A routine 20-week antenatal ultrasound scan showed a congenital lesion of the left fetal lung, measuring approximately 25 mm x 25 mm x 30 mm. The mass showed no sonographic change through the remainder of an uneventful pregnancy. The baby was delivered by elective caesarean section at 38 weeks’ gestation, with a birth weight of 2 900 g, and had no postnatal complications. A chest radiograph performed in the early neonatal period was normal, but a contrasted single-slice helical computed tomography (CT) chest scan at age 6 weeks demonstrated the small, oval, solid mass in the left lower lobe, with no associated mediastinal shift (Fig. 1). The vascular supply of the lesion could not be identified on this scan.

Over the ensuing 9 months the baby has been well, without respiratory symptoms.

A follow-up contrasted CT scan was performed at age 7 months on a 64-slice multi-detector scanner. This showed the mass to be morphologically unchanged. However, the multiplanar reconstructions possible on the multislice scanner facilitated a definitive diagnosis. The mass was shown to have a systemic arterial supply, via a 3 mm diameter vessel arising from the descending thoracic aorta, with venous drainage through 3 radicals into the hemi-azygous vein (Figs 2 - 3). These findings are consistent with extralobar pulmonary sequestration.

Discussion

Radiological advances

This case report indicates how radiological advances have enhanced both antenatal identification of congenital pulmonary abnormalities and their postnatal diagnosis. Previously such asymptomatic pulmonary lesions may have remained
Fig. 3. Reconstructed parasagittal image, demonstrating the venous drainage of the lesion. Note confluence of three draining venules (white arrow), into the hemi-azygous vein (black arrow), lying dorsal to the descending thoracic aorta (A).  

undetected until the onset of complications in later life. Furthermore a specific diagnosis would have required invasive procedures in the form of angiography or biopsy.

Radiation risk from paediatric CT has received particular attention in the last 5 years, prompting strategies for dose reduction that will largely preserve image quality. As with any radiological procedure, chest CT should be used for appropriate clinical indications, bearing in mind that the radiation dose is equivalent to approximately 250 standard antero-posterior chest radiographs.

Pathology and management issues

The spectrum of congenital lung lesions includes sequestration, congenital cystic adenomatoid malformation (CCAM), lobar emphysema and bronchogenic cysts. Classically, sequestration describes a bronchopulmonary mass with either no connection, or an abnormal connection to the bronchial tree. It typically has an anomalous systemic arterial supply, and may be intralobar (within the pleura of the involved lung) or extralobar. An intralobar sequestration usually has normal pulmonary venous drainage, while the extralobar lesion characteristically has aberrant drainage into the right atrium, vena cava orazygous systems, as in this patient.44

Extralobar sequestrations are less common than their intralobar counterparts, have a 4:1 male to female preponderance and occur on the left side in 90% of cases. Up to 65% are associated with other congenital anomalies. Since congenital pulmonary lesions may have overlapping features, the broader term ‘congenital thoracic malformation’ is recommended in preference to applying a pathological diagnosis in a clinical setting.2

Many such malformations detected at the 20-week antenatal scan regress or disappear by term. Although sequestrations are rarely symptomatic at birth, symptoms can be severe and are determined by the size and location of the lesion. Initially asymptomatic lesions may become symptomatic owing to the development of complications, in which case surgical removal is indicated.4

Therapy for asymptomatic lesions is controversial and the optimal management strategy is not clearly defined. Options include conservative management with careful long-term follow-up, surgical removal, or embolisation of the systemic feeder vessel. Reasons for surgical intervention or embolisation include the risk of recurrent infections, an increase in the arteriovenous shunt, pressure effect on adjacent normal lung, airway compression and malignant degeneration.6,10 The latter has been reported in congenital malformations, particularly CCAMs.9,10

Some experts advocate resection of all lesions irrespective of symptoms because of the potential for complications, while others have promoted conservative management owing to the lack of data on the long-term natural history of asymptomatic lesions and the possibility of their spontaneous regression.4 The optimal timing of surgery is also unclear.11

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