Neonatal surgical problems of the chest

Kokila Lakhoo

Abstract

Neonatal surgical problems of the chest include a wide spectrum of anomalies that extend from chest wall deformities to developmental cardiac malformations. To remain within the scope of this publication, cardiac anomalies and chest wall deformities are excluded.

Most neonatal chest conditions have other associated anomalies such as chromosomal anomalies that may form part of more complex syndromes. In the era before prenatal diagnosis many of these conditions would present as fetal loss or asymptomatically and discovered incidentally during postnatal imaging. Prenatal diagnosis has revolutionized the diagnosis and treatment of chest conditions.

Congenital cystic lung lesions have a prenatal diagnostic accuracy of almost 100% and are mainly asymptomatic at postnatal presentation. The diagnosis may be confirmed on postnatal computerized tomography (CT) scan, however the treatment of these asymptomatic lesions remains controversial. Most institutions offer surgery to avoid future complications however surveillance with or without imaging an alternate management.

More than 60% of cases with prenatally diagnosed congenital diaphragmatic hernia do not make it beyond the neonatal period. Complex associated anomalies and lung hypoplasia are the prime causative factors for this grim outcome. Even those infants that survive post-surgical repair some will have respiratory, gastrointestinal and neurological sequeli.

Chylothorax and thoracic teratoma are other thoracic conditions discussed.

Diagnosis of oesophageal atresia is less accurate on prenatal scan. Surgery could be challenging and outcomes are dependent on associated anomalies.

Keywords congenital lung cysts; diaphragmatic hernia; neonatal chest/thoracic conditions; oesophageal atresia

Introduction

One third of infant deaths are due to congenital anomalies and in the developing countries these anomalies are one of the leading causes of death in the newborn period. Congenital malformations of the chest account for the highest cause of infant mortality and comprise abroad spectrum of abnormalities ranging from chest wall deformities to cardiac anomalies. For the purpose of this review on neonatal surgical problems of the chest, common conditions affecting the diaphragm, oesophagus and lower respiratory tract are discussed. Cardiac abnormalities and chest wall deformities are beyond the scope of this review and have been excluded.

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Cystic lung lesions

Congenital cystic lung lesions are the commonest lung parenchymal lesions diagnosed on prenatal scan. This anomaly is rare when compared to gastrointestinal or cardiac malformations and has an incidence of 1 in 10,000 to 1 in 35,000 reported in the literature. Congenital cystic adenomatoid malformations (CCAMs), bronchopulmonary sequestrations (BPS) or 'hybrid' lesions containing features of both are common cystic lung lesions noted on prenatal scan. Less common lung anomalies include bronchogenic cysts, congenital lobar emphysema and bronchial atresia. Traditionally, cystic lung lesions were classified according to the size of the cyst but later modified to macro and microcystic lesions based on prenatal diagnosis. However due to mixture and overlap of the various cystic lung lesions, the pathology is presently seen as a spectrum and reported in descriptive terms.

Prenatal detection rate of lung cysts at the routine 18–20 week scan is almost 100% and may be the commonest mode of actual presentation. Most of these lesions are easily distinguished from congenital diaphragmatic hernia however sonographic features of CCAM or BPS are not sufficiently accurate and correlate poorly with histology. Prenatal magnetic resonance imaging (MRI) though not routinely used, may provide better definition for this condition; however inaccuracies were reported in 11% of cases.

In utero diagnosis of bilateral disease and hydrops fetalis are indicators of poor outcome whereas mediastinal shift, polyhydramnios and early detection are not poor prognostic signs. In the absence of termination the natural fetal demise of antenatally diagnosed cystic lung disease is 28%. Fetal cystic lung lesions may decrease in size relative to fetal growth, but complete postnatal resolution is rare. The apparent spontaneous 'disappearance' of antenatally-diagnosed lesions should be interpreted with care, as most of these cases subsequently require surgery.

In only 10% of cases the need for fetal intervention arises. The spectrums of intervention include simple centesis of amniotic fluid, thoracoamniotic shunt placement, percutaneous laser ablation and open fetal surgical resection. Maternal steroid administration has also been reported to have a beneficial effect on microcystic CCAMs although the mechanism is unclear. A large cystic mass and hydrops in isolated cystic lung lesions are the only real indication for fetal intervention.

Normal vaginal delivery in recommended unless maternal condition indicates otherwise. Large lesion is predicted to become symptomatic shortly after birth (10%), thus delivery at a specialized centre would be appropriate. Smaller lesions are less likely to be symptomatic at birth and could be delivered at the referring institution with follow up care in a paediatric surgery centre.

Postnatal management is dictated by clinical status at birth. Symptomatic lesions require urgent radiological imaging, ideally CT scan followed by surgical excision (Figure 1). In asymptomatic cases a CT scan of the chest at 6–8 weeks after birth is recommended, even if regression or resolution is noted on prenatal scanning. True resolution of these lesions is exceptional.

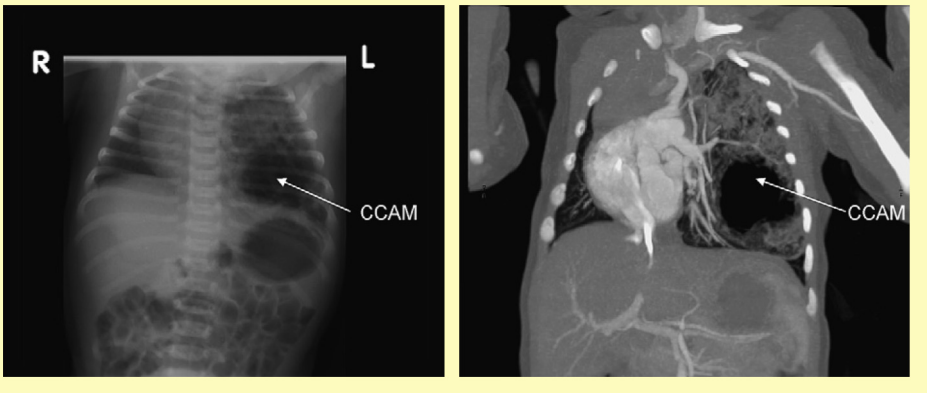

Figure 1 Chest radiograph and CT Scan showing right lower lobe CCAM.

Plain radiography should not be relied upon since over half of these lesions would be missed or underestimated.

Surgical excision of postnatal asymptomatic lesions remains controversial. Some centres have opted for conservative management by means of an annual follow up with a CT scan. The approach to treating this asymptomatic group has evolved in most centres, whereby a CT scan is performed 6–8 weeks post birth, followed by surgery before 6 months of age due to the inherent risk of infection and a small risk of malignant transformation. Small lesions, less than 1 cm may not be CCAM thus expectant management is acceptable. Successful outcome of greater than 95% have been reported for these surgically managed asymptomatic lung lesions.

Congenital lobar emphysema/overexpansion (CLE/O) is defined as an over inflation of one or more pulmonary lobes, secondary to external bronchial compression or defect in the bronchial cartilage.

Antenatal diagnostic accuracy is almost 100%. The typical postnatal presentation is of respiratory distress which may necessitate excision of the affected lobe. However many patients are asymptomatic and can be managed expectantly (Figure 2).

Bronchogenic cysts are found in the mediastinum in up to 2/3rd of cases, lying adjacent to the major airways, heart or oesophagus, with the remainder found within the lung parenchyma. Presentation is usually with pulmonary infection, but a proportion of cases are diagnosed incidentally. Surgical excision is curative and can be achieved by simple cyst removal and rarely segmentectomy or lobectomy if intraparenchymal.

Congenital diaphragmatic hernia (CDH)

CDH accounts for 1 in 3000 live birth and challenges the neonatologist and paediatric surgeons in the management of this high-risk condition. Mortality remains high (more than 60%) when the 'hidden' mortality of in utero death and termination of pregnancy are taken into account. Lung hypoplasia and pulmonary hypertension account for most deaths in isolated CDH. Associated anomalies (30%–40%) signify a grave prognosis with a survival rate of less than 10%.

In the UK most CDH are diagnosed at the 20-week anomaly scan with a detection rate approaching 70%, although detection as early as 11 weeks gestation has been reported. In the fetus,

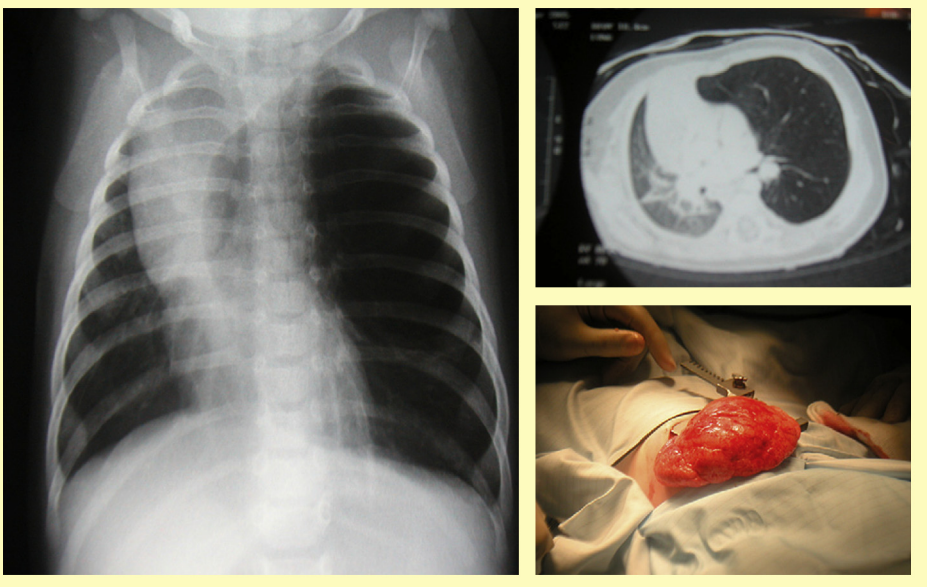

Figure 2 Congenital lobar overexpansion of the left upper lobe.