Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/mjafi

Images in Medicine

Congenital cystic adenomatoid malformation: Antenatal ultrasonography features

Lt Col D.S. Grewal^{a,*}, Maj Arun Kant Singh^b

^aClassified Specialist (Radiodiagnosis), Military Hospital Allahabad, UP 211001, India

^bMedical Officer (Radiodiagnosis), Military Hospital Allahabad, UP 211001, India

ARTICLE INFO

Article history:

Received 23 October 2016

Accepted 13 March 2017

Available online 7 April 2017

Keywords:

Congenital cystic adenomatoid malformation (CCAM)

Antenatal

Ultrasonography (USG)

size 3 mm and 4 mm were seen in apical and basal region of echogenic mass lesion (Figs. 3 and 4). Color Doppler did not reveal any demonstrable flow in this lesion. Based on these classical findings diagnosis of CCAM was made.

Discussion

Congenital cystic adenomatoid malformation (CCAM) is an uncommon fetal lung anomaly. Pathologically various theories like overgrowth, hyperplasia, hamartomatous changes

Introduction

Congenital cystic adenomatoid malformation (CCAM) recently being referred as congenital pulmonary airway malformation (CPAM), is a rare congenital pulmonary anomaly with reported incidence of 1 in 25,000–35,000 live births.¹ On antenatal ultrasonography (USG), characteristic appearance of lesion not only helps in diagnosing it but also excludes other fetal lung anomalies. We present here a case of CCAM.

Clinical and imaging findings

A 26 year old female patient (gravida:2, para:1), reported for anomaly scan at 18th week of gestation. Anomaly scan revealed an echogenic mass lesion occupying the left hemithorax (Fig. 1) and was displacing the heart toward right (Fig. 2). Stomach was normally located (Fig. 3). Two cystic lesions of

Fig. 1 – Axial USG image through fetal thorax: Arrowhead showing echogenic mass occupying left hemithorax.

* Corresponding author.

E-mail address: drdsinghgrewal@gmail.com (D.S. Grewal).

<http://dx.doi.org/10.1016/j.mjafi.2017.03.006>

0377-1237/© 2017 Published by Elsevier B.V. on behalf of Director General, Armed Forces Medical Services.

Fig. 2 – Axial USG image through fetal thorax: showing abnormal fetal cardiac position with fetal heart being pushed into right hemithorax by echogenic mass.

Fig. 3 – Oblique coronal section showing normally located stomach. Arrowhead depicting cyst within mass lesion located in apical region.

have been postulated to explain defect at level of terminal bronchioles.²

Based on clinical and pathological spectrum, CCAM was classified into 3 groups by Stocker et al.³ These are: Type I – is most common, account for 50–70% of cases, has single or multiple large cysts of size >2 cm. Type II – account for 18–40% of cases, is composed of multiple medium sized cysts (<2 cm size), has association with multiple congenital anomalies. Type III – account for 10% of cases, composed of bulky lesion with evenly distributed tiny cysts, prognosis of this category is poor because of association with congenital anomalies and respiratory compromise. Though this classification system remains most widely accepted however based on the site of origin of CCAM, this classification was subsequently expanded into 5 types by Stocker. A new term CPAM has been used for CCAM because lesion is cystic only in three types (out of 5 types) and adenomatoid in type III.⁴ Adzick et al. proposed a simple classification which is based on gross anatomy and USG findings. These groups are: (A) macrocystic group – lesion with single or multiple cysts of size ≥ 5 mm, (B) microcystic group – predominantly solid lesion with cyst size <5 mm.⁵ Our case belongs to microcystic group.

Fig. 4 – Axial USG image showing another cyst within echogenic mass at level of fetal heart.

CCAM is usually unilateral and is more common in left lung as is seen in our case. However right sided lesions and bilateral lesions are also known. Various renal, intestinal, cardiac and skeletal anomalies are associated with CCAM. In our case CCAM was an isolated anomaly. Karyotype was not done as there is no known association of CCAM with chromosomal anomalies.⁶

On antenatal sonography bronchopulmonary sequestration (BPS) and congenital diaphragmatic hernia (CDH) are the two most important differential diagnosis of CCAM. CCAM (other than type III) can be differentiated from BPS by demonstrating cyst in lung mass. In addition color Doppler or power Doppler can depict systemic blood supply of BPS, pulmonary blood flow in CCAM and thus can be used to differentiate type III CCAM from BPS. In our case color Doppler did not reveal any blood flow in echogenic lung mass at 18 weeks of gestation. Similar findings have also been reported by Ruano et al.⁷ On the contrary coexistent CCAM and BPS, CCAM with systemic blood supply has also been reported in literature. These variations if present can cause great diagnostic challenge. Small feeding artery may further pose a challenge, as it could be difficult to detect on Doppler. Another important differential diagnosis, CDH can be differentiated from CCAM by demonstrating normal stomach bubble and absence of peristalsis in CCAM.

Overall prognosis in fetuses with CCAM is good. In our case parents were explained the prognosis, but they opted for termination of pregnancy. Development of hydrop, polyhydramnios, mediastinal shift, bilateral lung lesions and associated congenital anomalies are main cause of adverse outcome.^{8,9}

Conflicts of interest

The authors have none to declare.

REFERENCES

1. Laberge JM, Flageole H, Pugash D, et al. Outcome of prenatally diagnosed congenital cystic adenomatoid lung malformation: a Canadian experience. *Fetal Diagn Ther*. 2001;16(3):178–186.

2. Di Prima FAF, Bellia A, Inclimona G, Grasso F, Meli MT, Cassaro N. Antenatally diagnosed congenital cystic adenomatoid malformation (CCAM): research review. *J Prenat Med.* 2012;6(2):22–30.
3. Stocker JT, Madewell JE, Drake RM. Congenital cystic adenomatoid malformation of the lung. Classification and morphologic spectrum. *Hum Pathol.* 1977;8:155–171.
4. Stocker JT. Congenital pulmonary airway malformation: a new name and an expanded classification of congenital cystic adenomatoid malformations of the lung. Symposium 24: non-neoplastic lung disease. *Histopathology.* 2002; 41(2 suppl):424–430.
5. Adzick NS, Harrison MR, Glick PL, et al. Fetal cystic adenomatoid malformation: prenatal diagnosis and natural history. *J Pediatr Surg.* 1985;20:388–483.
6. Iavazzo C, Eleftheriades M, Bacanu AM, Hassiakos D, Botsis D. Congenital cystic adenomatoid malformation: is there a need for pregnancy termination? *Case Rep Med.* 2012;2012:397508.
7. Ruano R, Benachi A, Aubry MC, et al. Prenatal diagnosis of pulmonary sequestration using three-dimensional power Doppler ultrasound. *Ultrasound Obstet Gynecol.* 2005;25: 128–133.
8. Bunduki V, Ruano R, da Silva MM, et al. Prognostic factors associated with congenital cystic adenomatoid malformation of the lung. *Prenat Diagn.* 2000;20(6):459–464.
9. Usui N, Kamata S, Sawai T, et al. Outcome predictors for infants with cystic lung disease. *J Pediatr Surg.* 2004;39(4):603–606.