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Imaging of congenital lung malformations

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ABSTRACT

Congenital lung malformations are a heterogeneous group of anomalies that involve the lungs and tracheobronchial tree (congenital airway pulmonary malformation, bronchial atresia, bronchogenic cyst, congenital lobar overinflation, pulmonary cyst, hamartoma, pulmonary isomerism and azygous lobe), vascular abnormalities (arteriovenous malformations, anomalous pulmonary venous return, pulmonary artery sling, interrupted pulmonary artery, pulmonary varix, pulmonary vein stenosis and pulmonary lymphangiectasia), or frequently both entities (pulmonary sequestration, pulmonary maldevelopment and scimitar syndrome). Advances in diagnostic imaging (including sonography, multi-detector computer tomography, magnetic resonance imaging and angiography) have increased their detection during both antenatal and postnatal periods, and radiological characterisation, which in turn influence patient counselling and management stratification. An educational illustration of the clinical application in characterisation of these malformations is presented.

Introduction

Congenital lung malformations comprise a spectrum of anomalies involving the lung parenchyma and its bronchovascular structures. They account for 5–18% of all congenital anomalies, with a cumulative incidence of 30–42 cases per 100,000 population, although this likely underestimates the true prevalence due to the frequency of undetected/asymptomatic lesions. Advances in diagnostic imaging have contributed to increased detection of lesions, both antenatally and in later life as clinically occult lesions identified incidentally in patients examined for other clinical indications. Given the wide variability yet often comparable clinical presentation, accurate diagnosis is critical for counselling, with implications on pregnancy viability and surgical planning. We therefore review the utility of radiological modalities available in the diagnostic evaluation antenatally and postnatally, present an educational outline of the radiographic characterisation of the most prevalent congenital lung malformations, and briefly discuss the impact of diagnostic imaging on clinical management strategies.

Diagnostic imaging modalities

Ultrasound

Ultrasound usually forms the first imaging in the detection of congenital lung malformations that are identified during antenatal screening. Its benefits of accessibility, reproducibility and radiation safety further enables surveillance in the antenatal period. Lesions most commonly manifest as echogenic masses within the chest, and infrequently in the abdomen with extralobar pulmonary sequestrations (PSs). Other features include the presence of cysts, effusions/hydrops and with large unilateral lesions, mass-effect causing contralateral mediastinal shift. Lesions are assessed in orthogonal planes and where feasible characterised using a high-frequency linear transducer (12–15 MHz in postnatal children) to achieve best resolution. The utility of B-mode Doppler can further characterise associated vascular anatomy such as the presence of anomalous vessels in PSs.

Plain radiographs

This usually forms the first imaging in the postnatal period, with the lesion manifesting as a dark hyperlucent or white radio-opaque mass within the chest. Large lesions may present with changes affecting an entire hemithorax [e.g., congenital pulmonary airway malformation (CPAM)], overinflation, mediastinal shift and complications such as pneumothorax, whilst vigilance may yield more subtle features including vascular abnormalities

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(e.g., scimitar syndrome), airway abnormalities including tracheal shift or carinal splaying with bronchogenic cysts, and associated anomalies (e.g., cardiac and spine).

Computerised tomography

Contrast-enhanced volumetric multi-detector computerised tomography (CT) is established as the gold-standard imaging modality in the radiographic characterisation of congenital lung malformations, conferring advantages of high spatial resolution and short acquisition times, thereby precluding requirement for general anaesthesia that may be necessary for other modalities such as magnetic resonance imaging (MRI). However, its clinical application is less uniform. Lesion characterisation with CT may influence antenatal counselling as well as surgical planning postnatally, whilst the risk–benefit balance including radiation considerations in asymptomatic children renders advocates of conservative management of cystic lung malformations, preferring to use it sparingly for patients who become symptomatic or where selective surgical intervention is merited.

Given considerations of radiation safety, particularly in the radiosensitive paediatric population and risks of iodine contrast, it is imperative that the examination is both justified and optimised. The recommended field of examination extends from the lung apices to the abdominal aorta down to the second lumbar vertebral level, to ensure assessment of the lung lesion and also potential anomalous vessels that may arise as inferiorly as from the abdominal aorta in extralobar PSs. Intravenous contrast (e.g., *Iopamidol*, Bracco Diagnostic Inc.) may be administered, usually as a single bolus, by hand or via automated pump injection, based on 2 mL/kg body weight volume, and scanning commenced to coincide with arterial system opacification (approximately 20 seconds post-injection) and delineation of anomalous system arterial vessels. A modification of this technique using a 50/50 split bolus of the contrast volume separated by a small-volume saline "chaser" may alternatively be employed, particularly if there are diagnostic concerns that necessitate optimal opacification of both arterial and pulmonary venous systems (e.g., anomalous venous return). Scanning parameters such as peak kilovoltage, milliamperage and axial-slice thickness are also optimised using paediatric age-adjusted algorithms.[1] Finally post-scanning reconstruction is employed to provide high-resolution multi-planar reformatted (MPR) imaging and 3-dimensional reconstructions of lesions and associated anomalous vascular anatomy.

Magnetic resonance imaging

The application of magnetic resonance imaging (MRI) has evolved, particularly in perinatal imaging, where it offers benefits of high contrast resolution and obviating ionising radiation to both mother and child, and enables quantification of congenital lesion size and volume of uninvolved lung that influence foetal prognostication, counselling and management. However, its longer scanning time necessitating general anaesthesia in most young children over 6 months of age (when a "feed and wrap" may be precluded), and its poor spatial resolution, particularly for assessment of lung parenchymal detail, ultimately confer significant disadvantages over CT in characterising most congenital lung malformations.

A cardiac coil with gating is employed to minimise cardiac motion artefact degradation of images. Imaging is performed in the axial and coronal plane (along the axis of the foetus in antenatal imaging), predominantly using T2-weighted fast spin echo, which outlines lesions as T2 hyperintensities due to the fluid within the lungs and airways antenatally, and hypointense postnatally once the fluid clears. In addition, T1-weighted sequences may be obtained with fat-saturation in the axial and coronal planes pre- and post-gadolinium contrast, the latter used sparingly to avoid potential nephrotoxicity particularly in neonates with immature kidneys. This will characterise vascular lesions, whilst 3D angiography will delineate anomalous vascular anatomy.

Catheter angiography

Catheter angiography is the gold standard in determining arterial anatomy, and whilst rarely necessary in diagnostic assessment of congenital lung malformations, may be employed in therapeutic embolization of PSs.

Congenital lung malformations and its radiographic characterisation

Congenital lung malformations may be broadly grouped as abnormalities predominantly relating to the lung parenchyma and tracheobronchial tree, those having both lung/airway and vascular entities to the abnormality, and those that are specifically vascular abnormalities. We will focus on the former two groups that involve the lung and airways, with review of pulmonary vascular abnormalities (e.g., arteriovenous malformations, anomalous pulmonary venous return, pulmonary artery sling, interrupted pulmonary artery, pulmonary varix, pulmonary vein stenosis, and pulmonary lymphangiectasia) beyond the scope of this article.

Parenchymal and airway abnormalities

Congenital pulmonary airway malformations

Congenital pulmonary airway malformation (CPAM) is the most common (30–40%) of congenital lung malformations and represents a hamartomatous proliferation of cystic spaces that resemble terminal bronchioles, being lined by respiratory epithelium, but lacking cartilage or bronchial glands.[2,3] The previous nomenclature (congenital cystic adenomatoid malformation; CCAM) is superseded based on recognition that lesions may contain solid as well as cystic components. They communicate with the bronchial tree (distinguishing it from bronchogenic cysts and PSs) and have blood supply derived from the pulmonary artery (distinguishing it from PSs) as well as pulmonary venous drainage.

The current Stocker classification of CPAMs,[4] modified on the original classification,[3] comprises five types based on the size of cysts and its resemblance to the bronchoalveolar tree. Type 0 involves multilobar acinar dysgenesis/dysplasia of major tracheobronchial airways, and being bilateral, is incompatible with life. Type 1 (70%) comprises macrocysts of bronchial or bronchiolar origin, with the largest cyst exceeding 2 cm. Type 2 (15–20%) is similar to type 1 but with cysts 0.5–2 cm in size. Type 3 (10%) is a predominantly solid lesion associated with microcysts less than 0.5 cm, of bronchoalveolar duct origin, and is the only type that is adenomatoid. Type 4 ($<5\%$) consists of large air-filled or fluid-filled cysts of distal acinar origin typically affecting a single lobe of the lung.

CPAMs are often identified on antenatal ultrasound as an echogenic mass (particularly type 3 lesions) with variable sized cysts, dependent on its subtype. Further evaluation with foetal MRI typically identifies T2-hyperintense lesions and may delineate the architecture of individual cysts within the mass. Both ultrasound and MRI may also demonstrate mass effects (mediastinal shift, hydrothorax) and vascular supply, including presence of any anomalous systemic arterial supply that may suggest a "hybrid" lesion comprising mixed CPAM-PS components (see below).

Postnatally, chest radiograph is usually the first investigation performed, which may demonstrate signs of a CPAM, typically as a hyperlucent/multi-cystic region (particularly type 1 and 2 lesions),