Case Report

Congenital pulmonary airway malformation – hybrid lesion – a case report

Chitra Andrew[1*], Shivani Gopal[2], Hemalatha Ramachandran[3], Umamaheshwari Balakrishnan[4], Usha Vishwanath[5]

{[1]Associate Professor, [2]Sr. Resident, [3]Assistant Professor, [5]Professor, Department of Obstetrics and Gynecology}
{[4]Assitant Professor, Department of Neonatology}
Sri Ramachandra Medical College, No. 1 Sri Ramachandra Nagar, Porur, Chennai – 600116, Tamil Nadu, INDIA.
Email: chitraandrew@gmail.com

Abstract

Introduction: Congenital pulmonary airway malformation (CPAM) is a fetal condition usually diagnosed in the early second trimester. This is a mass of lung tissue which has proliferation of bronchial structures. The space occupying nature of the lesion may result in mediastinal shift, or pleural effusion or hydrops due to increased intra-thoracic pressure. Post-natally, the CPAM tissue is unavailable for gas exchange leading to respiratory compromise. The condition is known to regress in utero but needs to be assessed for progression with evaluation of the CPAM volume ratio (CVR) calculated as CPAM volume divided by head circumference of fetus; to anticipate the occurrence of hydrops. Fetal surgery has been performed in a few centres for fetuses with a high CPAM ratio to alleviate hydrops and improve development of normal lung tissue for perinatal oxygenation. **Case report:** We describe the diagnosis and fetal management of a hybrid CPAM diagnosed at 20 weeks and monitored to term for an optimal outcome. An increase in CVR was noted at 30 weeks, following which antenatal corticosteroids were administered to the patient following which there was a rapid reduction in the size of the lesion. The use of corticosteroids in cases with CPAM has been reported to reduce the size of the lesion leading to avoidance of fetal compromise, and this has been the case in our patient also **Conclusion:** CPAM has a natural history of spontaneous regression in utero without any medical intervention. The use of steroids in selected cases with increasing CVR may be an additional tool to delay or avoid fetal surgery by causing regression of the lesion. Further study including randomization of selected cases is required to conclude on the use of antenatal steroids in the fetal management of CPAM.

Keywords: CCAM volume ratio, congenital pulmonary airway malformation, corticosteroids, fetal hydrops, hybrid lesion

[*]**Address for Correspondence:**
Dr. Chitra Andrew, F3 Ward, OB- GYN Department, Ultrasound unit, Sri Ramachandra Medical College and Research Institute, No. 1, Sri Ramachandra Nagar, Porur, Chennai-600116, Tamil Nadu, INDIA.
Email: chitraandrew@gmail.com
Received Date: 11/09/2014 Accepted Date: 18/09/2014

Access this article online

Quick Response Code:

Website: www.statperson.com

DOI: 20 September 2014

INTRODUCTION

Prenatal imaging has resulted in an increase in the number of prenatal diagnoses of lung lesions. These are more in number previously seen only at birth since the lesions have a natural history of regression in utero. The accurate diagnosis, counselling and fetal management of these lesions is therefore gaining more importance. Additional imaging tools such as MRI are utilized where there is ambiguity in the diagnosis.[1] Congenital pulmonary airway malformation (CPAM) is a condition where there is proliferation of the bronchial tissues which form a mass causing mass effect within the fetal lung. Post natally, the neonate has poor oxygenation in large lesions. Although the lesions may regress in utero, about 20 – 25% of cases can progress in size resulting in mediastinal shift. The increasing intrathoracic pressure result in hydrops which increases the perinatal mortality greatly.[2] Broncho-pulmonary sequestration (BPS) is a condition which closely mimics CPAM but is lung tissue with no bronchial connections. The non- functioning lung

How to site this article: Chitra Andrew, Shivani Gopal, Hemalatha Ramachandran, Umamaheshwari Balakrishnan, Usha Vishwanath. Congenital pulmonary airway malformation – hybrid lesion – a case report. *MedPulse – International Medical Journal* September 2014; 1(9): 572-575. http://www.medpulse.in (accessed 21 September 2014).

tissue may also result in mass effect similar to a CPAM. The difference between the lesions is that arterial supply to CPAM is from the pulmonary artery whereas BPS has a systemic vascular supply. The CPAM volume ratio which is the ratio of the volume of the mass divided by the head circumference is used to normalize the size of the tumour to the fetal size.[3] A CPAM volume ratio [CVR] of >1.6 has been associated development of hydrops and therefore poorer outcome and is an indication for fetal surgery in many centres. The fetuses with CVR less than 1.6 are managed conservatively with close fetal monitoring for the appearance of complications and the objective is to achieve a gestational age which is compatible with optimal survival.[2] Administration of antenatal steroids to reduce the size of lesions and delay the progression of the lesion has been documented and is now being studied more in an effort to reduce the number of cases needing fetal surgery. This is because, although the results of fetal surgery are good, the procedure requires anaesthetic and surgical expertise and is also associated with maternal morbidity and fetal mortality.[1]

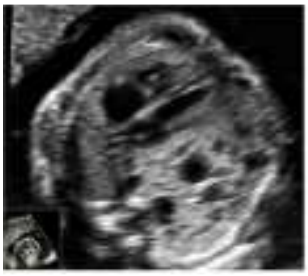

Figure 1

Figure 2

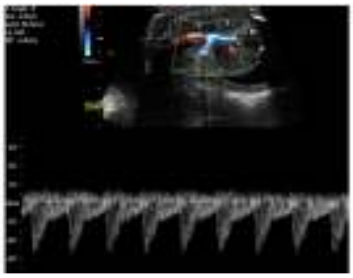

Figure 3

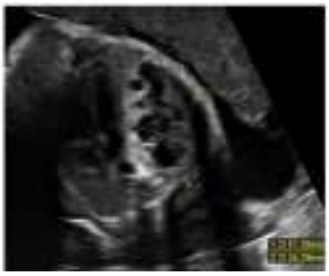

Figure 4

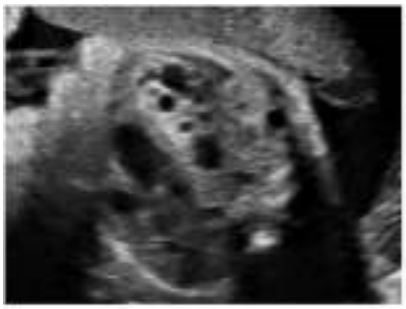

Figure 5

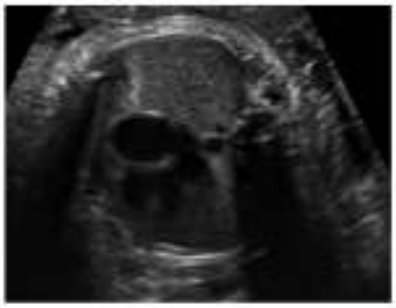

Figure 6

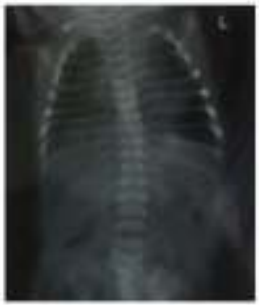

Figure 7

Figure 8

Legend

Figure 1: Lesion in left lung; **Figure 2:** Systemic blood supply to mass; **Figure 3:** Pulse Doppler arterial flow pattern; **Figure 4:** Lesion at 30 weeks; **Figure 5:** Lesion at 30 weeks showing cysts; **Figure 6:** No lesion detectable at 33 weeks; **Figure 7:** Normal x ray at day 1 of life; **Figure 8:** Healthy neonate at 4 months

CASE REPORT

Mrs. S. aged 28 years, married 2 years, 3 degree consanguineous marriage, Gravida 2with one previous abortion, was referred for a targeted scan at 20 weeks gestation. She hadpreviously undergone a spontaneous abortion at 8 weeks gestation. In the present pregnancy, the viability scan was corresponding to dates. No first trimester scan or combined biochemistry had been done. Her targeted scan at 20 weeks gestation revealed single live intrauterine gestation with normal liquor, growth and fetal activity. On detailed evaluation of the fetal anatomy, a mass was seen in the right side measuring 4.3 x 4.4 x 2.6cms. (Fig.1) The mass was echogenic and had multiple small cysts within the largest of which measured 10 x 9mm. The cardiac axis 41 degrees indicating there was no mediastinal shift. On colour and power Doppler the vascular supply to the lesion (Fig. 2, 3) was confirmed to be from the abdominal aorta suggesting that this was a CPAM [congenital pulmonary airway malformation] hybrid lesion. The CPAM volume ratio was 1.1. There was no evidence of hydrops in the fetus which was otherwise structurally normal. A detailed fetal ECHO was also done which revealed no structural cardiac malformations. The couple was counselled in detail regarding the nature of the lesion and that the outcome could be favourable as the CCAM volume ratio was less than 1.3 at the point of examination. They were advised that serial monitoring every fortnight would be carried out to look for development of hydrops and effusion. The couple was anxious and had many questions regarding

potential outcome. They were assured that CPAM could potentially have a favourable outcome with reduction of the size of the mass with advancing gestation and even apparent disappearance of the lesion. They were cautioned regarding the need for careful regular monitoring to look for complications. When reassured in this manner they opted to continue the pregnancy and agreed to the regular follow up visits. The follow up visits at 23, 25 and 27 weeks showed similar findings with a CPAM volume ratio of 1.1 and no evidence of mediastinal shift or fetal hydrops. When examined at 30 weeks gestation,(Fig. 4,5) the mass had increased in size over three weeks measuring 5.4 x 2.9 x 4.9cms although there was no evident mediastinal shift or fetal hydrops. The CPAM volume ratio at this point of examination was 1.5. As there was as sudden increase in the size of the lesion, the fetal medicine team met with the neonatology team to discuss further management of the baby. The option of antenatal steroids was discussed which was shown to have some benefit in few case reports. Since the fetus was at 30 weeks and the steroids would benefit the fetal lung maturity, a course of maternal steroids was decided upon. Betamethasone 12 mg was administered intra muscularly to the mother as two doses 24 hours apart. The fetus was examined 10 days later at 31 weeks +4 days. The CPAM volume ratio was 1.2. The fetus was otherwise normal. A repeat examination at 32 weeks + 4 days [a week later] showed a CPAM volume ratio of 0.5. Successive examinations at 33 weeks and 34 weeks showed further reduction of the size of the mass following which the lesion could not be visualized from 36 weeks onward (Fig.6) . The fetal growth, activity and liquor continued to be along normal limits. The baby girl was born at 38 weeks gestation weighing 3.25 kgwith APGAR at 1 minute of 8/10 and 5 minutes of 9/10. The neonate had no respiratory distress at birth and had an uneventful hospital stay. The postnatal chest X ray at 24 hours of life showed no lesion (Fig. 7) and as the baby continued to remain well and able to take breast feeds, she was discharged home on day 3 of life. We reviewed the baby at 4 months of life (Fig. 8) and there were no respiratory symptoms and the child was thriving well. A review with the paediatrician and CT scan has been planned prior to pediatric surgeon's opinion.

DISCUSSION

The lung lesions that were considered in the differential diagnosis of this particular case included CPAM [congenital pulmonary airway malformation] and broncho-pulmonary sequestration. CPAM has various types including micro-cystic [cysts<5mm] and macrocystic [cysts>5mm]. The microcystic type has a worse prognosis and often leads to the complications discussed earlier.[4] Broncho pulmonary sequestration is a mass of lung tissue that does not communicate with the trachea-bronchial tree. It is usually an echo-dense lesion which is seen to have its own vascular supply from the systemic circulation of the fetus. While about three quarter of antenatally diagnosed conditions regress spontaneously, the lesion can be complicated by mediastinal shift, hydrops and pleural effusion which lead to perinatal morbidity.[2] In our case the sonographic findings were of a macrocystic type with multiple cysts measuring 5 – 10 mm and a systemic vascular supply. This is consistent with a hybrid lesion which displays the echo-characteristics of a CPAM but takes the vascular supply from the aorta, similar to broncho-pulmonary sequestration.[4]. Hybrid lesions generally have a good outcome. Maternal corticosteroids antenatally administered causing reduction in CPAM volume ratio and improvement of hydrops was reviewed by Morris *et al*. In their study 15 cases of CPAM who had been given maternal steroids were reviewed. They found a 53% survival rate in high risk patients given steroids and opined that placebo controlled trials would be required to conclude on the beneficial effects of steroids in high risk CPAM.[5] Lob *et al* when comparing the outcomes of steroids versus fetal surgery in high risk microcystic CPAM noted that the steroid group had a 92% survival which was higher than the 82% of the fetal surgery group. They recommended that steroid therapy may be a preferred first line in the therapy compared to fetal surgery.[6] Derderian *et al* in a recent paper have described a series of cases with multiple doses of antenatal steroids in fetuses with suboptimal response to a single dose of steroid therapy.[7] They concluded that multiple dose regimens may reduce the need for fetal surgery. Neonatal management of the CPAM includes resuscitation and planning for surgery after stabilizing the oxygenation of the neonate.[8] If symptomatic, surgery may need to be planned in the immediate neonatal period. Asymptomatic infants can be assessed at 6 months age and decision of timing for surgery made at that time.[8]

CONCLUSION

CPAM when diagnosed in utero requires close fetal monitoring. The condition usually regresses but may progress to life threatening conditions including hydrops fetalis. Serial monitoring using CVR is the standard of care for this condition. Antenatal steroids are an option in the management of high risk cases to avoid surgery. In a country where fetal surgery is still in the early stages this would reduce the perinatal morbidity and mortality of these babies.

REFERENCES

1. Wall J., Coates A, Prenatal imaging and postnatal presentation, diagnosis and management of congenital lung malformations Current Opinion in Pediatrics: June 2014 - Volume 26 - Issue 3 - p 315-319 doi: 10.1097/MOP.0000000000000091 Pulmonology: Edited by David N. Cornfield
2. Ed. Bianchi D. W., Crombleholme T.M., D' Alton M.E., Fetology : Diagnosis and management of the fetal patient,2nd edition, pp 255 – 271, © Mc Graw Hill 2010
3. Crombleholme T.M., Coleman B., Hedrick H., Cystic adenomatoid malformation volume ratio predicts outcome in prenatally diagnosed cystic adenomatoid malformation of the lung. J Ped Surg, Volume 37, Issue 3, March 2002, Pages 331–338, doi:10.1053/jpsu.2002.30832
4. Azizkhan,R. G., Crombleholme T.G., Congenital cystic lung disease: contemporary antenatal and postnatal management Ped Surg Int., June 2008, Volume 24, Issue 6, pp 643-657 Date: 05 Apr 2008,
5. Morris L. M., Lim F. Y, Livingstone J.C., et al, High-risk fetal congenital pulmonary airway malformations have a variable response to steroids J. Ped. Surg. Volume 44, Issue 1, January 2009, Pages 60–65, doi: 10. 1016/j.jpedsurg.2008.10.012
6. Loh K.C, Jelin E., and Hirose S., et al Microcystic congenital pulmonary airway malformation with hydrops fetalis: steroids vs open fetal resection J. Ped. Surg. Volume 47, Issue 1, January 2012, Pages 36–39, doi:10.1016/j.jpedsurg.2011.10.015
7. Derderian S.C., Coleman A.M., Jeanty C et al, Favorable outcomes in high-risk congenital pulmonary airway malformations treated with multiple courses of maternal betamethasone, J. Ped. Surg. Corrected Proof, in press, doi:10.1016/j.jpedsurg.2014.08.019
8. Keswani S., Crombleholme T.M., Chapter 17.2, Fetal Tumours, Clinical Management, pp 331 – 333, Book: Fetal therapy : Scientific basis and critical appraisal of Clinical Benefits. Ed. Kilby M.D., Oepkes d., Johnson A., © Cambridge university Press 2013.

Source of Support: None Declared
Conflict of Interest: None Declared