Refresher Course

Ultrasound Assessment of a Fetus with Congenital Adenomatoid Malformation

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INTRODUCTION

Congenital adenomatoid malformation (CAM) is a rare abnormality of the lung which is characterised by excessive growth of the terminal respiratory elements.[1,2] Ultrasound examination can diagnose these lesions in utero.[3-5] The natural course of CAM varies greatly from neonatal death related to hydrops[6] and pulmonary hypoplasia, to excellent outcome, with spontaneous improvement in utero as have been reported in some cases.[7]

Classification and prognosis

CAM has been classified into three types, which were first identified by Stocker,[8] based on pathological appearance. The prognosis is different for each type.[8] Type I CAM consists of single or multiple large intercommunicating cysts, usually confined to one lobe, while type II includes smaller cysts and is often associated with anomalies such as sirenomelia, renal agenesis and extra lobar sequestration. Type III is often a bulky lesion occupying an entire lobe or lung with extremely small cysts. The reported survival rates of neonates are related to the CAM classification. Survival is best for type I cases at 90%, for type III, 60%, for type II 56%, due to the severity of the associated anomaly.

The important determinants of prognosis are the presence of associated anomalies and the amount of lung affected. A large space occupying lesion can interfere with the development of the adjacent lung and cause dyspnea due to a shift of mediastinal structures. Sometimes there is an occurrence of slow clearing of lung liquid.

The value of ultrasound assessment

Serial ultrasound examinations can be used to determine the type of lung lesion and its progression in utero. Additional benefits of ultrasound assessment in a CAM patient include monitoring fetal growth, estimating amniotic fluid volume and excluding other fetal anomalies which may accompany CAM. Prenatal ultrasonography also enables the physician to determine the need for elective surgery early in infancy before the child develops respiratory distress or a potentially life-threatening infection.[4,9] Fetal surgery undertaken in utero has also been reported. A fetus with CAM and secondary hydrops at 23 weeks gestation underwent in utero resection of the involved right middle lobe of the lung. The surgery was successful and there were no related maternal or fetal complications. However, a neonate was delivered at 30 weeks gestation due to premature rupture of the membranes. Another report[4] described a fetus with normal-appearing lung fields at the time of prenatal diagnosis, and other fetuses who had surgical resection of the affected lobe in utero with positive results and a lower rate of pneumonia infection 3-6 years after the operation. There is also a report recommending that conservative management is indicated in cases of cystic adenomatoid malformation of the lung without acute polyhydramnios or hydrops; a fetal survival rate of 100% has been reportd in these cases.[10]

In a recent report on a prospective study of 29 cases with a prenatal diagnosis of CAM, thoraco-amniotic shunt was offered only when hydrops or polyhydramnios was detected. The 17 cases without

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these complications were managed conservatively and survived[10]. The result of this prospective study has confirmed previous retrospective data which demonstrated the favourable outcome of CAM without accompanying hydrops or polyhydramnios.[3,11]

The most commonly CAM

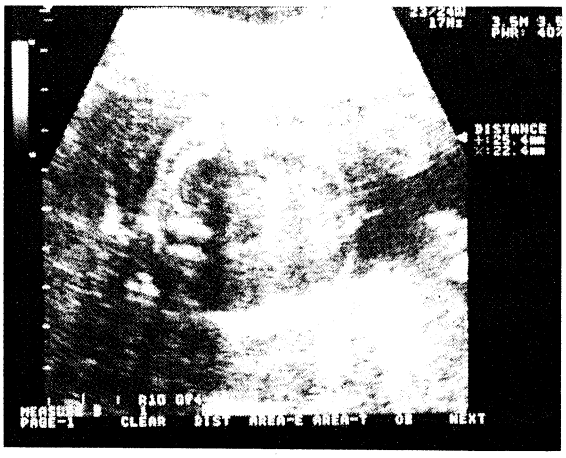
CAM (most commonly type III) can cause pulmonary hypoplasia and hydrops due to compression of the healthy lung tissue and heart respectively.[11,12] Pregnancies with CAM (25-50%) can be complicated by nonimmune hydrops fetalis and polyhydramnios. The mortality may be almost 100% if CAM is associated with hydrops.[3,11,13] Poor outcome is also associated with significant mediastinal shift.[11] Spontaneous decrease in size of the CAM or even complete regression may be seen in 20-40% of cases[5,14,15] and is a good prognostic sign.[3] A growing lesion was seen in 20% of the pregnancies and a stable lesion in 60%.[5,14] The initial CAM/chest ratio, the location of the CAM and the age of the fetus at the time of diagnosis did not correlate with the outcome. Sonographically predicted pathological type may not correlate with pathological diagnosis after surgery or with prognosis.[3]

One study reported that the outcome did not appear to be related to the anatomic type of CAM or to the presence of moderate polyhydramnios, but was related to the degree of mediastinal compression and to the existence of hydrops[15]. Poor prognostic signs for the fetus include polyhydramnios, ascites, mediastinal shift and noncystic type III CAM.[14,17]

Associated fetal anomalies

The prevalence of associated fetal anomalies in CAM including spinal and limbs deformities, various cardiac malformations, e.g., tetralogy of Fallot and truncus arteriosus and bilateral renal agenesis or dysplasia is rare. Isolated CAM is usually not associated with chromosomal abnormalities.[18] However, 2 of 25 reported fetuses with echogenic or complex lung masses had additional structural abnormalities with abnormal karyotyping.[11]

There is a spectrum of severity of CAM. The lesion can either regress and the infant be asymptomatic at birth, or it can progress to cause either fetal death from hydrops or neonatal death from associated pulmonary hypoplasia. These findings should be considered in prenatal counselling for CAM.[17]



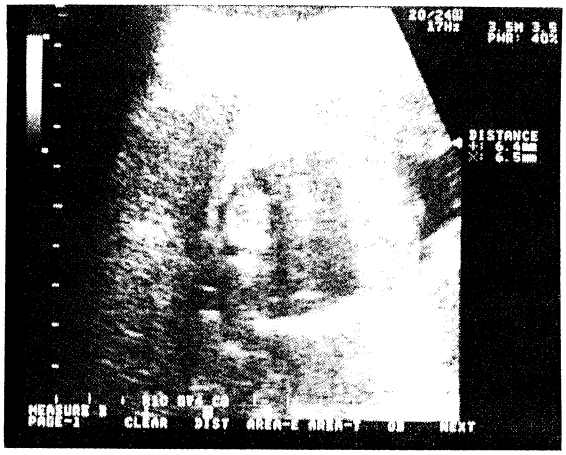


Figure 1,2. Transabdominal ultrasound scan. Transverse view of fetal chest, The macrocystic adenomatoid malformation of the lower lobe of right lung measuring 25.4×22.4 mm and 6.4×6.5 mm were seen.

CONCLUSION

Serial ultrasound examinations in the patient with CAM should be performed in order to detect early complications. In the absence of hydrops, polyhydramnios or severe mediastinal shift, a conservative management of the pregnancy is selected. There is also a report that children with "disappearing" fetal lung masses have persistent abnormalities after birth that are often subtle on radiographs but are well demonstrated with CT. Therefore, CT scanning is advised in cases of "disappearing" fetal lung masses in utero for accurate diagnosis and later management.[7]

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