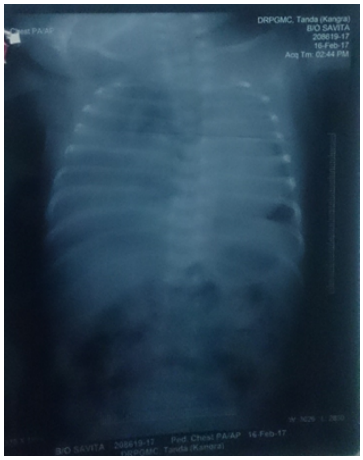
INTRODUCTION

Congenital cystic adenomatoid malformation is defined as condition with a mass of cysts lined by proliferating bronchial or cuboidal epithelium with intervening normal portions of lung. Its incidence varies from 1 in 25000 to 1 in 35000.[1] Approximately 25 % of affected infants are still birth while 20 % may have other associated anomalies. It may be associated with other pulmonary conditions like sequestration. Among cardiac associations Tetrology of fallot is the most common.

CASE REPORT

We present a case of a newborn whose mother was referred to our tertiary care center for delivery, in view of congenital cystic adenoid malformation in the baby, as observed in the mother's antenatal ultrasound at 30week 5 days gestation. Her antenatal period was uneventful. The ultrasound scan report at 30 weeks 5 days gestation reported enlarged right lung showing multiple cystic lesions with polyhydraminos and ascites suggestive of congenital cystic adenomatoid malformation with mass effect on heart (with possibility of fetal hydrops). A repeat ultrasound scan at 39weeks 3 days showed multiple cystic hypoechoic lesions completely covering the right hemothorax. No other anomalies were noted. The baby was born by vaginal delivery, cried at birth and developed respiratory distress in the first hour of life. This baby was admitted to Special Newborn Care Unit of our hospital and was put on free flow oxygen. The baby was investigated to rule out other causes of respiratory distress at birth. Chest radiograph showed multiple cystic areas, largest measuring 3.5 cm, in right hemithorax, shifting the heart to left side (figure 1).

Figure 1: Xray showing cystic lesions.



Despite all measures, the respiratory distress persisted and the baby was referred to higher center for surgical intervention.

DISCUSSION

Congenital cystic lesions of the lung are rare. Congenital cystic adenomatoid malformation is a hamartomatous lesion containing tissues from different pulmonary origins. Another common malformation of the lower respiratory tract is bronchopulmonary sequestration. Both these conditions have malignant potential.[2]

Congenital cystic adenomatoid malformation occurs as a result of abnormal maturation of bronchopulmonary tree.[3]The cause of abnormal maturation is not known accurately, however hamartomatous change in terminal bronchioles could be the cause. Also, arrest during embryological development of lung between 7 weeks and 15 weeks of gestational age (pseudoglandular phase) may cause abnormal maturation of bronchopulmonary tree. Another hypothesis states that the endodermal and mesodermal tissues fail to interact leading to an imbalance with increased cell proliferation and decreased cell death. There are few genes implicated in this condition namely HOXB5, Fgf7, PDGFB.[4,5]

There are five types of congenital cystic adenomatoid malformation. Type 0 is the rarest and is lethal. Type I is the most common form, constituting 50 % to 70 %, arising from distal bronchus or proximal bronchiole. The size of cyst varies between 3 cm to 10 cm. Type II accounts for 15 % to 30 % of cases. Cyst size varies from 0.5cm to 2cm. They arise from terminal bronchioles. Type II have the highest incidence of associated anomalies, upto sixty percent. Type III account for 5 % to 10 % of cases. They arise from acinar –like tissue. The size of cysts is very small. Type IV constitute 5 % to 15 % of cases. They are alveolar in origin and cysts are as large as 10 cm. They may have malignant potential and are associated specifically with pleuropulmonary blastoma. On antenatal ultrasound scan we divide these lesions as macrocystic (>5mm) and microcystic (< 5mm). Microcystic lesions are the ones associated with poorer prognosis. In our case the baby had macrocysts. Some cysts especially type I cysts show compression of heart leading to mediastinal shift, polyhydraminos, fetal hydrops. Our case had features of mediastinal shift and polyhydraminos.

The condition may be antenatally diagnosed or the baby may present with respiratory distress or the baby may be asymptomatic with incidental finding on chest x-ray. The signs

and symptoms include tachyapnoea, poor feeding, lethargy. Majority of infants are asymptomatic. Some may develop respiratory distress, recurrent chest infections, lung abscess, bronchiectesis later on. About 20 % are associated with anomalies (pulmonary, renal or cardiac).

The differentials of this condition consists of bronchopulmonary sequesteration, bronchial cyst, neurenteric cysts, congenital lobar emphysema, congenital diaphgramatic hernia. Poor prognostic findings include hydrops fetalis, ascites, polyhydraminos, bilateral lung involvement, lung to thorax transverse area ratio of less than 0.25, cystic adenomatoid malformation volume ratio (estimated volume of malformation divided by head circumference) > 1.6. Antenatally diagnosed congenital adenomatoid cystic malformation has an excellent prognosis if no respiratory distress is present.

Those who are antenatally diagnosed must undergo genetic counselling and parents must be explained the future implications and the prognosis as per the findings. The baby should be delivered in a tertiary care with Neonatal intensive care unit facilities. Asymptomatic baby must have the diagnosis confirmed by ultrasonography and if lesions are present then must undergo a CT scan. Elective resection of the affected portion must be done between 3 to 6 months of age to prevent respiratory infections, pneumothorax, malignancy. For symptomatic babies rule out other causes of respiratory distress and once stabalised carry out resection of the affected portion of lung.

CONCLUSION

When diagnosed antenatally parents must be given a detailed information about this condition and prognosis, which may vary with type and other congenital anomalies associated with this condition. A follow up ultrasound scan must be done after the mid trimester anatomy scan as cyst size decrease after 29 weeks gestation in about 20 % cases. Also, parents must be genetically counselled as this condition may recur in the next sibling too. Once born with this condition then surgery is the definitive treatment. Definite diagnosis is made by histopathological analysis after resection.

REFERENCES

1. Nagata K, Tsukimori K, Taguchi T. Management and outcome of antenatally diagnosed congenital cystic adenomatoid malformation of the lung. Hong Kong Med J 2007; 13:31-9.
2. Argeitis J, Botsis D, Kairi-vassilatou E, Hasiakos D, Papakonstantinou K, Kondi- Pafiti A. Congenital cystic adenomatoid lung malformation: report of two cases and literature review. Clin Exp Obstet Gynecol. 2008;35(1):76-80.
3. Bianchi DW, Crombleholme TM, D'Alton ME, Malone FE. Diagnosis and management of the fetal patient. 2nd ed. New York, NY: McGraw Hill; 2010
4. Jancelewicz T, Nobuhara K, Hawgood S. Laser microdissection allows detection of abnormal gene expression in cystic adenomatoid malformation of the lung. J pediatr Surg. 2008;43(6):1044-51.
5. Volpe MV, Pham LD, Lessin M, Ralston SJ, Bhan Ina, Cutz E et al. Expression of Hoxb-5 during human lung development and in congenital lung malformations.Birth defects research part A clinical and molecular teratology. 2003;67(8):550-6.