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Case report

Prenatal diagnosis of cystic adenomatoid malformation of the lung in a twin pregnancy

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Most pregnant women in the United Kingdom will have at least one antenatal ultrasound examination[1] and many women will have several scans performed. This widespread use of diagnostic ultrasound during pregnancy and the continually improving quality of ultrasound equipment has resulted in the increased detection of unsuspected fetal malformations. In some cases, particularly when cysts in body cavities or minor degrees of dilatation of the urinary collecting system are seen, the exact diagnosis and prognosis may be uncertain. This often results in considerable anxiety of mothers and other family members and may lead to dilemmas about management. This report describes the management of a twin pregnancy in which a large intrathoracic cystic lesion was detected antenatally in one of the fetuses.

CASE REPORT. A 24-year-old woman, gravida three, para two, was found on routine ultrasound examination at her first antenatal visit to have a twin pregnancy of approximately 23 weeks gestation. No fetal abnormality was apparent on that occasion. At 27 weeks gestation, she complained of abdominal pain and ultrasound examination revealed polyhydramnios. Both fetuses were appropriately grown for gestational age, but a cystic lesion 4 × 4 × 3 cm was noted in the thorax of the second twin (Fig 1). The mass appeared unilocular, and was situated mainly in the right side of the chest, extending posterior and superior to the heart (Fig 2). The stomach was identified normally placed in the abdomen and the diaphragm appeared to be intact. These findings excluded a congenital diaphragmatic hernia and the most likely diagnosis was thought to be a cystic lesion of the lung. No other congenital malformation was detected.

The patient was admitted to hospital for rest. She was treated with oral ritodrine to relax the uterus and prevent preterm labour, and with weekly courses of intramuscular corticosteroid to improve fetal lung maturity. The problem was discussed with a variety of experts (perinatologist, geneticist, neonatal paediatricians and paediatric surgeon). It was decided not to perform amniocentesis or cordocentesis for chromosome studies because the pregnancy was multiple. It was thought that *in utero* aspiration of the lesion would be unwise as its exact nature was unknown. It was planned to deliver the patient by elective caesarean section in the Royal Maternity Hospital when appropriate.

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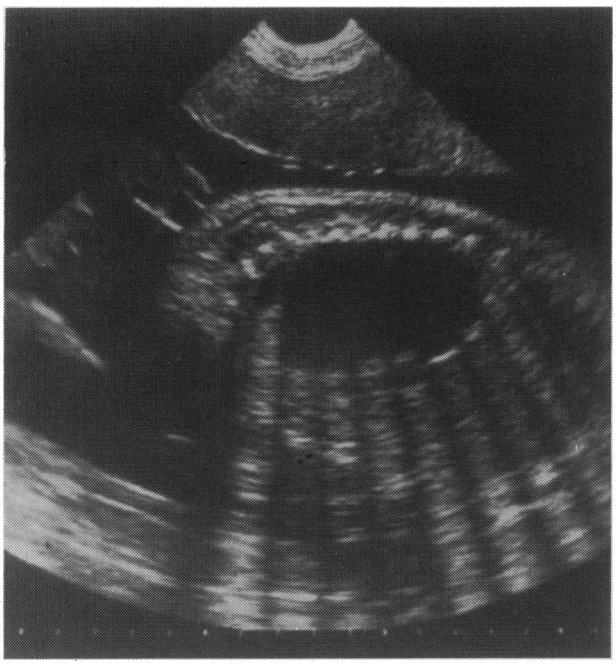

Fig 1. Longitudinal ultrasound scan of fetal thorax (Twin II) at 33 weeks gestation showing large sonolucent intrathoracic cyst.

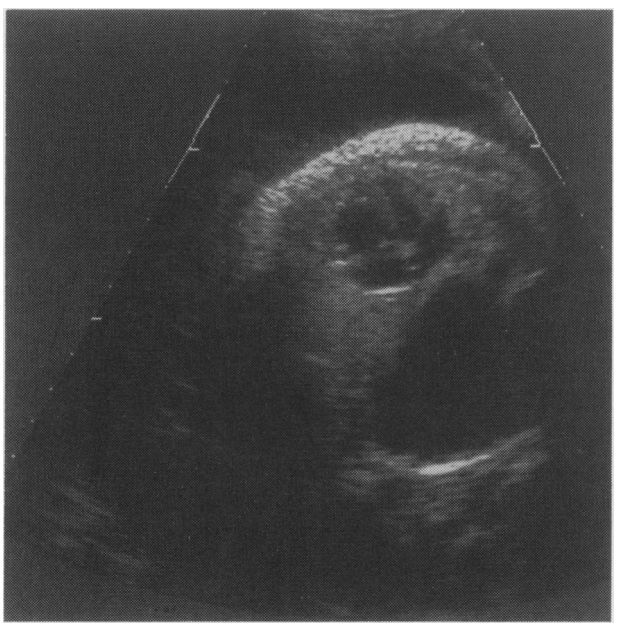

Fig 2. Transverse ultrasound scan of fetal thorax (Twin II) at 33 weeks gestation showing cystic lesion posterior to the heart.

The patient rested in hospital until 34 weeks gestation when she had recurrent episodes of uterine contractions and was transferred to the Royal Maternity Hospital. Serial ultrasound examinations showed continuing polyhydramnios, satisfactory growth of both fetuses, and gradual enlargement of the cystic lesion to 4 × 4 × 7 cm. Elective caesarean section was performed at 35 weeks gestation, this time being chosen to avoid emergency delivery as there was increasing uterine irritability and maternal anxiety, and the fetuses were now reasonably mature.

Twin one was a healthy baby girl weighing 2240 grams who had no subsequent problems. Twin two, a baby girl weighing 2395 grams, did not establish spontaneous respiration until five minutes old and remained cyanosed despite 100% oxygen given by face mask. Intermittent positive pressure ventilation was commenced. Chest X-ray confirmed the presence of a cystic lesion in the right side of the chest with mediastinal shift towards the left but was otherwise normal. The baby was transferred to the Infant Surgical Unit and underwent thoracotomy when aged three days. The middle and lower lobes of the right lung were replaced by a cystic mass and were removed. The baby has made a good recovery. The diagnosis of cystic adenomatoid malformation was confirmed histologically.

DISCUSSION

Intrathoracic extracardiac malformations are usually extrinsic, such as congenital diaphragmatic hernia which occurs in approximately 1 in 2000 births.[2] Intrinsic intrathoracic lesions are relatively rare. They include bronchogenic cysts, congenital cystic adenomatoid malformation of the lung, bronchopulmonary sequestration, primary pulmonary hypoplasia, chylothorax, idiopathic hydrothorax and mediastinal masses. Accurate prenatal diagnosis may be difficult.[3] A careful search should be made for the presence of other malformations which may be associated with some of the above conditions and consideration given to excluding possible chromosomal abnormalities.

Cystic adenomatoid malformation is a hamartomatous lesion of the lung. Stocker et al (1977) described three types,[4] on several criteria but basically on the size of the cysts. All three are characterised by a proliferation of structures resembling terminal bronchioles, increased elastic tissue and polypoid columnar or cuboidal epithelial proliferation. Type I, found in the case reported here, consists of multiple large cysts up to 7 cm in diameter and has the best survival rate. Types II and III are composed of multiple small cysts and may appear solid and hyperechoic on ultrasound. Type II lesions may be found in association with other malformations and have a poor prognosis.[4] Type III lesions are rare, often very large, and have a poor prognosis.

Detection of congenital malformations during prenatal life is important so that management may be planned to ensure the best possible outcome. Large intrathoracic masses may be associated with fetal hydrops secondary to obstruction of the venous return to the heart and pulmonary hypoplasia secondary to compression.[5] At delivery, severe respiratory distress is likely because of pulmonary compression, pulmonary hypoplasia, prematurity, or any combination of these, and the baby is likely to require major surgery within a few hours or days of birth. For all of these reasons, an experienced peadiatrician should be present at

the birth, and delivery should be in a unit with neonatal intensive care facilities, ideally close to an infant surgical unit. Discussion beforehand with everyone involved in the management of the patient allows delivery to be planned at the most suitable time. The method of delivery will depend on the individual circumstances. In the case described, caesarean section was performed because of breech presentation of the second, abnormal twin.

Space-occupying lesions in the fetal thorax, if large, are liable to cause respiratory embarrassment at delivery. Modern ultrasound equipment has made the antenatal detection of such lesions possible, allowing planning of the time, place, and mode of delivery to ensure the most favourable outcome. In the future, fetal surgery may become a realistic possibility.

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