OBJECTIVES:

1. Monitoring of the baby for any signs of respiratory distress.
2. Rule out the presence of other congenital anomalies.
3. Serial follow-up to observe for complications
4. Repeat USG and CT scan to check for regression of size of the cysts.

AIMS:

After delivery, postnatal management depends on the severity of the lesion, presentation at birth and presence or absence of complications like hydrops. Due to increased risk of recurrent infections and malignancy further in life, many authors recommend surgical resection even when infants are asymptomatic. However, another school of thought advocates that no intervention is needed, if perinatal and neonatal course is uncomplicated and serial ultrasonographic examinations confirm regression. Where conservative management is preferred; a regular follow-up is required during infancy and early childhood. By this strategy, we can avoid unnecessary surgical intervention at an young age.

INTRODUCTION

The most common malformations of the lower respiratory tract are congenital cystic adenomatoid malformation (CCAM), also known as congenital pulmonary airway malformation, and bronchopulmonary sequestration (BPS). CCAM is a hamartomatous lesion containing tissue from different pulmonary origins. BPS is made of extraneous and nonfunctioning lung tissue that has separated itself from the normal pulmonary structure. CCAM is lung tissue is replaced by cystic tissue which is non functional lung tissue. Both lesions have malignant potential.

The reported incidence of CCAM ranges from 1 in 11,000 to 1 in 35,000 live births. The reported perinatal mortality of antenatally diagnosed CCAMs has varied greatly, ranging from 9% to as high as 49%.

With the advent of newer and advanced techniques in ultrasonography and improved technical aspects of imaging, more number of cases are being detected antenatally. Exact location and size of the cysts can be made out accurately. CCAM is suspected antenatally when ultrasound findings include an increase in lung echodensity with or without cyst formation. Ultrasonography also helps to assess the regression of cysts on serial followup, making the need of more advanced imaging techniques and surgery less, thus sparing economic, financial and psychological burden on the family.

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AIMS:

To evaluate a neonate with antenatally diagnosed CCAM and follow-up during infancy, with an aim to assess the progression and intervene, if clinically needed.

CASE DETAILS:

A girl baby of birth weight 3.56kg, was born to a G.P.L.A mother; at 37 weeks of period of gestation; by emergency LSCS with breech presentation. Child cried immediately after birth and no resuscitation was needed, APGAR at 1 minute was 9. Baby was shifted to NICU for further observation. Baby had tachypnea for 4 days of life (60-70); other vitals were stable and the baby never had oxygen dependency. On a routine USG scan done at 37 weeks of gestation, a few small cysts were noted in basal region of left lung.

Antenatal USG at 37wkpOG:

Few small cysts of various sizes noted largest measuring 1.4cm possibly in left lung basal region – ? Congenital cystic adenomatoid malformation.

Chest X-ray. USG chest were planned to know the extent of lesion and its severity. The infant was evaluated by paediatric surgery team. A computed tomography chest scan with contrast revealed a lesion in left lower lobe in the posterior and medial basal segments.

CT PLAIN

ABSTRACT

Congenital Cystic Adenomatoid Malformation of the lung (CCAM) is a form of congenital cystic lung disease resulting from arrest in fetal lung development. These were once considered the exclusive domain of the surgeon, who had the authority to operate on all congenital cystic lung abnormalities regardless of size or clinical signs in order to avoid the risk of cancer and improve lung growth in even asymptomatic infants. Clinicians are reconsidering this approach in the light of the spontaneous improvement and possible resolution that occurs over months to years with many of these lesions, thinking about the opportunity to take a more conservative approach in many minimally symptomatic or asymptomatic infants in the early months of life. The risks of subsequent cancer are poorly understood and probably overstated. Many centers advocate surgery only in cases of symptomatic or significant lesions, although there is little consensus as to what constitutes a significant lesion. This article aims to evaluate a neonate, born in our hospital and was antenatally diagnosed with CCAM. This study also focuses on follow-up of the infant during infancy, with an aim to assess the progression and intervene, if clinically needed.

KEYWORDS

CCAM, BPS, antenatal USG

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Congenital Cystic Adenomatoid Malformation of the lung (CCAM) is a form of congenital cystic lung disease resulting from arrest in fetal lung development. These were once considered the exclusive domain of the surgeon, who had the authority to operate on all congenital cystic lung abnormalities regardless of size or clinical signs in order to avoid the risk of cancer and improve lung growth in even asymptomatic infants. Clinicians are reconsidering this approach in the light of the spontaneous improvement and possible resolution that occurs over months to years with many of these lesions, thinking about the opportunity to take a more conservative approach in many minimally symptomatic or asymptomatic infants in the early months of life. The risks of subsequent cancer are poorly understood and probably overstated. Many centers advocate surgery only in cases of symptomatic or significant lesions, although there is little consensus as to what constitutes a significant lesion. This article aims to evaluate a neonate, born in our hospital and was antenatally diagnosed with CCAM. This study also focuses on follow-up of the infant during infancy, with an aim to assess the progression and intervene, if clinically needed.

KEYWORDS

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CT CONTRAST:

USG abdomen, cranial USG scan and 2D Echo were normal.

On serial outpatient follow up, child remained well with no intercurrent respiratory infections and normal vitals and examination. She is conservatively managed, with a plan to repeat CT scan of the chest, closer to 1 year of age, to assess the status of cystic lesions in the lungs.

CONCLUSIONS

With the advent of advanced ultrasound imaging, more cases of congenital cystic lung conditions are being detected antenatally. Recent advances suggest that, if perinatal/neonatal course is uncomplicated and serial ultrasonographic examinations confirm regression, no further intervention is needed, and can be managed conservatively by regular follow-up, avoiding unnecessary surgical intervention at an young age.

REFERENCES: