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CASE REPORT

Intrathoracic extra-mediastinal cystic hygroma in infancy

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An infant with intrathoracic extra-mediastinal cystic hygroma is described. Fluid collection within the fetal chest was noted on routine antenatal ultrasound scan and this was subsequently drained. Postnatally, echocardiogram and thoracic CT scan demonstrated a cystic space between the pericardium and right mediastinal pleura. Thoracotomy performed at six weeks of age showed a multiloculated cystic mass adherent to the right pericardium and to the medial aspect of the diaphragm. Histology revealed the tumour to be a cystic hygroma (lymphangioma). Intrathoracic cystic hygroma occurring outside the mediastinum is extremely rare and has never been diagnosed previously in infancy. □ *Cystic hygroma (lymphangioma), extra-mediastinal, infant*

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Cystic hygroma (lymphangioma) is an uncommon tumour in childhood and is usually confined to the head and neck or axilla. The present report documents the first case of intrathoracic extra-mediastinal cystic hygroma presenting in a neonate.

Case report

A male infant was born by spontaneous vertex delivery at 38 weeks' gestation, weighing 3.79 kg, with Apgar scores of 9 and 10 and 5 min, respectively. The mother was a 29-year-old primigravida and during this pregnancy routine antenatal ultrasound examination at 20 weeks' gestation revealed fluid collection within the fetal chest. Mild ascites and nuchal oedema were also noted at this stage, although both had resolved spontaneously by 22 weeks' gestation. Fetal echocardiogram, karyotype and maternal viral infection screen were normal. By 28 weeks' gestation, it was felt that the fluid collection in the chest was pericardial rather than pleural, and immediately before delivery the fluid was partially drained under ultrasound control.

Initial chest X-ray showed irregularity of the right border of the upper mediastinum, with an echocardiogram demonstrating a large anterior soft tissue mass. Initial cerebral ultrasound was normal and the infant was admitted to the postnatal ward, but at 48 h of age he had a right-sided convulsion and was admitted to the neonatal unit. He continued to have clonic convulsions for which he required iv phenobarbitone. CT scan of the head showed blood within both lateral ventricles, the falx cerebri and the fourth ventricle. Blood pressure was 58/30 mmHg and clotting studies and platelet count were normal. Serum alpha-fetoprotein (AFP) concentration was increased at 36 800 μg/l, but serum human chorionic gonadotrophin (hCG) (< 4 IU/l (normal range < 5 IU/l)), urine vanillylmandelic acid (VMA) (4.5 μmol/mmol creatinine (< 21.1 μmol/mmol creatinine)) and urine HVA (8.8 μmol/mmol creatinine (< 21.7 μmol/mmol creatinine)) were all normal.

Repeat echocardiogram on day 16 revealed the cystic space to be between the pericardium and the right mediastinal pleura, and thoracic CT scan showed an anterior cystic space to the right of the heart which had soft tissue elements within it posteriorly (Fig. 1). The infant was readmitted for thoracotomy at six weeks of age, when a multiloculated cystic mass adherent to the right pericardium and to the medial diaphragm was found. The tumour was almost avascular with no solid element and was removed by a combination of blunt and sharp dissection. Histological examination of the tumour showed a cyst with a fibrous wall containing nodules of immature fat and lymphoid nodules. The appearances were those of a cystic hygroma (lymphangioma). There was no evidence of neoplasia.

The baby made a good postoperative recovery and serum AFP decreased to 49.2 μg/l. When last reviewed at 24 weeks of age, the baby was developing normally and cerebral ultrasound scan showed only mild asymmetry of the lateral ventricles.

Discussion

Cystic hygroma (lymphangioma) is a rare tumour in

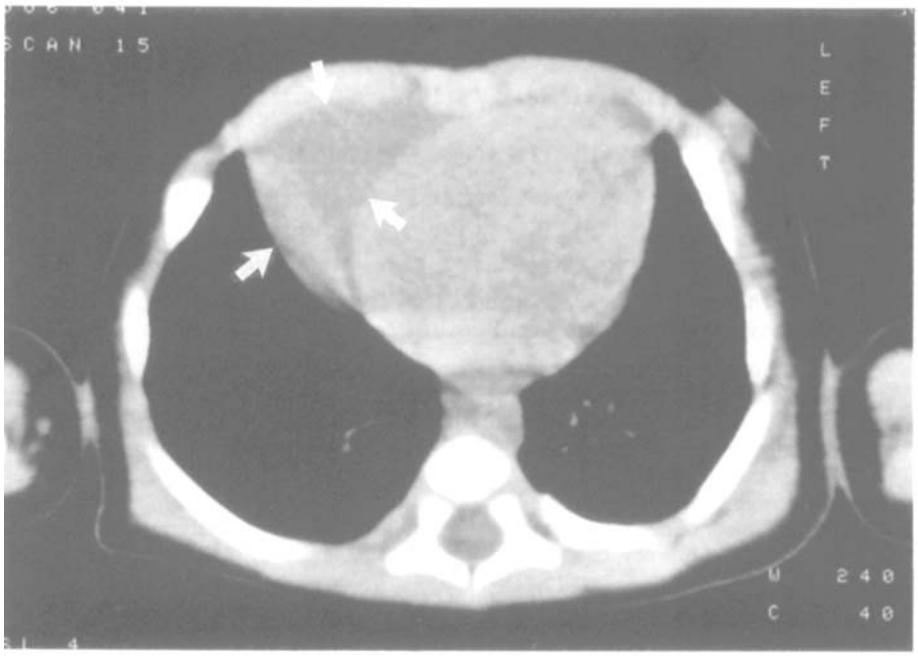

Fig. 1. CT scan of the chest, demonstrating the presence of an intrathoracic tumour.

childhood, most commonly found in the head and neck (78–90%) and axilla (20%) (1). These tumours usually present with a fluctuant cystic mass detected at birth, although the advent of routine ultrasound is making antenatal diagnosis of these tumours more common (2). Intrathoracic lesions occur in less than 10% and are usually cervico-mediastinal. Fewer than 1% of cystic hygromas are totally intrathoracic and these occur almost exclusively within the mediastinum. These mediastinal tumours may present in childhood, with large airway obstruction, pulmonary hypoplasia or, more commonly, they are found incidentally on routine chest radiographic examination (1).

Although there have been several case reports of intrathoracic extra-mediastinal cystic hygromas occurring in adults, only one case has been described previously in childhood (3). In that report, a six-year-old child presenting with pneumonia was found to have a lobulated cystic tumour between the diaphragm and under-surface of the left lower lobe. The tumour was removed at thoracotomy and found to be a cystic hygroma.

Cystic hygroma is thought to represent a hamartomatous malformation of the lymphatic system. It is postulated to develop as a consequence of a "jugular lymphatic obstruction syndrome"; ascites and pleural effusion may also develop although, as with this patient, spontaneous resolution can occur. In this patient, the serum alpha-fetoprotein (AFP) concentration was raised and decreased after resection of the cystic hygroma. AFP is elevated in both amniotic and hygroma fluid in cases of cystic hygroma (both due to Turner's syndrome and other causes). The raised AFP concentration was speculated to be due to leakage of fetal serum components into the cystic hygroma fluid and into amniotic fluid (4). The presentation of intraventricular haemorrhage and fits in a full-term baby was unexplained as there was no evidence of hypertension, and urinary VMA was not increased. The differential diagnosis of the intrathoracic lesion in this child included a pericardial cyst and teratomatous dermoid cyst. These were excluded histologically.

The present report describes a unique case of an intrathoracic extra-mediastinal cystic hygroma presenting in infancy. This was diagnosed antenatally together with ascites and nuchal oedema, both of which resolved spontaneously. As in this age group, the natural history of these tumours is unknown; it may be that a more conservative approach will be justified in future cases.

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