Prenatal ultrasonographic diagnosis and successful management of mediastinal teratoma

A case report

H. R. J. DUMBELL, A. C. COLEMAN, J. M. PUDIFIN, W. S. WINSHIP

Summary

A case of mediastinal teratoma, diagnosed in utero by real-time ultrasonography during a late 3rd trimester evaluation of polyhydramnios, is described. Prompt respiratory assistance to the infant at birth and early surgical intervention led to a successful outcome.

Case report

A 25-year-old primigravida was referred for investigation of rapidly developing polyhydramnios at 35 weeks' gestation; this was confirmed by real-time ultrasonography. The thorax of the fetus was seen to contain multiple cystic areas, which displaced the heart to the right (Fig. 1). The stomach was identified in the abdomen and appeared normal (Fig. 2). There were no ascites and the kidneys were normal; there was a small amount of skin oedema. A provisional diagnosis of either cystic malformation of the lung or a cystic mediastinal mass was made.

Labour was induced at 36 weeks' gestation following spontaneous rupture of membranes. A female infant weighing 2900 g was delivered. The baby required immediate active resuscitation but despite intermittent positive pressure ventilation, air entry could not be detected clinically. Apgar scores were 2, 3 and 5 at 1, 5 and 10 minutes, respectively. The infant was put onto a Bourne's ventilator in the neonatal unit. On examination she was cyanosed and oedematous with a distended abdomen and 5 cm hepatomegaly. There was bradycardia and the apex beat was displaced to the right. Chest radiography showed opacity of both lung fields (Fig. 3). Chest drains were inserted, and 30 ml of straw-coloured fluid was aspirated from the right pleural space after which some aerated lung was seen (Fig. 4).
An umbilical arterial line was inserted to measure the Astrup rating: pH 6.85; partial pressure of arterial carbon dioxide (Paco₂) 12.15 kPa; bicarbonate (HCO₃⁻) 16.1 mmol/l; base deficit (BD) 18.7 mmol/l; partial pressure of arterial oxygen (PaO₂) 7.3 kPa; and oxygen saturation 58.6% on 100% inspired oxygen administration.

The initial haemoglobin value was 16.0 g/dl and the white cell count 32.9 × 10⁹/l. Serum electrolyte and urea values were normal.

Management was aimed at improvement of the infant's respiratory and metabolic states, and the prevention of hypoxic encephalopathy. The blood gas status improved gradually and at 14 hours the Astrup rating on 85% inspired oxygen was: pH 7.29; Paco₂ 6.67 kPa; HCO₃⁻ 24.1 mmol/l; BD 2.5 mmol/l; PaO₂ 11.7 kPa; and oxygen saturation 95%.

The infant's condition remained stable and at 36 hours computed tomography (CT) was performed; this confirmed the presence of a cystic mass occupying most of the left hemithorax displacing the heart to the right (Fig. 5). A small fragment of calcium was seen within the mass suggesting the diagnosis of a teratoma (Fig. 6).
The baby's postoperative course was stormy and complicated by a Klebsiella pneumonia and left phrenic nerve paralysis. Examination under anaesthesia at the age of 5 weeks revealed paralysis of the left vocal cord and collapse of the left bronchial system. A tracheostomy was performed and artificial ventilation was continued.

Weaning from the ventilator was only possible after 3 months and the infant remained tachypnoic and oxygen-dependent via the tracheostomy. When the infant was 3½ months old it was apparent that the left diaphragm was moving paradoxically and was paralysed; it was plicated.

After this procedure there was a dramatic improvement in the infant's respiratory function and she was fit to be discharged home 3 weeks later.

The child has been followed up regularly, is now aged 18 months and has no respiratory difficulty. She has had normal phonation since the age of 13 months and has shown steady progress in growth and development. She now weighs 9.2 kg, measures 76 cm and her developmental progress is appropriate for her age.

Discussion

Polyhydramnios is commonly associated with fetal abnormalities. These fall into two main groups: (i) cardiovascular decompensation caused by severe anaemia or cardiac anomalies and arrhythmias; and (ii) obstructive malformations of the gastro-intestinal tract.

Ultrasonography has made it possible to investigate cases of polyhydramnios to determine a possible fetal cause so that an appropriate plan may be made for the delivery and management of the fetus.2

The discovery of a cystic mass occupying the left hemithorax of a fetus requires the identification of the stomach to exclude a left diaphragmatic hernia. Thereafter the differential diagnosis is between cystic conditions of the lung and teratoma. Cystic adenomatoid malformations of the lung (CAML) account for 25% of congenital lung malformations3 and of the three types described, type I has some large cysts while type II has multiple evenly sized cysts; type III is solid. Teratoma may be distinguished from CAML if calcium is detected on ultrasonography, but it can be difficult to pick up small flecks of calcium in the presence of polyhydramnios at 36 weeks' gestation, particularly if there is excessive fetal movement, as in our case. Calcium was, however, detected on CT post-natally.

Teratoma as a cause of polyhydramnios is rare and only 10% of cases of intrathoracic teratoma present with symptoms in the neonatal period.4 Those which cause symptoms and polyhydramnios are large and fill the left hemithorax. The mechanism by which polyhydramnios is produced is probably oesophageal compression causing decreased swallowing of amniotic fluid.5 Another complication of a large intrathoracic mass in the fetus may be obstruction of the venous return to the heart resulting in pleural effusion, ascites and even hydrops.6 The presence of a large pleural effusion can critically embarrass attempts to ventilate the compressed right lung and immediate drainage of the effusion is essential.

After the surgical removal of a large intrathoracic mass, hypoplasia of the compressed lung is always a threat to a successful outcome. This, however, should not be accepted as the only cause of failure to establish adequate ventilation. Paralysis of the ipsilateral diaphragm is not an uncommon complication and can be a critical factor. Plication of the paralysed diaphragm obviates paradoxical movement enabling the affected lung to expand and function effectively.

This case demonstrates that, alerted by the prenatal diagnosis of an intrathoracic mass by ultrasonography, intensive management and early surgery for a very large benign teratoma can have a successful outcome.

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REFERENCES