Postnatal Imaging of Bronchopulmonary and Mediastinal Malformations

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Introduction

Obstetrical ultrasound has induced a marked increase in the perinatal diagnosis of broncho-pulmonary and mediastinal malformations. The spectrum of malformations amenable to antenatal diagnosis is wide (Table 1). The numerous classifications and terminologies attempt to incorporate a common origin and association of many lesions, including variable foregut airway, lung, and vascular components. Aside from the classical anomalies, such as congenital cystic adenomatoid malformations (CCAM), and sequestration, other, less common ones can also be considered as part of the spectrum (pulmonary hypoplasia, pulmonary aplasia, tracheal bronchus, etc.).

Some authors have suggested that many of the bronchopulmonary malformations (BPMs) represent an obstructive malformation sequence with secondary pulmonary dysplastic changes. Differences in the level, timing, and degree of obstruction are responsible for the spectrum of anomalies. The vascular origin of the anomalies has also been emphasized. Pulmonary hyperplasia as encountered in CCAM (type III) or in laryngeal atresia is one type of dysplastic change. Microcystic parenchymal dysplasia as seen in bronchial atresia, sequestration, or congenital lobar emphysema (CLE) is the other type that is encountered.

There have been several reports on the natural history of such anomalies. Furthermore, nowadays, neonatal management is widely influenced by the prenatal findings.

The aim of the present work is to define criteria for the postnatal work-up of patients with antenatally suspected BPM and to concisely detail the findings on imaging techniques (mainly chest X-ray and CT scan) that, even in asymptomatic newborns, point to the presence of this group of malformations [1-3].

Table 1. Spectrum of bronchopulmonary malformations

Congenital cystic adenomatoid malformation
Sequestration
Congenital lobar hyperplasia
Bronchial atresia
Pulmonary cysts
(Laryngeal atresia)

Bronchopulmonary Malformations

In Utero

Bronchopulmonary malformations are most often detected during the second trimester. Sonographically, the most common presentation at that stage is a hyperechoic space-occupying lesion that is either homogenous or contains small or large cysts. The mass causes a contralateral shift of the mediastinum. Since the spectrum of malformations is wide, a specific diagnosis cannot be made in utero unless a systemic vessel is demonstrated that vascularizes the lesion, suggesting the presence of a sequestration. However, at that stage, an exact differential diagnosis is not necessary, since many lesions are hybrid and include several different dysplastic tissues. Instead, the search for bilateral lesions and for other systemic malformations (e.g., skeletal, cardiovascular) is more important, as these will influence the in utero and postnatal prognoses. For instance, fetal hydrops and effusion may indicate a poor prognosis.

An important differential diagnosis to consider is diaphragmatic hernia. Its sonographic presentation may be confusing but the demonstration of small bowel loops, colon, and/or stomach in the thorax suggest the diagnosis.

The natural history of many BPMs consists of a relative (or absolute) decrease in volume such that the anomaly becomes increasingly difficult to recognize. This is especially the case when the mediastinum returns to the midline or the hyperechogenicity of the mass diminishes. Nonetheless, in either case it should not be assumed that the mass has completely disappeared [1, 2, 4-7].

A BPM should also be included in the differential diagnosis of a mediastinal mass (Table 2). Cystic lesions could correspond to a vascular malformation; therefore, the use of

Table 2. The differential diagnosis of a mediastinal mass in utero

Cystic lesions
Bronchogenic cyst
Duplication cyst
Neurenteric cyst
Vascular malformation
Cystic lymphangioma
Cystic teratoma
Solid/mixed type lesions
Mediastinal sequestration
Mediastinal teratoma

color Doppler is mandatory. Solid or mixed type lesions are more difficult to demonstrate and should suggest a mediastinal sequestration or a mediastinal teratoma [1, 2, 8, 9].

The Neonatal Period

In the neonatal period, the newborn affected by a BPM can be completely asymptomatic or present with respiratory distress due to airway compression. Noteworthy is the fact that the fluid within cystic malformations may rapidly be replaced by air and, due to air trapping, the cysts may subsequently enlarge. In such cases, asymptomatic newborns may acutely become symptomatic. For this reason and as a rule, all newborns with a prenatal diagnosis of BPM should undergo a neonatal work-up in order to determine the best treatment and follow-up. It should also be underlined that not all BPMs are detected prenatally; rather they can be an 'incidental' finding in a neonate [2-4].

Imaging Techniques and Findings

Chest X-ray

Chest X-ray is the basis of the neonatal work-up. Supine, frontal, and lateral views should be obtained during inspiration, while X-rays taken during expiration may be of interest in order to demonstrate air trapping. The potential findings on chest X-ray are listed in Table 3.

Table 3. Findings on chest X-rays in newborns with bronchopulmonary malformations

Abnormal aeration
Hyperaeration of an entire lung Æ asymmetric aeration
Hyperaeration of a lobe or segment
Consolidation
Partially aerated consolidation
Round nodular lesion
Partially aerated round lesion
Fluid air level
Completely aerated cyst (bleb)
Increased vascular markings
'Silhouette' sign at the level of the diaphragm
Enlarged mediastinum X-ray

Several of the findings may be associated. Also, when successive X-rays are obtained within the first few days of life, an evolution from one pattern to another can be observed. This evolution is usually a clue to the diagnosis and corresponds to resorption of the fluid contained in a lesion (i.e., pulmonary cyst, CLE, CCAM). Associated vertebral anomalies should also be searched for.

It should be noted that a normal-appearing chest X-ray does not mean complete resolution of the malformation or the absence of anomaly. Therefore, complementary CT or MR imaging should be done as well [1, 10].

Chest CT

Helical chest CT is the method of choice for imaging a BPM and is particularly accurate in determining areas of abnormal aeration, air fluid level, bilateral lesions, and abnormal vascularization. Pre- and post-contrast enhancement scans are mandatory as they demonstrate abnormal systemic arterial vascularization in case of sequestration as well as peripheral dysplastic venous drainage. 3D volume rendering also allows better mapping of the anomaly. CT provides essential information regarding bronchial atresia, bronchomalacia, and tracheal bronchus. Finally, CT reveals cystic lesions in any location (parenchymal vs. mediastinal) and allows densities within the lesions to be measured [1, 10-12].

Chest Magnetic Resonance Imaging

This technique provides information similar to that obtained with CT, especially concerning vascularization of the malformation. T1, T2, and echo-gradient sequences should be obtained in order to optimally assess the anomaly. MR imaging of the chest does not allow parenchymal assessment but it may demonstrate any extension of these lesion towards the spine and spinal canal [9, 11].

Ultrasound

Ultrasound may be helpful in demonstrating lesions close to the diaphragm and in the mediastinum. It is also able to demonstrate systemic vessels emerging from the aorta towards a sequestration as well as the anatomical relationship between a cystic lesion and other structures of the mediastinum [10, 13].

Upper GI Tract Opacification

Since BPM, especially sequestration, may involve the esophagus or stomach, in selected cases (e.g., persisting opacity in a lower lobe) an upper GI series may be useful [1, 2].

Angiogram

Angiograms are no longer performed unless embolization of a sequestration is being considered as a thera-

peutic option. They are also periodically used to evaluate patients with complex cardiovascular malformations [1, 2].

Conclusions

The antenatal diagnosis of BPM has modified our postnatal approach and management of the newborns affected.

A systematic work-up should be proposed even in asymptomatic patients.

Chest X-ray and helical Ct should be performed in order to determine the best treatment.

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